Weber Christian Disease Presenting With Pancytopenia and Anticardiolipin Antibodies

Antikardiolipin Antikorları ve Pansitopeni ile Kararize Weber Christian Hastalığı

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Abstract
Weber-Christian disease is a controversial entity that represents recurrent fever, inflammation of adipose tissue and tender subcutaneous nodules. The etiology of WCD remains unknown. However, it has been related to an immunologically mediated reaction to diverse antigenic stimuli. We present a 50 year-old man with Weber-Christian disease presented with fever, recurrent pancytopenia attacks, IgG and IgM anticardiolipin antibody positivity. The patient showed fever and characteristic redish subcutaneous nodules. Biopsy taken from the lesion showed areas of fat necrosis, accumulation of leucocytes and macrophages with foamy cytoplasm, and no evidence of vasculitis. Histopathologic findings were consistent with Weber-Christian panniculitis. Bone marrow aspirate and biopsy disclosed mild hypercellularity with concomitant mild increasing lymphocyte and plasma cells. Patients with pancytopenia, cutaneous lesions and ACA positivity must be searched for WCD.

Key words: Weber-Christian disease, Pancytopenia, Anticardiolipin antibody.

ÖZET

Anahtar kelimeler: Weber-Christian hastalığı, Pansitopeni, Antikardiolipin antikor


INTRODUCTION
Weber-Christian disease (WCD) is a rare idiopathic lobuler panniculitis characterized by fever, arthralgias, subcutaneous nodules and plaques located mainly in the extremities which resolve leaving characteristically depressed atrophic areas (1). There has been an attempt to classify the panniculitis into lobular and septic types and WCD falls into the lobular type. Lobular panniculitis may be seen with infections, alpha-1-antitrypsin deficiency, malignant disease, pancreatitis, systemic lupus erythematosus and cytophagic histiocytic panniculitis (2). Histologic findings include areas of fat necrosis with an inflammatory infiltrate showing a lobular pattern and usual presence of macrophages with foamy cytoplasm (2,3). We present a case of WCD presenting with recurrent pancytopenia and anticardiolipin antibody positivity.

CASE REPORT
The patient was 50 year-old man who had pallor, fever, fatigue and artralgia. In physical examination fever was 39.3°C, oral mucosa was pallor and dry. There was no lymphadenopathy, hepatomegaly splenomegaly and cutaneous lesions. His past medical history, had brucellosis one year ago and using rifampin and streptomycin. Laboratory studies showed a hemoglobin level 3.3 gr/dL, White Blood cell count of 1.410 x 109/L, neutrophil count of 0.144x109/L, and platelet count of 19.800 x 109/L, ESR 120 mm/h. Coagulation and liver function tests were normal but serum concentration of lactate dehydrogenase was elevated to 728 IU/L (Normal:230-460). There was polyclonal gammopathy in serum protein electrophoresis. ANA and anti dsDNA were negative. Brucella aglutination was 1/80 positive. Antibodies including to CMV, EBV, HSV and HIV were negative. Parvo virus IgG and HbAg were positive but, HBV DNA was negative. Bone marrow aspiration and biopsy disclosed mild hypercellularity with concomitant mild increasing lymphocytes and plasma cells. It was thought reactive changes. In thorax CT, fibrotic bands was seen in apical regions. Neutropenic fever was treated with meropenem, amicacin, teicoplanin and amphoterisin B. Pancytopenia resolved and fever declined 25 th days after starting therapy.
Three months after resolving of pancytopenia attack, he suffered from tender, swelling, redish subcutaneous nodules at lower extremities (Fig. 1). Biopsy taken from the this lesion showed areas of fat necrosis, accumulation of leucocytes and macrophages with foamy cytoplasm, consistent with lobular panniculitis (Fig. 2). There was no evidence of vasculitis. Complete blood count showed pancytopenia. With using ELISA technique elevated anticardiolipin antibodies (ACA), IgG ACA 65 GPL/U (N=0-18) and IgM ACA 14.3 MPL/U (N=0-13) was determined. Bronchoscopic examination was performed, bronchoscopic fluid as microbiologically and cytologically was normal. The patient was treated with steroid for six months period and then dosage tapered and stopped. For neutropenic fever; meropenem, amicacin, vancomycine were used. 15th day of therapy; fever declined thrombocytopenia and leucopenia were resolved. In 20th day cutaneous lesions dissappeared leaving depressed atrophic areas. For 6 months he has required no treatment, and remained well and asymptomatic.

**DISCUSSION**

We present a patient with WCD who has relapsing pancytopenia attacks and ACA positivity. The patient showed fever, characteristic cutaneous nodules and skin biopsy changes. In up to 25% of the reported cases of WCD has been associated with glomerulonephritis, scleroderma, morphe and dermatomyositis, systemic lupus erythematosus, rheumatoid arthritis, auto immune chronic hepatitis and haemolytic anemia (4). The etiology of WCD remains unknown. However, it has been related to an immunologically mediated reaction to diverse antigenic stimuli, because of an association in some patients with elevated levels of circulating immune complexes.

Increased serum sIL-2R concentration, which represent T-cell activation and high level of interferon gamma can be seen (1,5). Systemic involvement in WCD remains controversial, however numerous reports suggests a systemic form of the disease. Cutaneous manifestations usually precede systemic involvement. Mesenteric, hepatic, myocardial, retrobulbar and perivisceral fat involvement may occur (6,7,8).

Abnormal analytical results are common particularly hypocomplementemia, hypercomplementemia, antinuclear antibodies, antinitochondrial antibodies, elevated immunoglobulin levels, hypergammoglobulinemia (9). We observed ACA positivity in our patient. We do not know what significance the association ACA with WCD and pancytopenia. It may also be the result of immunologically mediated reaction to diverse antigenic stimuli. Our patient has presented no thrombotic events, although WCD has been associated with mesenteric, inferior vena cava, pelvic, internal jugular vein and intracranial thrombosis (9).

In the management of the patient with WCD underlying causes must be searched for and treated if found. Treatment is symptomatic, systemic corticosteroids are helpful but, recurrences are frequent as the dosage is reduced (10). Azathiopurine, mycophenolate, cyclophosphamide may also be useful (11,12,13). Cyclosporine A is considered to the drug of choice in WCD and it is effective against the disease via suppression of T-cell reactions (14). In conclusion, patients with pancytopenia, cutaneous lesions and ACA positivity should be searched for WCD.
REFERENCES


