

Weber-Christian disease associated with lupus anticoagulant and anticardiolipin antibodies

SIR, Weber-Christian (W-C) disease is a process of unknown aetiology characterized by recurrent fever and inflammation of the adipose tissue. Pathological studies disclose areas of fat necrosis with an inflammatory infiltrate showing a lobular pattern and the usual presence of macrophages with foamy cytoplasm [1]. The clinical signs include tender, palpable nodules, located mainly in the extremities, and fever, although abdominal pain, arthritis and arthralgias, and hepatosplenomegaly have also been reported.

Although the cause of this disease is unknown, an immune mechanism has been implicated in the pathogenesis, and it is frequently associated with haematological abnormalities and autoimmune diseases.

We present a case of W-C disease in which the presence of lupus anticoagulant and anticardiolipin antibodies (ACA) was detected. To the best of our knowledge, there are no previous reports of this association (MEDLINE, from 1966 to 1998).

The patient was a 54-yr-old man who began to note fever, which persisted over 1 month, abdominal pain and the presence of tender s.c. nodules in both thighs in 1992. Physical examination revealed painful s.c. nodules measuring between 1 and 3 cm in both thighs and epigastric pain on palpation; there was no evidence of masses or organomegaly. Laboratory analyses disclosed the presence of leucocytosis with neutrophilia (22 000 cells/mm³; 85% segmented) and an erythrocyte sedimentation rate of 35 mmHg; the haematocrit and haemoglobin levels and platelet count were normal. There was no evidence of abnormality in amylase and lipase levels, eosinophil chemotactic activity, α -1-antitrypsin levels and liver and kidney functions. The serum electro-

phoretic mobility and immunoglobulin levels were normal, as were rheumatoid factor, antinuclear antibodies, and complement components C3 and C4. The patient tested positive for lupus anticoagulant (DVVtest and DVVconfirm, Russell's Viper Venom Time test by American Diagnostica). During the course of active disease, he presented elevated ACA levels: IgG ACA, 126 U/GPL; IgA ACA, 11 U/APL; normal value: <22 U (indirect ELISA using REAADS anticardiolipin IgA and IgG semiquantitative 96-microwell test). Blood cultures were negative, as were the Ziehl-Neelsen stain in sputum, urinalysis and serological tests for the most common microorganisms in our environment. Bone marrow biopsy and aspirate disclosed mild hypercellularity, with concomitant granulocyte hyperplasia and reactive changes. Chest X-ray demonstrated the presence of a small pleural effusion and linear atelectasis at the base of the left lung. Colonoscopy and barium enema revealed no abnormal findings. Abdominal computed tomography demonstrated diffuse thickening of the mesenteric fat. The biopsy of the s.c. nodules revealed a lobular inflammatory lesion presenting acute and chronic infiltration with abundant foamy histiocytes and giant cells; there was no vascular involvement, but small areas of necrosis were observed. The patient was treated with steroids over a 1-yr period, during which time improvements were observed in the clinical and analytical findings and imaging studies. The symptoms recurred upon discontinuation of therapy, which was immediately reinitiated. During the period of clinical remission, the patient presented lower titres of ACA (IgG ACA, 66 U/GPL; IgA ACA, 7 U/APL). There was no evidence of thrombotic events during the follow-up period.

In up to 25% of the reported cases, W-C disease has been associated with other diseases [2], some of them immune related [3], including glomerulonephritis [4], scleroderma, morphea and dermatomyositis [5], systemic lupus erythematosus [2, 6], rheumatoid arthritis [2], autoimmune chronic hepatitis and haemolytic anaemia [3]. Abnormal analytical results are common, particularly hypocomplementaemia, hypercomplementaemia, antinuclear antibodies, antimitochondrial antibodies, anti-smooth muscle antibodies, elevated immunoglobulin levels, hypergammaglobulinaemia and circulating immune complexes [2, 3, 7]. However, a review of the literature has revealed no reports of the association of lupus anticoagulant or ACA that we observed in the presented case. Our patient has presented no thrombotic events, although W-C disease has been associated with mesenteric thrombosis [8], inferior vena cava, pelvic and iliac vein thrombosis [9], internal jugular vein thrombosis, intracranial thrombosis [10] and arterial thrombosis; most of these developments were detected in necropsy studies, and some of them were probably related to the local inflammatory phenomenon that accompanies this disease.

We do not know what significance the association of lupus anticoagulant and ACA with W-C disease may have, but we consider that the presence of both should

be added to the list of findings associated with this intriguing disease.

J. JIMENEZ-MAZUECOS, M. YEBRA-BANGO, A. SANCHEZ-RUIZ, M. VILLARREAL-GARCIA-LOMAS

Internal Medicine Service, Clinica Puerta de Hierro, Universidad Autonoma de Madrid, Madrid, Spain

Accepted 5 March 1999

Correspondence to: M. Yebra-Bango, Servicio de Medicina Interna I, Clinica Puerta de Hierro, C/San Martin de Porres, 4, 28035-Madrid, Spain.

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