

**Conclusions:** Teaching knee aspiration to medical students is effective and feasible. Furthermore it improves junior doctor's confidence and translates into improved clinical practice. The possibility of extending this training across other medical schools should therefore be considered.

**Disclosure:** The authors have declared no conflicts of interest.

#### 437. ALL WALES PATIENT QUESTIONNAIRE: UNDERSTANDING OF ANTI-TNF THERAPY DECLINES AFTER THE FIRST YEAR OF TREATMENT

C. E. Page, B. Rhys-Dillon, S. J. Evans and U. Srinivasan  
*Rheumatology Department, Princess of Wales Hospital, Bridgend, United Kingdom*

**Background:** Patients being considered for anti-TNF therapy are given information on the goals and risks associated with therapy and on what to do if problems are experienced. We previously reported that despite a dedicated education programme, gaps in knowledge still exist. We report on the extension of this survey across Wales.

**Methods:** All Rheumatology patients in Wales receiving anti-TNF therapy (excluding 2 pilot hospitals) were sent anonymous true/false questionnaires about indications for their therapy, side effects and what to do if they had an infection or needed a surgical procedure. We analysed data received with particular reference to mode of administration and duration of therapy postulating that those on intravenous treatment may have benefited from frequent contact with the healthcare team and therefore score higher, as might those who had more prolonged time on treatment.

**Results:** 893 questionnaires were distributed with a 72% response rate. 632 replies were analysed: 384 (61%) Etanercept patients, 134 (21%) Adalimumab and 106 (17%) Infliximab. Duration of therapy: 310(49%)  $\geq 2$  years, 167 (26%) 1-2 years, 135 (21%)  $< 1$  year. Of the 9 questions analysed, 40% of respondents answered 8 or more correctly. Questions relating to indications for anti-TNF treatment had high rates of correct responses (77 - 91%) but questions about toxicity and infection gave more variable results. 35% of patients felt anti-TNF therapy was safe with no side-effects and 37% indicated that they would continue therapy (or were unsure) if they had an infection. Similarly, 37% were not aware that therapy should be stopped prior to surgery. There was no statistical difference in responses between those on subcutaneous or intravenous treatment. However, there was a significant difference in the number of correct answers with respect to duration of treatment. A statistically higher proportion of patients on treatment for less than one year scored 8 or more correct answers ( $p < 0.001$ ). This difference at 1 year was particularly apparent with questions relating to toxicity. ( $p < 0.04$ ) and was lost when comparing treatment duration more or less than 2 years.

**Conclusions:** We had a high response rate in this national survey. Patients are well informed of the goals of anti-TNF therapy. However a significant proportion (37%) did not recognise active infection as a reason for temporary cessation of therapy and a similar number of respondents felt there were no significant risks associated with treatment. Reassuringly, patients receiving subcutaneous therapy were as well informed as those on intravenous therapy. Our results suggests that recall of knowledge (particularly in relation to infection) appears to decline after 12 months of treatment and further educational sessions at this point may address this issue and promote optimal knowledge retention.

**Disclosure:** The authors have declared no conflicts of interest.

#### 438. POSTGRADUATE PAEDIATRIC RHEUMATOLOGY (PRh) TRAINING WITHIN PAEDIATRICS IN THE UK

Sharmila Jandial and Helen E. Foster  
*Musculoskeletal Research Group, Newcastle University, Newcastle upon Tyne, United Kingdom*

**Background:** The PRh clinical service in the UK is currently successfully delivered in many areas by adult rheumatologists with training in PRh, working in clinical networks with PRh multidisciplinary teams. However, current adult rheumatology training does not include PRh - future delivery of PRh clinical service in the UK relies on adequate PRh trainees being recruited from within paediatrics. Specialist training within UK paediatrics follows a structure comprising 'Core' training (two years, competency based framework [www.RCPCH.ac.uk]) before progressing to a competitive entry point for "Grid" speciality training (usually three years) The aim of this study was to establish the extent of PRh exposure for trainees within general paediatrics, as exposure is likely to influence future sub-speciality career choice.

**Methods:** All national programme directors in paediatrics ( $n = 17$ ) were contacted by email and invited to complete a web-based anonymised questionnaire about PRh training in their Region.

**Results:** The response rate was 14/17 (82%). The median number of Specialist Registrars (SpRs) in general paediatrics per Region was 64 (range 36 - 110). Commonest specialities offered to SpRs were endocrinology, neurology, respiratory and oncology ( $n = 13$ , 93%). Less commonly offered were adolescent medicine or clinical pharmacology ( $n = 1$ , 7%), metabolic medicine ( $n = 2$ , 14%), education or immunology ( $n = 3$ , 21%), and PRh ( $n = 4$ , 28%). Trainees were invariably not offered choice in their speciality placements in Core training ( $n = 5$ , 36%). PRh training was provided at 'Core' in 4 centres (28%) and post 'Core' at 8 centres (57%) all of which were tertiary hospitals. Three Regions (21%) permitted trainees to pursue PRh training outwith their given Region. Notably only three Regions had candidates apply for 'Grid' training in PRh and all had provided PRh experience in Core training. Within Regional teaching programmes for SpRs, PRh was included in

10/12; taught by general paediatricians ( $n = 4$ ), paediatric rheumatologists ( $n = 5$ ) and adult rheumatologists ( $n = 3$ ).

**Conclusions:** Opportunities for PRh training within general paediatrics are limited and confined to PRh tertiary centres. The RCPCH core competency framework for all paediatricians includes musculoskeletal medicine and few Regions in the UK offer appropriate training. This is likely to adversely affect recruitment to the PRh sub-speciality & the achievement of core competencies required by all paediatricians. Given that future adult rheumatologists will not be trained to deliver PRh services as at present, this study suggests a shortfall in paediatric trainees in PRh with major implications for manpower planning for the future provision of PRh services in the UK. This needs to be addressed both in general paediatric training and by adult rheumatology services.

**Disclosure:** The authors have declared no conflicts of interest.

## Case Reports (I)

#### 439. HYPERTROPHIC PULMONARY OSTEOARTHROPATHY MIMICKING ATYPICAL RHEUMATOID

Sarah E. Medley and A. L. Dolan  
*Rheumatology, Queen Elizabeth Hospital, Woolwich, United Kingdom*

**Background:** A 52 year old lady presented to orthopaedics with arthralgia affecting knees, shins and wrists. She was arthroscoped and synovial biopsy of the right knee revealed no synovitis. Ongoing arthralgia was diagnosed as rheumatoid arthritis and treated with monotherapy methotrexate and then sulphasalazine with added prednisolone. IM steroid was ineffective. A second diagnosis of facet joint arthritis in part explained her need for additional tramadol, meloxicam and gabapentin for pain control. Rheumatoid factor was negative and inflammatory markers rose from ESR 27 to 101 during treatment. This and her pain, contributed to a very high Disease Activity Score of 8.2. She was a smoker of at least 45 pack years.

**Methods:** Her symptoms failed to respond to treatment and TNF was planned. Only then was a chest x-ray ordered, which revealed a large right apex mass, which proved to be a squamous carcinoma of the lung. Subsequent resection resulted in rapid and significant improvement in her articular symptoms.

**Results:** Her rheumatological care was subsequently transferred to our hospital. All medication except analgesics had been stopped as she was now receiving adjuvant chemotherapy. On examination there was clubbing but no synovitis. There were no tender joints in the upper limbs, but tenderness over the shins and the metatarsophalangeal joints persisted. Radiographs showed metacarpal shaft thickening in the hands compatible with hypertrophic pulmonary osteoarthropathy (HPOA). Periosteal reactions were also evident on x-ray of the tibia, and metatarsals. There were no erosions.

**Conclusions:** The possibility of HPOA had been raised by an earlier nuclear medicine bone scan, but this was only in a final report that took over 3 months to be authorised. A preliminary report had not suggested this possibility.

This case illustrates the need for a chest x-ray in new RA. The diagnosis was atypical with negative rheumatoid factor and synovial biopsy. Pain and a raised ESR influenced DAS unduly. Atypical unresponsive RA requires review of diagnosis rather than just treatment escalation.

Hypertrophic osteoarthropathy is rare, but may be primary or secondary. In the case of malignant lung tumors, arthropathy with synovitis may present in advance of clubbing. This case summarises a case of inflammatory arthritis, which was unresponsive to standard therapy. The subsequent findings of clubbing in a smoker, and radiographic changes made the diagnosis of HPOA likely. The resolution of articular symptoms post lung resection was striking.

**Disclosure:** The authors have declared no conflicts of interest.

#### 440. AN UNUSUAL CASE OF HEPATIC OSTEODYSTROPHY

Sathish Kallankara<sup>1</sup> and Tim Gillott<sup>2</sup>  
<sup>1</sup>Department of Rheumatology, Diana, Princess of Wales Hospital, Grimsby, United Kingdom and <sup>2</sup>Department of Rheumatology, Diana, Princess of Wales Hospital, Grimsby, United Kingdom

**Background:** Hepatic osteodystrophy (HOD) refers to a combination of osteoporosis and osteomalacia occurring in chronic liver disease. HOD occurs most frequently with longstanding cholestasis and is common in primary biliary cirrhosis (PBC). It is very infrequent at presentation in PBC.

**Methods:** We present an interesting case of hepatic osteodystrophy associated with PBC, in which the patient presented with osteoporotic fracture well ahead of clinical presentation of PBC.

**Results:** A 63 year old lady presented in November 2003 with low back ache and loss of height. She did not have any clinical fracture or other systemic symptoms. She attained menopause at 55 years and did not have any other risk factors for osteoporosis. She denied any personal or family history of significant medical diseases. Clinical examination was unremarkable except for marked kyphosis.

X-ray revealed compression fractures of several dorsal and lumbar vertebrae. DEXA scan showed osteoporotic spine and osteopaenic femoral head. Her base line blood tests were normal including the liver function tests (LFT). She was started on once weekly Risedronate with Adcal D3 in January 2004. In November 2004 her

routine 3 monthly blood tests revealed abnormal LFTs (ALT - 91 IU/L, Alkaline Phosphatase (AP) - 238 IU/L).

Further tests revealed normal TFT, PTH, N-Terminal Telopeptide levels and protein electrophoresis. Hepatitis profile and Coeliac screen were negative. 25 - hydroxy vitamin D(25-OH D3) level was 25 units which was considerably low even after correcting for weather conditions. Autoantibody profile showed positive anti mitochondrial antibody IgG(1:640) and anti smooth muscle antibody. Ultrasonography of liver was normal.

She was seen by gastroenterologist who started her on Ursodeoxycholic acid (UDCA) in October 2005. She remains clinically stable with no fractures and the latest blood reports showed an ALT of 22, AP 88 and 25 -OH D3 level of 96.

**Conclusions:** HOD can develop even in the pre-cholestatic phase of PBC. Ursodeoxycholic acid which is the corner stone for treatment of PBC can lead to better outcome in HOD.

**Disclosure:** The authors have declared no conflicts of interest.

#### 441. PRIMARY SEPTIC ARTHRITIS OF THE ACROMIO-CLAVICULAR JOINT

Peter Raite<sup>1</sup>, Karthikeyan P. Iyengar<sup>1</sup>, Shashank Chitgopkar<sup>1</sup>, Jayant B. Nadkarni<sup>1</sup>, Peter Hughes<sup>2</sup> and William Y. Loh<sup>1</sup>  
<sup>1</sup>Orthopaedics, Southport and Ormskirk NHS Trust, Southport, United Kingdom and <sup>2</sup>Radiology, Southport and Ormskirk NHS Trust, Southport, United Kingdom

**Background:** Primary septic arthritis of the A-C joint is an uncommon disorder and is rarely seen even in the immuno-compromised individual. We report a case of de novo primary septic arthritis of the acromio-clavicular joint (A-C) caused by *Staphylococcus aureus*. This has not been previously reported. Risk factors for septic arthritis include local conditions like pre-existing joint disease (e.g. trauma, osteoarthritis), systemic vulnerability of the host to infection due to disease or medication.

**Methods:** A fit and healthy 42 year old right handed male farmer presented with a 2 day history of sudden onset left shoulder pain. This was associated with fever and restricted shoulder movements.

Local examination revealed a diffuse swelling and tenderness of the A-C joint. The shoulder movements were restricted. He had a raised CRP of 274mg/L, ESR of 50mm/h with a neutrophil leucocytosis. Plain radiographs of the left shoulder revealed small erosion at the superior aspect of left clavicle with no evidence of periosteal reaction. Magnetic Resonance Imaging (MRI) scan revealed marked soft tissue swelling centered on the A-C joint. These features were enhanced following Gadolinium contrast injection. Computerized axial scan (CT) showed cartilaginous and bony erosions of the AC joint. Both synovial fluid aspirate and blood cultures confirmed a heavy growth of *Staphylococcus aureus*.

**Results:** He was treated successfully with IV Flucloxacillin (2g qds) and oral Fusidic acid (0.5g tds) for 6 weeks. He regained full function of his left shoulder at 3 months with a Constant Shoulder Score of 100%. A follow-up MRI scan and CT scan showed resolution of the acute infection.

**Conclusions:** History and clinical examination is vital in suspecting a diagnosis of A-C joint infection and can help to differentiate it from gleno-humeral arthritis. However arthrocentesis remains the gold standard for diagnosis of septic joint. Imaging generally plays an adjunctive role. MRI is superior to ultrasonography and can detect a septic joint as early as 24 hrs after infection.

Early MRI and CT investigations and a positive joint aspirate allowed accurate diagnosis and prompt treatment of this unusual primary septic arthritis of the A-C joint.

**Disclosure:** The authors have declared no conflicts of interest.

#### 442. A PARANEOPlastic PRESENTATION OF RAPIDLY PROGRESSIVE SYSTEMIC SCLEROSIS

Sarah J. Evans and Margaret M. O'Sullivan  
*Rheumatology, North East Wales NHS Trust, Wrexham, United Kingdom*

**Background:** Systemic sclerosis (SSc) can present as a paraneoplastic manifestation with a reported incidence of between 3% and 7%.

We present a patient with a silent lung carcinoma presenting with diffuse SSc. **Methods:** A 67-yr old male smoker with advanced chronic obstructive pulmonary disease presented with a six month history of Raynauds and progressive skin tightness affecting all of his body. He had constitutional symptoms of malaise, anorexia and significant weight loss. There were no progressive respiratory symptoms.

Physical examination revealed widespread scleroderma affecting his face, limbs and trunk. There were marked flexion contractures of his fingers but no digital ulceration, clubbing, and no palpable lymphadenopathy. Chest examination revealed fine end inspiratory crackles at both bases. Cardiovascular and neurological systems were unremarkable.

**Results:** Investigations revealed a low serum albumin of 32 g/l (normal range 35-50). CRP, plasma viscosity, and renal function were normal. ANA was positive (mixed nucleolar/speckled pattern, titer 1:300) and Scl-70 was positive. There was no proteinuria, haematuria or urinary casts. CT chest showed an irregular spiculated mass in the left lower lobe suggestive of malignancy. A positron emission tomography (PET) scan was arranged to guide a biopsy site at bronchoscopy. A combined PET/CT scan showed a markedly avid lesion in the left lower lobe with increased radionuclide uptake of mediastinal and hilar lymph nodes. Basal pulmonary fibrosis was also noted, with ground glass opacification and increased 18 F-fluorodeoxyglucose (FDG) uptake consistent with active

inflammation. Bronchoscopy was carried out and biopsy confirmed squamous cell carcinoma which was inoperable.

Three intravenous pulses of 1g methylprednisolone were given followed by prednisolone 10mg daily alleviating the constitutional symptoms. One month later he developed painful ischaemic ulcers on his fingertips and elbows. There was a poor response to iloprost and he subsequently had a stellate ganglion block with some improvement.

**Conclusions:** This case illustrates that SSc can follow a rapidly progressive course, and when associated with a short history in an older patient with prominent constitutional symptoms, the presence of underlying neoplasia should be suspected.

**Disclosure:** The authors have declared no conflicts of interest.

#### 443. BEAUTY AND THE BEAST; RECALCITRANT PSORIASIS INDUCED BY ETANERCEPT

Batsi Chikura, Vipul Vagadia and Eddie J. Tunn  
*Rheumatology, Royal Liverpool University Hospitals, Liverpool, United Kingdom*

**Background:** Etanercept is a tumour necrosis factor (TNF) blocker and is effective in treating psoriatic arthritis and psoriasis. Paradoxically it can induce psoriasis which usually resolve after treatment discontinuation. We wish to report a case of severe palmo-plantar psoriasis induced by etanercept and resistant to treatment after withdrawal of etanercept.

**Methods:** A 63 year old female patient with a 40 year history of seropositive rheumatoid arthritis presented with a psoriatic eruption 6 months after initiation of etanercept therapy. She had no previous personal or family history of psoriasis. In August 2005 after failing methotrexate, sulfasalazine and leflunomide, she was started on etanercept following the British Rheumatology Society (BSR) guidelines. She presented with a psoriatic eruption mainly on her hands, lower limbs and feet. The diagnosis of psoriasis was confirmed by a dermatologist and histopathological examination.

She continued with etanercept at the same time being treated for psoriasis by the dermatologists using the standard therapies for psoriasis. Her psoriasis did not improve. Etanercept was discontinued and the rash disappeared everywhere else except for the soles of her feet. Her rheumatoid arthritis flared up four months after etanercept was discontinued and a trial of reintroduction of etanercept led to a flare-up of her psoriasis in other parts of her body. Etanercept was discontinued for the second time and once again the psoriasis disappeared everywhere else except the soles of her feet.

The psoriatic plaques under her feet have been resistant to all forms of treatment currently available for treatment of severe psoriasis.

**Results:** Pirard et al reviewed in 2006 thirty cases of psoriasis induced by anti-TNF therapy and concluded that the psoriasis was self limiting and did not require treatment discontinuation. Skin lesions usually resolve after discontinuation of anti-TNF therapy and patients respond to basic treatments such as topical creams. This certainly is not our experience.

Infliximab, adalimumab and etanercept therapies are effective in the treatment of psoriasis. Paradoxically, anti-TNF therapy can induce psoriasis and this observation begs the question as to the role of TNF in the pathogenesis of psoriasis? Anti-TNF therapy can also exacerbate existing psoriasis. Interestingly, anti-TNF therapy can also induce and treat lung fibrosis. We find these paradoxical effects intriguing. All anti-TNF therapies have been associated with the induction of psoriasis and we can conclude that this is a class effect.

**Conclusions:**

1) We feel that there is a need to raise awareness of anti-TNF induced recalcitrant psoriasis.

2) The incidence of this adverse drug reaction, the risk factors and the mechanisms by which anti-TNF therapy induces psoriasis are unknown and needs investigating.

3) We find the paradoxical effects of anti-TNF intriguing.

**Disclosure:** The authors have declared no conflicts of interest.

#### 444. WAS IT THE CURRY? AN UNUSUAL VASCULAR COMPLICATION OF SLE

Dimitra Doufexi<sup>1</sup>, Mark Lloyd<sup>2</sup>, Peter Clarkson<sup>3</sup> and Andrew Hatrick<sup>4</sup>  
<sup>1</sup>Medicine, Frimley Park Hospital NHS Foundation Trust, Frimley, Camberley, United Kingdom, <sup>2</sup>Rheumatology, Frimley Park Hospital NHS Foundation Trust, Frimley, Camberley, United Kingdom, <sup>3</sup>Cardiology, Frimley Park Hospital NHS Foundation Trust, Frimley, Camberley, United Kingdom and <sup>4</sup>Radiology, Frimley Park Hospital NHS Foundation Trust, Frimley, Camberley, United Kingdom

**Background:** Renal artery stenosis (RAS) is a recognised complication of systemic lupus erythematosus (SLE). We report a case of SLE in which RAS presented as life-threatening flash pulmonary oedema. To our knowledge this is the first time this rare complication of RAS has been described in SLE.

**Methods:** Case: A 53 year old female developed a sudden onset of chest and abdominal pain, dyspnoea and wheeze following a meal in an Indian restaurant. On admission she rapidly deteriorated and required intubation and ventilation. She was an ex-smoker with a longstanding history of SLE (rash, joint pain, ANA +ve, SSA +ve, aCl -ve). She was known to have ischaemic heart disease (stent insertion 2004 and 2006) and emphysema. There was no history of hypertension. Rx: MMF 1g bd, prednisolone 10mg od, atorvastatin, aspirin, diclofenac.

Her pre-admission exercise tolerance was 20m for the past 3 years (FEV<sub>1</sub> 1.0). **Investigations:** CXR: severe pulmonary oedema. ECG: low voltage complexes. Catheter studies: CO 2.0, PCWP 24, SVRI 5060 (indicating low output and high vascular resistance); troponin I 0.42 (high); CRP 25; TTECHO: good LV function (EF ~ 50%), no significant valvular lesion, normal dimensions; CTPA: no PE, L pleural effusion and bibasal atelectasis. CT head, abdominal US normal.

**Results:** She remained in ITU for 12 days, initially causing diagnostic confusion. Initial treatment was furosemide, inotropes and an increase in steroids to cover possible lupus myocarditis. Troponin fell to normal. Later MRA showed stenosis on the proximal R renal artery, infrarenal aortic disease and focal stenoses on L common and L external iliac arteries. Coronary angiography demonstrated multiple vessel disease worse on R coronary artery (85% complex stenosis). She was discharged on day 17, and 2 days later had R renal artery stent insertion; 1 month later, R coronary artery re-stent. Her exercise tolerance since then has improved considerably, allowing her to go back to work.

**Conclusions:** The mechanisms causing flash pulmonary oedema in RAS are poorly understood. In patients with concomitant hypertension diastolic dysfunction is thought to be important; labile hypertension and acute salt and water retention are also thought to play a role. Our case is unusual in that there was no preceding history of hypertension and her RAS was unilateral. The importance of looking for less common vascular complications of SLE, and of aggressively managing vascular risk factors is emphasised.

**Disclosure:** The authors have declared no conflicts of interest.

#### References

- Manzi S et al, Am J Epidemiol 1997 Mar 1;145(5):408–15.
- Pickering TG et al, Lancet 1988 Sep 3;2(8610):551–2.

#### 445. BEWARE ANGIOEDEMA WITH ETANERCEPT

Sarah Moore<sup>1</sup>, David Palmer<sup>2</sup> and George Kallarackal<sup>1</sup>

<sup>1</sup>Rheumatology, Kettering General Hospital, Kettering, United Kingdom and

<sup>2</sup>General Practice, Forest Gate Road surgery, Corby, United Kingdom

**Background:** A 44 year old lady with a seven year history of seropositive rheumatoid arthritis was commenced on etanercept following the failure of disease control with three different disease modifying agents.

**Methods:** Two days after her third etanercept injection she awoke with severe facial swelling. She had no symptoms or signs of airway compromise.

**Results:** Her etanercept was stopped and her general practitioner gave her a short course of oral steroids and anti-histamines. The swelling resolved gradually over a number of days.

**Conclusions:** Many drugs have previously been linked with angioedema including ACE-inhibitors and NSAIDs. It is important for clinicians to be aware of this reaction in patients being commenced on etanercept as angioedema is potentially life threatening. Similar reactions have been reported with anti-TNF agents and we feel that it is important to highlight this issue to the rheumatology community.

**Disclosure:** The authors have declared no conflicts of interest.

#### 446. A CASE OF FOCAL NEUROMYOTONIA ASSOCIATED WITH RECURRENT ANGIOEDEMA/URTICARIA

Elena Nikiphorou, Tanya Baqai and Ali S. Jawad

Rheumatology, Barts & The London NHS Trust, The Royal London Hospital, London, United Kingdom

**Background:** Neuromyotonia is a rare condition which lies in the spectrum of disorders of peripheral nerve hyperexcitability. It involves dysfunction of Voltage-Gated-Potassium-Channels and can be classified as one of the channelopathies.

**Methods:** A 65-year-old white man initially presented with disabling distal limb muscle cramps and visible myokymia. It coincided with the onset of urticaria and angioedema. Subsequent attacks of muscle cramps occurred with relapse of the urticaria/angioedema. Electromyography confirmed the presence of frequent spontaneous fasciculation potentials and cramp discharges but without fibrillation potentials and no evidence of inducible discharges on repetitive stimulation. The features were supportive of peripheral nerve hyperexcitability. Voltage-gated potassium, calcium and sodium channel and serum anti-GAD antibodies were persistently negative, with normal complement and immunoglobulin levels. No other auto-immune serology was identified. A diagnosis of focal neuromyotonia was made. MRI of the lower limbs revealed symmetrical bilateral subcutaneous oedema along the fascia of the lower leg musculature without real oedema of the muscles. A paraneoplastic process was ruled out by negative CT imaging and serological antibody tests.

**Results:** He had initial good response to low dose phenytoin and acute attacks have been terminated by intravenous diazepam. His course however had been complicated by suboptimal response or gastrointestinal intolerance to carbamazepine, gabapentin, pregabalin, baclofen, amitriptyline and antihistamines, including cetirizine and chlorpheniramine. He now has progression in attack frequency and distribution of muscle involvement to involve pharyngeal, laryngeal and possibly trunk musculature, despite the above treatments (images and video clip available).

**Conclusions:** The clinical features and electromyography findings are diagnostic of focal neuromyotonia. Having no identified autoimmune serology, as literature suggests is often the case, he poses certain management dilemmas as regards second line immunomodulatory treatments. Few anecdotal reports have suggested benefit from use of immunosuppressant agents, IVIG and plasma exchange.

We currently propose the use of Rituximab through its properties of B lymphocyte depletion.

**Disclosure:** All authors have declared no conflicts of interest.

#### 447. A RARE CASE OF MULTIFOCAL PIGMENTED VILLONODULAR SYNOVITIS PRESENTING IN AN ADULT

Venkat Reddy, Ferhat Uddin and Ian Chikanza

Rheumatology, Barts and the London NHS Trust, London, United Kingdom

**Background:** Pigmented villonodular synovitis (PVNS) is a benign proliferative disease of the synovium that affects joints, bursae or tendon sheaths. Common presenting complaints are pain, swelling, mechanical derangement and decreased range of movement. The most commonly affected joints are the knee (80%), hip and ankle.

The common diffuse form of PVNS usually presents in the 3rd or 4th decades of life, and is almost always unilateral and monoarticular.

Focal PVNS commonly occurs in the 5th or 6th decades of life and affects joints or tendon sheath. It has a strong female predilection, affects the digits of the hand or foot, and presents as a slow-growing painless mass.

Multifocal PVNS has only been reported to occur in children very rarely. To our knowledge multifocal PVNS has not yet been reported in an adult.

**Methods:** A 49 yr old Bangladeshi gentleman had first presented at the age of 38 with a bilateral painful stiff wrists and knees. He worked as a chef and involved standing on his feet for long hours.

Clinical examination of the right knee revealed crepitus, synovial thickening and prominent osteophyte. Flexion of the wrist was limited to ten degrees.

The patient had also been diagnosed with pleural and peritoneal tuberculosis.

**Results:** Full blood count, urea, electrolytes, liver function tests and inflammatory markers were normal.

X-Rays revealed changes of osteoarthritis in the right knee and advanced osteoarthritic changes and collapse of the proximal row of carpal bones in the left wrist.

Arthroscopy of the right knee revealed synovial thickening and extensive proliferative villous vascular synovium.

MRI of the left wrist showed an effusion in the radio-ulna joint, with low signal areas due to haemosiderin. Bony erosions were seen in the ulna, distal radius and in the carpal bones especially the scaphoid and triquetrum.

MRI of the right knee showed synovial thickening, moderate effusion with low signal areas due to haemosiderin deposits, but no bony erosions.

Histological features of synovial tissue from the wrist and knee revealed villous synovial tissue, diffuse chronic inflammatory cell infiltrate rich in plasma cells and marked synovial hyperplasia with surface fibrin deposition. Collections of haemosiderin laden macrophages were noted. Appearances are those of a chronic synovitis with features suggestive of PVNS.

On follow up after eight years since synovectomy no recurrence of symptoms were noted.

**Conclusions:** Multifocal PVNS occurring in an adult is very rare. MRI findings may be diagnostic. Synovial biopsies of the involved joints may be necessary to secure this rare diagnosis. Synovectomy of multiple joints may be required.

**Disclosure:** The authors have declared no conflicts of interest.

#### 448. EXACERBATION OF PSORIASIS AND PSORIATIC ARTHRITIS INDUCED BY INTERFERON THERAPY

Aruna s. Malipeddi<sup>1</sup> and George Kallarackal<sup>2</sup>

<sup>1</sup>Rheumatology, University Hospitals of Leicester, Leicester,

United Kingdom and <sup>2</sup>Rheumatology, Kettering General Hospital, Kettering, United Kingdom

**Background:** Interferon is an immuno-modulator used in the treatment of various medical conditions. Although rare there have been reports of exacerbation or occurrence of autoimmune conditions following its use. We present a case of psoriasis and psoriatic arthritis exacerbated by interferon therapy.

**Methods:** A 43 year-old man presented with severe exacerbation of psoriasis and psoriatic arthritis for a week with worsening of pain and swelling in his hands and knees. On examination he had active synovitis of the small joints of the hands and knees and extensive psoriatic plaques. The erythrocyte sedimentation rate was 73 on admission.

He was diagnosed with psoriasis as a child and psoriatic arthritis for 10 years both of which were controlled on methotrexate 17.5milligrams/week. He was diagnosed with hepatitis C 3 years previously. He was started on interferon and ribavirin. 7 weeks after starting the interferon he noted exacerbation of psoriasis and psoriatic arthritis. Interferon and ribavirin were stopped and he was treated with intensive topical therapy with betnovate and Lassar's paste for psoriasis and increasing the dose of methotrexate for psoriatic arthritis, following which his symptoms improved.

**Results:** Interferon is used for various medical conditions including hepatitis. It's use has been associated with occurrence of autoimmune phenomenon including autoimmune thyroiditis, thrombocytopenia, anaemia, sarcoidosis, exacerbation of psoriasis and psoriatic arthritis (1-6). The exact mechanism is unknown. Withdrawal of interferon, topical treatment for psoriasis and steroids for psoriatic arthritis leads to improvement.

**Conclusions:** Interferon is not contraindicated in patients with psoriasis and psoriatic arthritis but may aggravate the condition. All patients should be counselled

for potential exacerbation prior to starting interferon. Symptoms usually subside following withdrawal of interferon.

**Disclosure:** The authors have declared no conflicts of interest.

#### References

1. C Taylor, DA Burns, MJ Wiselka. Extensive psoriasis induced by interferon alfa treatment for chronic hepatitis C. *Postgrad Med J* 2000; 76: 365-67.
2. Makino Y, Tanaka H, Nakamura K, Fujita M, Akiyama K, Makino I. Arthritis in a patient with psoriasis after interferon - alpha therapy for chronic hepatitis C. *J Rheumatol* 1994 Sep; 21(9): 1771-2.
3. Burman P, Karlsson FA, Oberg K, Alm G. autoimmune thyroid disease in interferon-treated patients. *Lancet* 1985;63:1086-90.
4. Abdi EA, Gia-Khanh N, Ludwig RN, Dickout WJ. Pulmonary sarcoidosis following interferon therapy for advanced renal cell cancer. *Cancer* 1987;59:896-900.
5. Akard LP, Hoffman R, Elias L, Saiers JH. Alpha-interferon and immune hemolytic anemia. *Ann Intern Med* 1986;105:306.
6. Abdi EA, Venner PM. Immune thrombocytopenia after  $\alpha$ -interferon therapy in patients with cancer. *JAMA* 1986;255:1878-79.

#### 449. A CASE OF SEVERE LEG ULCERS IN A PATIENT WITH RHEUMATOID ARTHRITIS WHICH IMPROVED ON STOPPING LEFLUNOMIDE

Susie C. Earl and Peter J. Prouse  
*Rheumatology, North Hampshire Hospital, Basingstoke, United Kingdom*

**Background:** Patients with rheumatoid arthritis (RA) are predisposed to developing chronic leg ulcers (1,2). The causation of leg ulcers in patients with RA is multifactorial, including vasculitis, venous insufficiency and Felty's syndrome (3).

**Methods:** Mrs VD is a 57 year old Caucasian female with aggressive RA and severe leg ulcers. She also suffers from premature cardiac disease, type II diabetes, and marked lipodystrophy. RA was diagnosed in 1990 and was initially well managed on methotrexate but over the course of a few years became very nodular and increasingly aggressive. She failed to respond to methotrexate and sulphasalazine and both drugs were stopped in 2000. Leflunomide was commenced in 2000 initially 40mg daily but then reduced to 20mg due to pancytopenia. In 2003, Mrs VD developed severe bilateral leg ulcers and in 2004 had failed split skin grafts to both ulcers within days of the grafting. Her RA was increasingly active and she was considered for biological agents but unfortunately her infection risk was felt to be too high. She continued with leflunomide. By 2007, the leg ulcers became increasingly necrotic (see photos) and this prompted her admission to the North Hampshire Hospital initially under the care of the general surgeons as her leg ulcers were so severe that she was considered for a left below knee amputation. The leg ulcers were debrided and vac pumps applied. Examination revealed widespread symmetrical nodules with a number of vasculitic lesions. She had classical rheumatoid changes affecting the hands and feet. Bilateral necrotic leg ulcers were present over the lateral malleoli with the most severe changes affecting the left leg (see photos). Peripheral pulses were intact.

**Results:** Investigations: Blood tests - Hb 10.2, WCC 8.3, neut 5.4, ESR 37, normal U&Es/LFTs, CRP 39. Immunology - Rheumatoid factor 640, anti CCP antibodies 161, ANA negative, cryoglobulins normal, anti-cardiolipin antibodies negative. Xrays - see images. USS abdomen - splenomegaly (15.5cm). Arteriogram - Mild arterial disease in infra popliteal vessels with distal small vessel disease consistent with diabetic type disease. Skin biopsy - no evidence of vasculitis.

Leflunomide was stopped due to case reports (4,5) suggesting that it may worsen leg ulcers and a 10 day washout with cholestyramine was commenced. Four weeks after the washout period, the leg ulcers were improving and the vasculitic lesions started to resolve. Her joints flared and a combination of prednisolone 7.5mg daily and azathioprine (dose escalated to 200mg daily) was commenced. In September 2007, Mrs VD underwent split skin grafts to both leg ulcers with very good results. Mrs VD is now being considered for biological agents.

**Conclusions:** This case highlights a difficult case of RA complicated by severe leg ulcers which improved upon stopping leflunomide.

**Disclosure:** The authors have declared no conflicts of interest.

#### 450. ASYMPTOMATIC INTRA CARDIAC MASS (ICM) & ANTI-PHOSPHOLIPID SYNDROME (APL) - A RARE ASSOCIATION?

Mytheen S. JasminSajna and Kuntal Chakravarty  
*Rheumatology, Queen's Hospital, Romford, United Kingdom*

**Background:** Presence of an ICM, as revealed on echocardiography, can be a diagnostic challenge. Most often such findings suggest a neoplastic lesion arising from the atrium or the ventricle. We describe a case of a young woman who was being investigated for infertility with an incidental finding of elevated apL and an asymptomatic intracardiac mass, which disappeared without surgery.

**Methods:** A 30- year old married woman was being investigated for suspected infertility when she was found to have a prolonged activated partial thromboplastin time. She admitted to have suffered from Raynaud's phenomenon for over a decade without any history of thrombo-embolic disease or miscarriage. Further

evaluation revealed that she had a transient history of pleuritic chest pain with no associated dyspnoea.

Physical examination was normal apart from reduced air entry in the right lung base with a possible pleural rub. Her chest x-ray showed right basal shadowing and computerised tomography pulmonary angiogram confirmed pulmonary embolism of right lower lobe.

**Results:** Further immunological investigations showed positive antinuclear factor with normal double stranded DNA. IgG acL and IgM acL were >100(0-9.4) and 24.3(0-10.7) respectively, associated with positive lupus anticoagulant (LAC percentage correction-47.8(0-9.9),corrected partial thromboplastin time 45.8(24.0-38.0) and positive anti-SSA antibody. She was commenced on low molecular weight heparin, warfarin and low dose Aspirin. A transthoracic echocardiogram suggested an ill-defined mass in the right atrium. A trans oesophageal echocardiogram confirmed a mass at the junction of the inferior vena cava with the right atrium close to the base of the tricuspid valve. Further investigations including a cardiac MRI showed a cardiac mass. The diagnosis remained unclear suggesting the possibility of an embryonic remnant with or without an intracardiac thrombus.

In view of the history of apL she was advised to continue on high dose warfarin and aspirin. A repeat cardiac MRI, six months later showed marked reduction in the size of the mass suggesting that the ICM was more likely to be a thrombus than a neoplasm. She has now been advised to be on life long anticoagulation.

**Conclusions:** Cardiac manifestations of apL syndrome range from valvular and coronary artery disease to cardiomyopathy and intracardiac thrombosis. ICM due to intra-cardiac thrombosis may cause significant diagnostic confusion. It is not uncommon for such patients to be subjected to surgery with attendant risks. A greater awareness is necessary of such rare echo-cardiographic finding in apL syndrome as patients may be spared of unnecessary surgery.

**Disclosure:** The authors have declared no conflicts of interest.

#### 451. ASYMMETRIC NODAL OSTEOARTHRITIS IN THE CONTEXT OF OLD POLIO

Kieran P. Nunn<sup>1</sup>, Michelle L. Lee<sup>1</sup> and Louise Dolan<sup>2</sup>  
<sup>1</sup>*School of Medicine, Guy's, King's & St Thomas' Hospital's, London, United Kingdom* and <sup>2</sup>*Department of Rheumatology, Queen Elizabeth Hospital, London, United Kingdom*

**Background:** This atypical case of asymmetrical presentation of bilateral arthropathy gives insight into the factors influencing symmetry.

During an introductory education in Rheumatology, we learn about the key features of common disease states, such as the bilateral arthropathies including rheumatoid arthritis (RA) and osteoarthritis (OA). This marks the extent of many doctors' education in rheumatology, this case is an informative demonstration of how patients differ from textbook cases.

This patient presented with unilateral nodal OA in the context of existing polio-induced hemiplegia. Current understanding describes two aetiological theories for the unilateral presentation of a bilateral arthropathy; neurogenic and immobilisation theories.

**Methods:** A 74-year-old woman presented with a 5-year history of right hand pain and deformity of the right hand distal inter-phalangeal joints (DIPJ) which had become increasingly painful. She complained of approximately 30 minutes of morning stiffness and new inability at previous tasks such as knitting. Poliomyelitis was diagnosed aged 4 years, resulting in a flaccid left arm. Her arthritic complaint interestingly spared this limb. On examination there were marked Heberden's nodes of the right DIPJs and a weak hypotonic left arm with a comparatively smaller hand.

**Results:** Blood results excluded underlying inflammation (ESR 5, RF and autoantibody screen negative) and hand X-ray was consistent with OA change of the DIPJ on the right.

**Conclusions:** This case stimulated an analysis of the neurogenic and immobilisation theories of a spared limb in bilateral arthropathy. It appears that paralysis reduces penetrance of an autosomal dominant condition because Heberden's nodes are not use-related, and current evidence is in favour of the neurogenic theory.

The 'bilateral arthropathies' may present unilaterally in the context of pre-existing hemiplegia. Neurogenic and immobilisation theories exist to explain this presentation, but currently neither is substantiated by evidence.

Even if patients do not present with a classical rheumatological pattern of disease, they need to have complete investigation, while we trust the description of all symptoms; as rather than presenting commonly with something uncommon, they are likely to be presenting uncommonly with something common.

**Disclosure:** The authors have declared no conflicts of interest.

#### 452. CLINICAL RELAPSE IN RITUXIMAB TREATED REFRACTORY WEGENER'S GRANULOMATOSIS DESPITE PERSISTENT SUPPRESSION OF B-CELL FUNCTION

Roope Manhas, Michael Webley and Sally Edmonds  
*Department of Rheumatology, Stoke Mandeville Hospital, Aylesbury, United Kingdom*

**Background:** The anti B-cell drug rituximab has been reported to be efficacious in the treatment of Wegener's granulomatosis (WG) refractory to cyclophosphamide

(CTX) therapy. We present a patient who initially responded well to rituximab, but subsequently relapsed clinically with persistent suppression of B-cell function which precluded further treatment.

**Methods:** A 39 year old woman presented in 1995 with rheumatoid factor positive polyarthritis, initially diagnosed as rheumatoid disease. She soon developed a vasculitic rash over the lower limbs and painful oral ulceration. ESR 89, CRP 20, c-ANCA positive at 1:160, ANA & ENA both negative. There was no evidence of renal or respiratory disease. A diagnosis of WG was made. Intravenous CTX was administered (1 g fortnightly x4, then monthly for a total of 6 pulses) resulting in clinical resolution & fall in ANCA titre (to 1:10). Azathioprine (AZA) & low dose prednisolone was used as maintenance therapy, both being discontinued by December 2002 due to disease remission. She remained well until 2004 when rash & joint symptoms recurred & ANCA titre rose to 1:320 requiring re-treatment with CTX. After the second pulse she developed a new headache & chondritis of the right pinna. Contrast enhanced MRI brain showed diffuse enhancement of the meninges. Ocular examination showed nodular scleritis & papilloedema on funduscopy (with raised intraocular pressures) despite CSF pressure of 3cm H2O on lumbar puncture. Further pulses of CTX failed to settle the headache, & MRI changes progressed. The patient was treated with rituximab in December 2005, and received 1 g (plus 750 mg CTX and 250 mg methylprednisolone) on each of two occasions, a fortnight apart.

**Results:** Treatment improved the headaches dramatically (although without complete resolution), intraocular pressures normalised & fundoscopic appearances improved. However in June 2006 (6 months post therapy), headaches and papilloedema worsened again. Visual field testing revealed constriction of peripheral vision in left eye. MRI scan of orbits excluded optic nerve or chiasm involvement. Ophthalmic surgeons performed left optic nerve sheath fenestration in an attempt to improve ocular drainage, but vision continued to deteriorate. B-cell function remained suppressed (total CD-19 B-cells 0.01x10<sup>9</sup>/l). The patient currently awaits assessment of intra-ventricular pressures, with a view to placement of a ventricular shunt. The meningeal enhancement on MRI persists.

**Conclusions:** Rituximab can be a very effective treatment for refractory WG, but when disease relapses clinically in the context of persistent B-cell suppression, further treatment can be difficult. Subsequent use of drugs that modulate T-cell function are likely to significantly increase risk of serious infection & impair immune surveillance against malignant disease.

**Disclosure:** The authors have declared no conflicts of interest.

**453. MULTIFOCAL TUBERCULOUS OSTEOMYELITIS - A CASE REPORT**

Caitriona Buckley<sup>1</sup>, Anita Rai<sup>2</sup> and Jaia Ravindran<sup>2</sup>  
<sup>1</sup>Rheumatology Department, Worcester Royal Hospital, Worcester, United Kingdom and <sup>2</sup>Rheumatology Department, University Hospitals of Coventry and Warwickshire, Coventry, United Kingdom

**Background:** Mycobacterial infection is an unusual cause of osteomyelitis in the developed world. We describe a case of multifocal tuberculous osteomyelitis in an otherwise healthy young woman.

**Methods:** A 28 year old Asian woman presented with a one year history of progressive left ankle pain and swelling. She also had left elbow pain. Apart from an uncomplicated, closed fracture of her left ankle 2 years previously her past history was unremarkable. She had been living in the UK for 2 years prior to this presentation. On examination there was an erythematous, fluctuant warm swelling behind the left lateral malleolus. The left calcaneum was tender, as was the left olecranon. Physical examination was otherwise normal.

**Results:** MRI of the left ankle showed a 2cm bone abscess in the left calcaneum which communicated with a 5cm subcutaneous abscess via a fistula. MRI of the left elbow showed a 3 x 2 x 2cm bone abscess in the olecranon process extending down to the shaft of the ulna. A reddish brown fluid was aspirated from the left ankle which subsequently grew acid fast bacilli. Histology from the abscess in the left calcaneum showed multiple epithelioid granulomata and necrotic debris resembling caseating necrosis. Investigations to find any other foci of infection were all negative.

Incision and drainage of the abscess cavities in the left lateral malleolus and proximal left ulna was carried out. She was commenced on a regimen of Rifampicin, Isoniazid, Pyrazinamide and Ethambutol for 2 months which was then changed to Rifampicin and Isoniazid for 4 further months. Pyridoxine was given for 6 months. She has recovered well on this treatment.

**Conclusions:** Although not common in the UK, there are approximately 30 million people with tuberculosis worldwide of whom between 1 and 3% have involvement of the skeletal system. However, the incidence of TB in the UK is rising, largely due to immigration from areas of high prevalence. Osteoarticular TB predominantly affects the spine, but it has been reported in all bones of the body. Ankle and elbow involvement are rare, each accounting for just 2% of cases of skeletal TB. Multifocal disease occurs in 3% of cases. Skeletal TB occurs mostly during the first 3 decades of life, and in the elderly. Radiological investigations can be unreliable, so diagnosis must be established histologically. Treatment is with a prolonged course of anti-tuberculous chemotherapy. Modern drug treatment has substantially reduced the need for surgery in this condition.

This case highlights that although rare in affluent societies, mycobacterial infection should be included in the differential diagnosis of chronic oligoarthritis particularly in patients who have migrated from areas of high TB prevalence. This is a curable condition and outcome is best the earlier it is diagnosed.

**Disclosure:** The authors have declared no conflicts of interest.

**454. CAN ANAKINRA CAUSE ANTI-GBM DISEASE ?**

Vipul Vagadia, Batsi Chikura and Roger Bucknall  
 Rheumatology, Royal Liverpool University Hospital, Liverpool, United Kingdom

**Background:** Adult Onset Still's Disease(AOSD) is a rare systemic inflammatory disorder of unknown origin. Recently, two case studies reported the successful induction of remission with the interleukin 1(IL 1) receptor antagonist anakinra in refractory AOSD. Anakinra found to have a steroid sparing agent in AOSD and usually well tolerated without major side effects. We report an unusual case of AOSD who developed anti-glomerular basement membrane (anti-GBM)-good pasture's disease on anakinra.

**Methods:** A 60 year old Caucasian male, had been diagnosed to have AOSD since 2000, on the basis of polyarthralgia, fever, rash and raised ferritin. He was treated with prednisolone 60 mg/day and high dose aspirin which failed to control his symptoms. Despite successive treatment with high dose prednisolone and methotrexate, ciclosporin, the patients' AOSD remained active. In January 2006, he was started on etanercept with prednisolone 40 mg/day, which he tolerated well but this combination could not keep disease in remission. Due to severity of his recurrent symptoms of his AOSD, he was started on anakinra 100mg/day in June 2006, which kept disease in remission with CRP from 130 to 5 in few days and he could come off prednisolone.

In March 07, he presented with acute renal failure without any precipitating features secondary to anti-GBM disease.(See table).

**Results:** Anakinra was stopped in the assumption of anakinra induced anti-GBM disease. For the treatment of his anti-GBM disease, he had I/V methylprednisolone, plasma exchanges, cyclophosphamide. He had regular haemodialysis for his severe renal impairment. After 3 months of cyclophosphamide treatment he developed bone marrow suppression so, we withdrew it. He was started back on prednisolone 50 mg/day as he had recurrence of AOSD symptoms. He became persistently anti-GBM antibody negative since June 07. Currently, he is on prednisolone 30 mg/day and azathioprine which is partially controlling his symptoms of AOSD.

**Conclusions:** In animal studies, IL-1 blocker has been shown to be effective in ameliorating experimental anti-GBM associated glomerulonephritis. In contrast, our patient developed anti-GBM disease while been on anakinra and remain anti-GBM persistently negative since off anakinra. There are no reported cases of anti-GBM disease caused by anakinra. We could not prove any coincidence of anti-GBM disease in our patient but as he became persistently anti-GBM negative so, we assume that there could be direct causation between anakinra and anti-GBM disease.

**Disclosure:** The authors have declared no conflicts of interest.

|                         | March 2007   | July 2007                            |
|-------------------------|--|--------------------------------------|
| Urea(Jan 07 - 8.0)      | 54   | 8.8                                  |
| Creatinine(Jan 07 130)  | 1717   | 399                                  |
| Ultrasound of kidney    | no evidence of obstructive renal disease. kidney of normal size.     |                                      |
| ANCA, ANA               | Negative   | Negative                             |
| Anti- GBM (Normal <2.5) | 97   | Persistently Negative since June 07) |
| Renal biopsy            | resnetic glomerulonephritis consistent with anti-GBM disease         |                                      |
| CT scan of Chest        | No evidence of pulmonary haemorrhage or lung parenchymal involvement | No further changes in lungs          |

**455. A VERY WOOLLY DIAGNOSIS: COXIELLA BURNETI REACTIVE ARTHRITIS**

Helen Cohen<sup>1</sup>, Marina Morgan<sup>2</sup> and David Blake<sup>1</sup>  
<sup>1</sup>Rheumatology, Royal National Hospital for Rheumatic Diseases, Bath, United Kingdom and <sup>2</sup>Microbiology, Royal Devon and Exeter NHS Foundation Trust, Exeter, United Kingdom

**Background:** We describe the case of a professional musician presenting with a reactive arthritis due to Coxiella burneti. There is only one previous case report of reactive arthritis due to this organism. The case highlights the need for a thorough history in cases of reactive arthritis, and that zoonotic (organisms transmissible from man to animal and vice-versa) infections can present as acute rheumatological conditions.

**Methods:** A professional violinist presented to her GP with acute onset of malaise, joint pain, swelling and stiffness. Her doctor noted tender, hot swollen joints at the MCPs, PIPs, elbows and knees together with knee effusions. Diclofenac 50 mg tds was prescribed, and her joint swelling settled. Two weeks later, she complained of a further relapse lasting two days and of some stiffness in her fingers. Blood testing revealed a CRP of 13 mg/dl, Anti DS-DNA 138iu/ml, C-ANCA 1:80, PR3 +ve. She was concerned that further attacks might reduce dexterity in her hands, and referred for a rheumatology opinion.

On examination she was well with no signs of synovitis or joint inflammation. Closer questioning revealed that she had taken in three orphan lambs one week prior to the onset of her illness. She did not have any animal husbandry experience and was unable to cope with them running around her town house soiling the floors. One lamb became unwell. The clinical impression was of a zoonotic infection

causing a reactive arthritis. The case was discussed with a microbiologist with an interest in zoonoses, and an infection screen performed.

**Results:** Repeat antibody profiles were all negative. Serology was consistent with past infection with *Coxiella burnetii*.

**MANAGEMENT:**

In view of the potential risk of chronic Q fever endocarditis, a course of Doxycycline 200mg od for 3 weeks was given. She has remained well and continues to play the violin.

**DISCUSSION:**

There is one previous case report of *Coxiella burnetii* reactive arthritis, described as a 'migratory oligoarthritis' shortly after acute Q fever. The important reservoirs are cattle, goats and sheep. Transmission can occur through contact with contaminated dust, infected carcasses, contaminated milk and parturient material. Veterinary, farm and abattoir workers are high risk groups. The common practice in this country of reviving new born, hypothermic lambs in the bottom warming oven of an Aga is an effective way of generating rickettsial spores in household dust.

**Conclusions:** KEY MESSAGES: 1.Always take a thorough history in possible cases of reactive arthritis. 2.Acute infections may cause false positives to auto-antibody blood tests. 3.Zoonotic infections need to be considered in the urban population as well as the rural population. 4.Chronic Q fever often manifests as endocarditis with consequent high morbidity and mortality.

**Disclosure:** The authors have declared no conflicts of interest.

#### 456. THE SPECTRUM OF ARRHYTHMIAS IN CARDIAC SARCOID

Shweta Bhagat<sup>1</sup>, Sumeet Agrawal<sup>1</sup>, Damodar Makkuni<sup>1</sup>, Leena Das<sup>1</sup>, Frances Borg<sup>1</sup>, Halan Moorthy<sup>2</sup> and Bhaskar Dasgupta<sup>1</sup>  
<sup>1</sup>Rheumatology, Southend Hospital, Southend, United Kingdom and  
<sup>2</sup>Cardiology, Southend Hospital, Southend, United Kingdom

**Background:** Conduction abnormalities are potentially life threatening manifestations of cardiac involvement with sarcoidosis. We discuss the importance of being aware of cardiac involvement as a serious complication of sarcoidosis, imaging modalities and treatment aspects.

**Methods:** We present two interesting cases of multisystem sarcoid which represent two ends of the spectrum of these conduction abnormalities.

**Results:** The first case is of a 61 year old male who presented to rheumatology with multiple subcutaneous lumps and dactylitis. He subsequently presented to casualty with episodes of dizziness. Examination revealed a heart rate of 35/min. Electrocardiogram showed complete heart block. Further investigations revealed elevated serum calcium, serum creatinine and serum ACE, with elevated Troponin. Echocardiogram showed severe concentric hypertrophy and high resolution CT scanning revealed bilateral hilar lymphadenopathy and bilateral pleural effusions. Histology from the cutaneous lumps confirmed the diagnosis of saroidosis. He had a permanent pacemaker implanted. PET/CT scan showed uptake in the hilar lymph nodes and MIBI scan of the heart was abnormal. He was treated with oral prednisolone and methotrexate. The skin nodules and dactylitis disappeared completely and the pleural effusions resolved. However the cardiac rhythm has not normalised and the ventricular hypertrophy has not regressed.

The second case is that of a 44 year old male, known to have lupus pernio and pulmonary sarcoid, being treated effectively with corticosteroids. He presented with ventricular tachycardia (VT) which was refractory to initial treatment with amiodarone. Cardiac MRI showed myocardial scarring consistent with cardiac sarcoid. He had an implantable cardioverter/defibrillator inserted, and was also treated with long term prednisolone.

**Conclusions:** Cardiac sarcoidosis, despite being not so common clinically, accounts for close to one-fourth of all sarcoid-related deaths. Heart block develops mainly during the active phase of the disease. Early treatment with corticosteroids might improve AV conduction disturbance. However, sustained VT is not closely linked with disease activity and frequently develops in the advanced stage of disease. Our second patient developed VT while on corticosteroids, which are the mainstay of treatment and may halt worsening of left ventricular dysfunction. However there is a lack of long term data on the use of pacemakers/defibrillators and hence they are best left insitu even if the disease is in remission.

**Disclosure:** The authors have declared no conflicts of interest.

#### 457. GIANT CELL ARTERITIS - A PREVENTABLE STROKE OF THE EYE; THE IMPORTANCE OF RECOGNISING JAW CLAUDICATION AS A PREDICTOR OF NEURO-OPHTHALMIC INVOLVEMENT

Frances A. Borg, Shweta Bhagat, Leena Das and Bhaskar Dasgupta  
 Rheumatology Department, Southend Hospital, Westcliff-on-Sea, United Kingdom

**Background:** Guidelines for the early recognition and management of giant cell arteritis (GCA) have recently been produced<sup>1</sup>. Features such as visual disturbance and jaw claudication are prognostic of impending visual loss, and therefore should be considered as "TIAs of the eye"

We describe a case illustrating the necessity of early recognition of jaw claudication as a feature of GCA, and a missed opportunity to save sight.

**Methods:** An 81 year old woman presented with a three day history of complete visual loss in the right eye and two weeks of intermittent blurred vision in the left eye. She had consulted her General Practitioner four weeks previously with jaw claudication. There was no history of headache or scalp tenderness, and there were no features of polymyalgia rheumatica. She was advised to have blood tests but was not referred for exclusion of GCA, or started on steroids.

On examination, she was unable to perceive light in the right eye and had vision of 6/36 in the left eye. The right optic disc was swollen and pale, with attenuated retinal arteries. There was pallor and blurring of the nasal margin of the left optic disc. Both temporal arteries were thickened. The right was non-pulsatile, the left was still pulsatile. Full systemic examination was normal, including peripheral pulses, and there was no evidence of muscle tenderness. Initial blood tests revealed ESR 27, CRP 13 and were otherwise normal.

**Results:** A diagnosis of anterior ischaemic optic neuropathy was made. Temporal artery biopsy not only confirmed GCA but also showed severe mediointimal proliferation which is a marker of neuro-ophthalmic complications.

She was treated with methylprednisolone 500mg bd for three days. The vision in the left eye improved to 6/12 and CRP fell to 2. Unfortunately, on switching to oral prednisolone, the left sided vision deteriorated, with swelling and pallor to the entire left optic disc. Her vision is currently 6/60 in the left eye, and she remains unable to perceive light in the right eye. Having lived independently, she will now need residential care.

**Conclusions:** This case illustrates the importance of recognising jaw claudication as a predictor of visual loss in GCA. This patient had none of the other well recognised features of GCA, and only a very modest rise in her inflammatory markers. We suspect that if she had described headache, the diagnosis would have been recognised when she initially presented, and steroid therapy would have been instituted in time to save her vision. GCA should be thought of as a "preventable stroke of the eye", with jaw claudication considered to be a "TIA" of this.

**Disclosure:** The authors have declared no conflicts of interest.

#### Reference

1. Dasgupta B, Hassan N. Giant cell arteritis: recent advances and guidelines for management. Clin Exp Rheumatol 2007;25(1)Suppl 44:S62-5.

#### 458. DERMATOMYOSITIS ASSOCIATED WITH ASTROCYTOMA AND DENDRITIC CELL IMMUNOTHERAPY

Emma C. Derrett-Smith and David A. Isenberg  
 Centre for Rheumatology, University College London Hospital, LONDON, United Kingdom

**Background:** A 39 year old man presented with 4 weeks of muscle pain and weakness, fatigue, fever and a rash. A year earlier, he had developed a grade II astrocytoma. This had been debulked with 85% clearance leaving no neurological deficit. Follow-up MRI scans of the tumour over several months showed no changes.

**Methods:** There had been no indication for further conventional therapy but he contacted a centre in Germany specialising in dendritic cell (DC) immunotherapy. The treatment he received combined the standard use of DC with Newcastle Disease virus, one of the class of oncolytic viruses thought to promote tumour death. The combination was infused into the patient with additional interferon.

**Results:** The patient presented two weeks later with a rash classical of dermatomyositis. He had MRC grade 4 proximal muscle weakness. The only other clinical finding was of non-specific mucositis and oral candidiasis.

He had a low-grade inflammatory response with CRP 11, ESR 30, a creatine kinase (CK) 4280 iu/L and normal immunology. MRI brain revealed no change in tumour compared with the previous study. EMG revealed a proximal myopathy, MRI of the thigh revealed an inflammatory process most prominent in the adductors and a muscle biopsy confirmed the diagnosis. A PET scan revealed abnormal uptake in the proximal muscles only.

Treatment with intravenous and then oral steroid and a single dose of 7.5mg methotrexate did not result in a change in clinical or biochemical parameters.

The patient did, however, develop cellulitis at the site of his muscle biopsy and an elbow synovitis. He was treated with standard antibiotics and cultures later revealed a sensitive staphylococcus aureus from the elbow aspirate and negative cultures elsewhere.

Over two days, the cellulitis had spread and the site was opened surgically to reveal extensive and deep myonecrosis and necrotising fasciitis. The patient did not mount a clinical response to this and developed only a late modest rise in CRP to 110. He was treated with intravenous immunoglobulin (IVIg) and transferred for specialist plastic surgical care. Six further resections of necrotic tissue were required prior to a major skin graft and, following further IVIg, the patient recovered full muscle strength. CK returned to normal and the rash resolved completely.

**Conclusions:** We have presented this case to highlight a number of interesting points. If his condition were paraneoplastic, it is the first to be reported associated with a primary brain tumour. In the context of negative auto-antibodies, an unrelated immune pathology is unlikely. There are no reported cases of myositis related to DC therapy, but it can induce autoimmunity. Anecdotally, we felt that the patient did not mount the expected immune response to his infection, and that his immunotherapy may have contributed to this through attenuation of his immune response.

**Disclosure:** The authors have declared no conflicts of interest.

#### 459. BILATERAL PAROTID GLAND INVOLVEMENT IN WEGENER'S GRANULOMATOSIS

Amrit K. Saha<sup>1</sup>, Satish Rachapalli<sup>2</sup> and Patrick Gordon<sup>3</sup>  
<sup>1</sup>Rheumatology, Guy's and St Thomas' NHS Trust, London, United Kingdom,  
<sup>2</sup>Rheumatology, Ashford and St. Peter's Hospitals NHS Trust, Ashford, United Kingdom and <sup>3</sup>Rheumatology, King's College Hospital NHS Trust, London, United Kingdom

**Background:** Wegener's granulomatosis (WG) is a granulomatosis small vessel vasculitis. Classically, it is a disease having a predilection for the upper airways, lungs and kidneys associated with classical antineutrophil cytoplasmic antibodies (cANCA). Salivary gland involvement is a rare feature of this disease with bilateral parotid gland involvement rarer still.

**Methods:** We report a case of limited WG with bilateral parotid gland involvement. **Results:** An 18-year-old caucasian woman with asthma presented with sinusitis, bilateral parotid gland swelling, right sided facial nerve palsy, arthralgia, weight lost and elevated inflammatory markers. Magnetic resonance imaging showed enlarged parotid glands with histology from both parotids showing poorly formed granulomas. A presumptive diagnosis of sarcoidosis was made and she was started on 60 mg of prednisolone. In spite of this treatment, leakage of fluid from the biopsy sites continued and she later developed nasal bridge collapse with epistaxis. Further investigations showed normal renal function, urinalysis and Computed Tomography (CT) of the chest. Antinuclear antibodies and extractable nuclear antigens were not detected. Antiproteinase 3 antibodies were minimally elevated-9 (normal <5U/ml). A CT of her neck showed enlarged parotid glands and right lacrimal gland. Other salivary glands were normal. CT of the sinuses showed extensive defects within the nasal septum. A biopsy of the paranasal sinus and right nasal septum showed multinucleate giant cells with evidence of a vasculitis consistent with Wegener's granulomatosis. She was treated with co-trimoxazole, prednisolone and cyclophosphamide. With this treatment, antiproteinase 3 levels and inflammatory markers have normalised. Clinically, her parotid gland swellings are reducing and her facial weakness is improving.

**Conclusions:** In the whole literature, there have only been 5 definite cases of WG with bilateral parotid gland involvement. They have all been the limited form of WG and all have been successfully treated with cyclophosphamide and prednisolone. This case is unique in the fact that bilateral parotid gland swelling was the presenting complaint without any evidence of submandibular gland or lung involvement. The differential diagnosis of bilateral parotid gland swelling should include Wegener's granulomatosis and appropriate investigations should be carried out.

**Disclosure:** The authors have declared no conflicts of interest.

#### 460. REVERSIBLE HODGKIN'S LYMPHOMA ASSOCIATED WITH EPSTEIN-BARR VIRUS IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS ON LONGTERM AZATHIOPRINE THERAPY

Sarah J. Evans and Margaret M. O'Sullivan  
 Rheumatology, North East Wales NHS Trust, Wrexham, United Kingdom

**Background:** Patients with systemic lupus erythematosus (SLE) have an increased risk of developing hematologic malignancies. The risk of non-Hodgkin's lymphoma (NHL) is well documented but an increase in Hodgkin's lymphoma has more recently been reported. The link to explain this excess risk continues to be studied, but immunosuppressive drugs used in the treatment of SLE may be involved in their aetiology.

Patients treated with immunosuppressive agents following organ transplantation are at an increased risk of developing lymphoproliferative disorders. These lymphomas are strongly associated with Epstein-Barr Virus (EBV), and are closely linked to the competence of the immune system. We describe a case implicating azathioprine therapy in the development of an EBV-driven HL in a patient with SLE. **Methods:** A 43-yr-old female with SLE had been commenced on prednisolone, azathioprine and cyclosporin in 1998 for lupus pneumonitis. By 2005 the level of immunosuppression had been reduced and only azathioprine was continued. She remained well until presenting in April 2007 with a six week history of severe soreness of the tongue and throat, and fullness at the back of her mouth causing difficulty swallowing. This was associated with weight loss of 5 kg since January 2007 (less than 10% of her total body weight). She had never smoked.

**Results:** Physical examination revealed a 1.5cm firm indurated mass at the right postero-lateral border of the tongue. Blood results at the time showed a mild anaemia and lymphopenia in keeping with her SLE. C-Reactive Protein and plasma viscosity were normal.

MRI scan confirmed a mass at the base of the tongue extending into the pharyngeal tissue and oropharynx associated with local lymphadenopathy. Staging CT scan of the chest, abdomen and pelvis was unremarkable.

Histologic examination of the biopsy showed a mixed lymphoid infiltrate consisting of both T and B-cell blasts with strong positivity for the EBV marker EBER (EBV-encoded RNA). Subsequent clonality studies confirmed a HL of the nodular sclerosing type. Azathioprine was stopped and after six weeks the lesion had started to regress (documented by repeat MRI scan) and continues to resolve without chemotherapy.

**Conclusions:** This case implicates azathioprine in the development of an EBV-associated HL in a patient with SLE. Tumour regression after the withdrawal of azathioprine suggests that the therapy was a major factor in the generation of this lymphoma.

**Disclosure:** The authors have declared no conflicts of interest.

#### 461. ERECTILE DYSFUNCTION IN RHEUMATOID ARTHRITIS PATIENT ON ETANERCEPT

Abhishek Abhishek and Louisa Badcock  
 Department of Rheumatology, Derbyshire Royal Infirmary, Derby, United Kingdom

**Background:** Patients with rheumatoid arthritis (RA) are sexually less active than healthy population. This is multifactorial and correlates with disease activity (1). TNF- $\alpha$  is one of the mediators of erectile dysfunction (ED). Treatment with anti TNF- $\alpha$  in RA would therefore be expected to alleviate pre-existing ED. We report the first case of isolated reversible ED in a RA patient on anti TNF- $\alpha$  agent etanercept.

**Methods:** A 60yr old man with 23yr history of erosive RA was commenced on etanercept (50mg/week, subcutaneously) in October 2006. Before commencing etanercept he had failed hydroxychloroquine, D-penicillamine, gold, sulfasalazine, methotrexate and leflunomide. Prior to commencing etanercept, the DAS-28 score was 6.32 (SJC 10, TJC 20, VAS 65, ESR 18 mm/hr). Within 2 month of commencing on etanercept, he developed testicular pain and erectile dysfunction manifest as failure of erection. Around this time RA was well controlled with a DAS-28 score of 2.75 (SJC 2, TJC 1, VAS 18, ESR 9 mm/hr) and examination of other systems was normal. He stopped etanercept himself on which ED improved. Since ED is not a reported side effect of etanercept, it was started again. On restarting etanercept ED recurred. Investigations revealed a normal full blood count, urea electrolytes & creatinine, liver function tests, prostate specific antigen, serum protein electrophoresis, MRI of lumbar spine and testicular ultrasound. At urology and neurology consultation no other cause for ED was demonstrated and it was felt to be due to etanercept.

Etanercept was stopped again. Within a week of stopping etanercept, testicular pain and ED began to improve. He obtained normal sexual function within a month. After 6 month of discontinuing etanercept, the ED and testicular pain have not recurred. Even though he is not on any DMARD, RA remains in remission with DAS score of 2.35 (SJC 6, TJC 0, VAS 8, ESR 9 mm/hr).

**Results:** The occurrence of ED in our patient on anti TNF- $\alpha$  agent is counter-intuitive. TNF- $\alpha$  is believed to mediate ED. ED is associated with increased blood levels of inflammatory.

(IL-6, IL-1 $\beta$ , & TNF- $\alpha$ )mediators. These correlate negatively with sexual performance (2). Moreover TNF- $\alpha$  up-regulates phosphodiesterase type 5 (PDE-5) expression in cavernosal vascular smooth muscle cells. As PDE-5 hydrolyses cGMP, this effect might 'blunt' the pro-erectile actions of NO (3). Impotence has been reported in patients with RA on methotrexate but our patient was not on methotrexate at this time (4).

**Conclusions:** Our patient had an idiosyncratic reaction to an anti TNF- $\alpha$  agent. We would like to increase awareness among our colleagues about occurrence of this unusual side effect.

**Disclosure:** The authors have declared no conflicts of interest.

#### References

1. Clinical Rheumatology 2007; 26(1):30-8.
2. European Heart Journal 2006; 27(22): 2640-8.
3. British Journal of Urology International 2007; 99(3): 612-8.
4. Arthritis and Rheumatism 1989; 32(10):1341-2.

#### 462. AORTIC ANGIOSARCOMA MIMICKING TAKAYASU'S ARTERITIS: A DIAGNOSTIC DILEMMA

Maysam Abdin-Mohamed<sup>1</sup>, Joanna Ledingham<sup>1</sup>, Fiona Witham<sup>2</sup> and Fiona McCrae<sup>1</sup>

<sup>1</sup>Rheumatology, Queen Alexandra Hospital, Portsmouth, United Kingdom and

<sup>2</sup>Radiology, Queen Alexandra Hospital, Portsmouth, United Kingdom

**Background:** Takayasu Arteritis (TA) is a large vessel vasculitis that can cause stenosis and potential occlusion of the aorta. This form of vasculitis may present with ischemic symptoms or with less specific symptoms. We present a case ultimately diagnosed with Primary Aortic Angiosarcoma (PAA) that initially presented with symptoms consistent with TA.

**Methods:** A 55-year-old lady was referred to the Rheumatology department in April 2005 with symptoms of myalgia, arthralgia and painful thighs both at rest and on activity. Examination showed mildly reduced global power and reduced distal leg pulses. Investigations revealed elevated inflammatory markers (ESR 101 mm/hr, CRP 90 mg/l) but further detailed investigations, including tumour markers, immunology and vasculitis screens were normal. Isotope bone scan revealed increased uptake in the left tibia; subsequent x-ray showed features of a bone infarct only.

Abdominal CT however showed narrowing of the distal aorta and angiogram confirmed thickening with almost complete occlusion of the aorta just above its bifurcation. These findings supported a working diagnosis of type 4 TA.

**Results:** Sequential treatment with high doses of oral prednisolone (up to 80 mg) and 2 weekly infusions of cyclophosphamide (500 mg) and methylprednisolone (1 g) resulted in only minor improvement in symptoms and persistent raised blood inflammatory markers (ESR 100 mm/hr, CRP 142 mg/l). Claudicant symptoms continued despite insertion of an aortic stent. Anti-TNF treatment (infliximab) was commenced for a working diagnosis of treatment resistant TA with again a poor response. This lady developed new symptoms of left tibial pain and a raised serpiginous rash. Left knee x-ray showed a new lesion at the upper left tibia.

MRI and CT of the tibia revealed a destructive osteolytic lesion. Skin and bone biopsies were undertaken and revealed a high-grade multi-focal epithelioid angiosarcoma.

Immunosuppressant therapy was replaced by chemotherapy and palliative radiotherapy to her leg. Unfortunately this lady did not respond to treatment and her condition deteriorated until her death in November 2006. A post-mortem confirmed the diagnosis of PAA.

**Conclusions:** PAA is extremely rare (only 30 cases reported in the literature) and these tumours originate in the descending thoracic or abdominal aorta as in the case presented. Prognosis is poor due to their late diagnosis and early metastatic spread.

TA is a rare form of large vessel vasculitis and up to 50% of patients have been reported to have active disease despite steroids.

This case highlights the possible diverse presentations of malignancies and the high index of suspicion that must always be applied to patients with a presumed connective tissue disease or vasculitis that is unresponsive, or poorly responsive, to conventional treatments.

**Disclosure:** The authors have declared no conflicts of interest.

#### 463. MYCOBACTERIUM XENOPI WRIST ARTHRITIS IN A YOUNG IMMUNOCOMPETENT WOMAN

Mohamed M. Bakr<sup>1</sup> and Sally Edmonds<sup>2</sup>

<sup>1</sup>Rheumatology, Stoke Mandeville Hospital, Aylesbury, United Kingdom and

<sup>2</sup>Rheumatology, Stoke Mandeville Hospital, Aylesbury, United Kingdom

**Background:** Mycobacterium xenopi is a slow-growing, nontuberculous mycobacterium responsible for nosocomial and opportunistic infections, mainly the lung. Few cases of mycobacterium xenopi arthritis have been reported and only one case of wrist involvement (tenosynovitis in immunocompetent patient). To our knowledge, this is the first reported case of wrist joint infection.

**Methods:** A 39-yr-old female, presented with a two week history of painful and swollen right wrist. There was no history of preceding infection or trauma. In 1999 and 2002, She underwent right tenosynovectomy for de Quervain's tenosynovitis and in 2005, right ulnar decompression at the elbow.

On examination, she was afebrile and systemically well. The right wrist was swollen and exquisitely tender with limited movement.

**Results:** Laboratory investigations showed raised CRP at 61.9, WBC 12.5 and neutrophils 8.1. X-ray of the right wrist and a CXR were normal.

Ultrasound of the right wrist showed fluid around the distal radio-ulnar joint and tenosynovitis of the extensors. MRI of the right wrist demonstrated moderate inflammatory change throughout the soft tissues of the wrist with radio-ulnar and radiocarpal effusions. Microscopy of the joint aspirate showed few pus cells with no organisms or crystals (she was not on any antibiotics). Arthroscopy of the right wrist was performed approximately 40 days from the initial presentation due to persistence of pain and swelling. This showed florid synovitis throughout the wrist joint. Synovial biopsy demonstrated non-caseating granulomatous synovitis and synovial culture grew mycobacterium xenopi. Triple therapy with rifampicin, clarithromycin and ethambutol was started with gradual significant improvement of the right wrist.

**Conclusions:** Mycobacterium xenopi is a rare cause of septic arthritis especially in an immunocompetent individual. Our case was thought to be a result of previous instrumentation around her wrist. Early synovial biopsy should be encouraged for diagnosis before irreversible joint damage occurs especially in those with previous surgical procedures in or around the involved joint.

**Disclosure:** The authors have declared no conflicts of interest.

#### 464. A FEMORAL ANEURYSM IN BEHCET'S DISEASE: WHEN TO OPERATE?

Margaret H. Ma<sup>1</sup>, Pradip Nandi<sup>1</sup>, Edward Chaloner<sup>2</sup>, Ghada Yanni<sup>1</sup> and Vijay Hajela<sup>1</sup>

<sup>1</sup>Department of Rheumatology, University Hospital Lewisham, London,

United Kingdom and <sup>2</sup>Department of Vascular Surgery, University Hospital Lewisham, London, United Kingdom

**Background:** Behçet's Disease (BD) is a chronic, relapsing, inflammatory disease characterized by recurrent oral and genital ulcers. It may also be associated with inflammation affecting the joints, eyes, brain, skin and blood vessels. Men are affected more often and with more severe disease than women. Vascular involvement occurs in 7-29% of patients. There are 4 types of vascular lesions: arterial occlusions, venous occlusions, aneurysms and varices. Aneurysm formation accounts for most vascular deaths. Common sites include abdominal aorta, femoral artery, and thoracic artery.

**Methods:** A 36 year-old Turkish gentleman presented to the Emergency Department with a 1-month history of a swelling in the left groin. He had been diagnosed with BD 3 years previously. There was a history of recurrent oral and genital ulceration and erythema nodosum. There had been no other organ involvement and he had only been treated with colchicine.

**Results:** On examination, he was febrile. There was a pulsatile mass in the L groin. Peripheral pulses in his lower limb were present. He was also noted to have oral and scrotal ulcerations. There were no eye, skin or joint involvement. Haematological investigations showed Hb 13.4g/dL, WCC 13.3 × 10<sup>9</sup>/L, CRP 227mg/L and ESR 65mm/hr. Urine and blood culture were sterile. Upon

presentation, he underwent a CT angiogram which showed a left 5 cm common femoral aneurysm with mural thrombus.

He was commenced on high dose prednisolone (60mg) and aspirin (75mg). Antibiotics were also used to cover for cellulitis around the scrotal ulcers. Subsequently, he developed paraesthesiae in L3 dermatome and weak hip flexion (power 3/5). It became evident that the aneurysm was compressing the femoral nerve and urgent surgery was required. His fever subsided and CRP was improving with immunosuppression. 14 days after presentation, he underwent a repair of the aneurysm. 6 weeks post-operatively, he was treated with a course of intravenous cyclophosphamide and methylprednisolone. For maintenance therapy, azathioprine was used. He has remained well on follow-up. Repeat imaging has shown no further aneurysms.

**Conclusions:** Surgical treatment of BD aneurysms can be challenging. The aneurysm itself can be technically difficult to operate on and there is a higher rate of graft occlusion. It is important to avoid surgical intervention during the active stage of disease if this is possible to avoid breakdown of the anastomosis. However complications such as aneurysm rupture or compressive symptoms on surrounding tissue may necessitate urgent surgery as in this case. Close liaison between medical and surgical teams was important to help judge the timing and preparation for surgery. There is risk of new aneurysms developing post-operatively and so regular imaging was required post-operatively.

**Disclosure:** The authors have declared no conflicts of interest.

#### 465. MYCOPHENOLATE MOFETIL IN PAEDIATRIC DERMATOMYOSITIS AND ANTI-JO1 POSITIVE POLYMYOSITIS

David J. Armstrong<sup>1</sup>, Graham M. Raftery<sup>1</sup>, Ismael Atchia<sup>1</sup> and Bridget Griffiths<sup>2</sup>

<sup>1</sup>Rheumatology, University Hospital of North Durham, Durham, United Kingdom and

<sup>2</sup>Musculoskeletal Unit, Freeman Hospital, Newcastle-upon-Tyne, United Kingdom

**Background:** Mycophenolate mofetil (MMF) is a lymphocyte inhibitor used across a range of autoimmune conditions. Experience of its use in myositis is limited.

**Methods:** We describe two cases of longstanding myositis in which conventional therapy had been unsuccessful, but in which the use of MMF induced clinical remission.

**Results:** Patient one, a 40 year old female, presented 10 years ago with generalised myalgia affecting her arms, legs and jaws. Creatinine Kinase (CK) level was 922 U/l (<170), aldolase 41 U/l (<7.6); EMG was in keeping with myositis. Anti-Jo1 antibody was positive, without evidence of pulmonary involvement. There was a good response to Prednisolone 40 mg daily, but she developed a flare the next year when MMF was reduced to 10mg daily. She was treated over the next 10 years with Prednisolone at doses between 7 mg and 20 mg, and the introduction of azathioprine (up to 100 mg daily) and methotrexate (up to 12.5 mg per week) were ineffective. She was reluctant to receive cyclophosphamide. For most of this period the CK level fluctuated between 500 U/l and 1500 U/l, and at times she was significantly incapacitated. She had to give up work.

MMF was introduced (1g bd), and within 3 months the CK level had reduced to 109 U/l. Nine months later she remains in complete clinical and biochemical remission, taking Prednisolone 4 mg daily, and has returned to work.

Patient two, a 27 year old female, presented 16 years ago with an erythematous rash, proximal muscle weakness and dyspnoea; CK was 22,640 U/L and ANA 1:1280 (ENA negative). A diagnosis of dermatomyositis was made, initially controlled with ciclosporin and intermittent pulses of methylprednisolone. In 2002 she had worsening weakness (CK6700 U/L); prednisolone 60 mg daily and methotrexate 25 mg/week were commenced. CK remained elevated (8000 U/L) despite the addition of monthly intravenous immunoglobulin (IVIg) infusions. In May 2004 she developed further weakness, dysphagia, and dyspnoea. She received 6 pulses of cyclophosphamide and methylprednisolone, then commenced azathioprine (increased to 150 mg daily) and prednisolone 30 mg daily. Initial improvement in CK was not sustained; IVIg was restarted in November 2004. In Sept 2005 she remained fatigued (CK 2000 U/L) and was receiving treatment with prednisolone 20 mg daily, azathioprine 100mg daily and IV Ig 1 g/kg for 2 days every 4 weeks. MMF 500 mg daily was added, improving activities of daily living, reducing fatigue and facilitating a gradual reduction in prednisolone to 5 mg daily. There have been no further flares in disease activity and most recent CK was 630.

**Conclusions:** There is little information on the successful use of mycophenolate mofetil in Jo-1 positive polymyositis, or paediatric dermatomyositis. We believe it has a role to play in difficult or resistant cases.

**Disclosure:** The authors have declared no conflicts of interest.

#### 466. TRAUMA AS A CAUSE OF INFLAMMATORY ARTHRITIS - A SHORT CASE SERIES INCLUDING MRI FINDINGS

Angharad Roberts<sup>1</sup>, Shah Khan<sup>2</sup> and John E. Brockbank<sup>1</sup>

<sup>1</sup>Rheumatology, East Lancashire NHS Trust, Blackburn, United Kingdom and

<sup>2</sup>Radiology, East Lancashire NHS Trust, Blackburn, United Kingdom

**Background:** Although infrequent, there are several reports of trauma induced spondylo-arthropathy. Here we describe 3 further cases of post-traumatic inflammatory arthritis in young men, one of whom rapidly developed erosions, periosteal reaction and significant cartilage loss as seen on MRI. All cases were

asymptomatic prior to trauma, had a systemic acute phase response, and developed arthritis within 2 weeks of injury.

**Methods:** Case 1

A previously well 33 yr old man with psoriasis suffered a fracture of the distal phalanx of his thumb. 10 days after injury he presented with a diffusely swollen thumb and elevated C-Reactive Protein (CRP) (60 mg/l). In short succession he developed synovitis of both knees, right ankle, left 2nd PIP and an achilles enthesitis. Synovial fluid revealed no crystals or bacterial growth. He had a negative screen for alternative causes of reactive arthritis. The patient improved with joint injections and diclofenac. The thumb remained swollen and radiographs at follow-up showed marked periosteal reaction with expansion of the sesamoid bone and erosions at the 1st IP and MCP joint. MRI at 6 months confirmed synovitis, erosions, marked cartilage loss at both joints, tenosynovitis and oedema of the sesamoid.

**Case 2**

A young boy presented at age 11 with a swollen left knee after ankle trauma. MRI and arthroscopy at that time revealed only an idiopathic synovitis, which eventually settled after steroid injections. He re-presented at age 14 to orthopaedics after trauma to the right knee, which again became swollen (10 ml aspirated). On this occasion he also developed oral ulceration and an effusion (115 ml) of the opposite knee. CRP was elevated at 41 mg/l both knees were injected and his symptoms and inflammatory response settled.

**Case 3**

A 16 yr old boy suffered minor trauma to his left knee after a bike accident. The left knee remained swollen, but he developed swelling in his right knee at two weeks with significant early morning stiffness and inflammatory back pain. ESR was elevated at 22 mm/hr, radiographs of the spine and pelvis were normal. Aspirate from both knees showed marked leucocytosis. Again there was no other preceding infective illness or other risk factor. He was treated with diclofenac and steroid injection of both knees.

**Results:** Previous case reports and series suggests joint trauma may precipitate not only local inflammatory joint disease but also more widespread disease and the time-line in these cases would support this. It is postulated that self antigens exposed during injury may initiate an autoimmune reaction in genetically predisposed individuals. Deep Koebner phenomena and Substance P have also been implicated.

**Conclusions:** These cases support enquiry for trauma as a precipitating factor for arthritis in patients susceptible to spondylo-arthropathies.

**Disclosure:** The authors have declared no conflicts of interest.

#### 467. NOT 'AS' IT SEEMS

Vinodh Devakumar<sup>1</sup>, Khan Shah<sup>2</sup> and John Brockbank<sup>1</sup>  
<sup>1</sup>Rheumatology, East Lancashire NHS Trust, Blackburn, United Kingdom and <sup>2</sup>Radiology, East Lancashire NHS Trust, Blackburn, United Kingdom

**Background:** Typical presentations of Osteomalacia (OM) include fractures, Looser's zones or rickets. Hypophosphataemic OM can rarely present with a pattern similar to Ankylosing Spondylitis (AS) with proliferative vertebral syndesmophytes and ossification around the hips but with sparing of the sacro-iliac joints (SIJs). We present just such an unusual case who was initially managed as AS. We also present the first report of the Magnetic Resonance Imaging (MRI) findings in this condition.

**Methods:** A 23-year-old woman of Asian descent presented with lower back pain and stiffness after childbirth. Examination revealed restricted hip and spinal movements with a kyphosis. Modified schober's test was 3 cm. Pelvic and spinal X-rays were unremarkable, serum calcium normal with an alkaline phosphatase (AP) of 240 IU/l (38-126), thought to be raised postpartum. She was treated with physiotherapy and anti-inflammatories for several years during which time her pain and spinal restriction worsened. Her AP remained mildly elevated at 132 IU/l with normal calcium. Repeat X-rays showed squaring of the vertebrae in the lumbar spine and osteophyte formation in both hips. Most recent X-rays show diffuse bridging syndesmophytes throughout the lumbar spine. Pelvic X-rays revealed normal SIJs. In view of the unusual presentation an MRI was performed. This revealed normal appearances of both SIJs with no ossification of longitudinal or interspinous ligaments. There was straightening of the lumbar spine with superior end plate bowing of the vertebrae more in keeping with OM and no evidence of inflammation. Subsequent biochemical analysis revealed a raised parathyroid hormone (PTH) of 104 µg/ml (8-73), low 25OH vitamin D2 of <4.0 mmol/l, phosphate 0.73 mmol/l (0.8-1.5) and normal calcium. Review of her notes revealed a low phosphate level of 0.38 mmol/l a year prior to initial presentation. She was commenced on calcium and vitamin D supplements. After therapy her repeat PTH level was 17.2 µg/ml. Presently some of her musculoskeletal symptoms have improved but she continues to have severe restriction of spinal and hip movements.

**Results:** Hypophosphatemic OM mimicking AS is rare. It was first described by Frame et al. in 1961. The hip and spine are most commonly affected. Radiological findings include calcification and ossification occurring at the attachment of ligaments and tendons. Squaring of the vertebral bodies and bridging syndesmophyte formation in the lumbar and thoracic regions mimic AS. Calcification of the pelvic and sacroiliac ligaments may obscure the SIJs giving them an ankylosed appearance. MRI reveals the joints to be normal, with no evidence of inflammation or bone oedema but other changes of OM.

**Conclusions:** Hypophosphataemic OM is rare but early recognition and treatment may prevent serious disability. Unusual patients or findings on X-ray or MRI should prompt further investigation.

**Disclosure:** The authors have declared no conflicts of interest.

#### 468. OESOPHAGEAL INVOLVEMENT IN WEGENER'S GRANULOMATOSIS

Kiran K. Putchakayala, M. N. Choudhry, J. D. Schofield, L. Waller and I. N. Bruce  
 The Kellgren Centre for Rheumatology, Central Manchester & Manchester Children's University Hospitals NHS Trust, Manchester, United Kingdom

**Background:** Wegener's Granulomatosis (WG) is a systemic disease of unknown aetiology characterized by necrotising granulomatous inflammation, tissue necrosis and vasculitis affecting small and medium sized blood vessels. The classic clinical triad involves the upper respiratory tract, lungs and kidneys. Although multi-system manifestations are well recognized, involvement of the gastrointestinal tract is uncommon. We describe a patient with oesophageal involvement presenting with dysphagia and odynophagia.

**Methods:** A 44 yr old woman presented with fatigue, livedo reticularis and a haemorrhagic area on her soft palate in October 2005. She was diagnosed with limited WG based on presenting symptoms, a positive C-ANCA, and a gingival histology consistent with WG.

Over the next year she complained of recurrent mouth ulcers associated with rhino sinusitis, oral candidiasis and weight loss. In October 2006 methotrexate was added to prednisolone in an attempt to suppress disease activity. Six months later she developed dysphagia and odynophagia. Gastroscopy showed oesophageal candidiasis which was treated with fluconazole and a proton pump inhibitor. Her repeat gastroscopy with biopsy revealed extensive oesophageal ulceration, complete resolution of candidiasis, and changes consistent with active vasculitis.

**Results:** Aggressive immunosuppressive therapy was given in the form of pulsed corticosteroids and cyclophosphamide. Treatment was complicated by pancytopenia and deranged liver function tests. In October 2007 repeat gastroscopy demonstrated more extensive abnormalities along the entire length of the oesophagus but with no evidence of candidiasis. There were multiple areas of hyperplasia and sinus formation with epithelialized ulceration. Video fluoroscopy confirmed oesophageal ulceration with lesions also noted above the epiglottis.

In view of the refractory oesophageal involvement and progressive weight loss, treatment with Rituximab was commenced. Unfortunately the patient developed pulmonary haemorrhage and Pneumocystis jirovecii pneumonia which precipitated her demise.

**Conclusions:** This patient demonstrated oesophageal involvement due to WG with dysphagia and odynophagia resistant to immunosuppressive therapy. Our literature review revealed only four previous reports of symptomatic oesophageal vasculitis in patients with WG and only one case report with dysphagia as the dominant symptom.

**Disclosure:** The authors have declared no conflicts of interest.

#### 469. CASE REPORT- ANTIPHOSPHOLIPID SYNDROME AND TAKAYASU'S ARTERITIS

Lubna Aslam<sup>1</sup>, Fahim Tunekar<sup>2</sup> and David D'Cruz<sup>1</sup>  
<sup>1</sup>Rheumatology, Guy's & St.Thomas' Hospital, London, United Kingdom and <sup>2</sup>Histopathology, Guy's & St.Thomas' Hospital, London, United Kingdom

**Background:** Vasculitis and antiphospholipid syndrome (APS) rarely co-exist. We describe a man with APS and Takayasu's arteritis (TA) presenting with multiple episodes of thromboembolism secondary to a combination of thrombotic and inflammatory processes.

**Methods:** A 19 year old gentleman was admitted with left sided weakness in January 2005. MRI confirmed a left cerebellar infarct. He was sent home on low dose aspirin having fully recovered.

Two months later, he was re-admitted with a left superficial femoral artery occlusion requiring embolectomy. Histology revealed granulomatous intimal inflammation of femoral artery. Subsequently, his thrombophilia, vasculitis screen, and antiphospholipid antibodies were negative. He was sent home on long term warfarin.

He was referred to our unit in September 2005. He smoked 10-20 cigarettes a day with left leg claudication distance of approximately 200 meters. Examination showed livedo reticularis and diminished left foot pulses. His investigations were normal apart from a positive lupus anticoagulant on 2 occasions. He was treated with high intensity anticoagulation (target INR 3-4) in view of the previous arterial thromboses.

He stopped his warfarin in July 2006 and subsequently presented with acute left leg ischaemic pain and absent foot pulses. Ultrasound angiography of the left leg showed significant Lt. superficial femoral artery occlusion at its origin. Mesenteric angiogram showed multiple visceral aneurysms.

His PET scan was negative and his repeat inflammatory markers were normal. He received IV heparin, warfarin and iloprost infusion. Despite being on 20 mg of warfarin, his INR remained sub-therapeutic requiring enoxaparin 80 mg if INR <2.

In January 2007 he suffered an acute left middle cerebral artery territory infarct. CT angiography showed thickening of the left internal carotid artery suggestive of vasculitis. He was therefore given 3 infusions of methylprednisolone followed by cyclophosphamide pulses-500 mg fortnightly for 3 months and phenindione was commenced in view of his warfarin resistance.

**Results:** Currently, he is on maintenance treatment with azathioprine. His INR remains stable on phenindione with no further events.

**Conclusions:** The association between APS and TA is rare, however the combination not only results in catastrophic outcomes due to synergistic effects on vessel wall either secondary to thrombosis or vascular inflammation, but also

makes management a difficult challenge as anticoagulation in the presence of visceral aneurysms can lead to bleeding with grave consequences.

In summary, we describe a complex patient with a combination of TA and APS with a good clinical outcome following anticoagulation and immunosuppression.

**Disclosure:** The authors have declared no conflicts of interest.