Weber Christian Disease Presenting With Pacytopenia and Anticardiolipin Antibodies

Antikardiolipin Antikorları ve Pansitopeni ile Karakterize Weber Christian Hastalığı

MehmetYILMAZ[1], Simten DAGDAŞ[2], Nuran ALLI[3], Sibel ERDOGAN[4], S.Zeynep AKI[1], Gülsüm ÖZET[1]

[1]Gaziantep University School of Medicine Department of Hematology
[2]Ankara Numune Education and Research Hospital, Department of Hematology
[3]Ankara Numune Education and Research Hospital, Department of Dermatology
[4]Ankara Numune Education and Research Hospital, Department of Pathology

Abstract

Weber-Christian disease is a controversial entity that represents reccurent fever, inflamation of adipose tissue and tender subcutaneus nodules. The etiolgy of WCD remains unknown. However, ıt 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, reccurent pancytopenia attacks, IgG and IgM anticardiolipin antibody positivitiy.. 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 hypercellularitiy with concomitant mild increasing lympocyte 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

Weber-Christian hastalığı tekrarlayan ateş, yağ dokusunun inflamasyonu ve ağrılı ciltaltı nodülleri ile karakterize bir hastalıkdır. Weber-Christian hastalığının etyolojisi bilinmemektedir. Fakat çeşitli antijenik stimuluslara yanıt olarak meydan gelen immünolojik kökenli reaksiyonlarla ilişkili olduğu düşünülmektedir. Burada ateş, tekrarlayan pansitopeni atakları, antikardiyolipin IgG ve IgM antikor pozitifliği ile seyreden bir Weber-Christian olgusu sunulmaktadır. Hastada karakteristik kırmızımsı cilt altı nodülleri ve ateş saptandı. Lezyonlu bölgeden alınan biyopside vaskülit bulguları olmaksızın; köpüklü stoplazmalı makrofajlar, lökosit birikimi ve yağ nekrozu izlendi. Histopatolojik bulgular Weber-Christian panniküliti ile uyumlu idi. Kemik iliği aspirasyon ve biyopsisinde hafif lenfosit ve plasma hücre artışı ile birlikte hafif bir sellülarite artışı izlendi. Pansitopeni, cilt lezyonları ve antikardiyolipin antikor pozitifliği Weber-Christian hastalığı yönünden araştırılmalıdır.

Anahtar kelieler: Weber-Christian hastalığı, Pansitopeni, Anticardiyolipin antkor

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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 lobuler and septal types and WCD falls into the lobuler type. Lobuler panniculitis may be seen with infections, alpha 1-antityripsin deficiency, malignant disease, pancreatitis, systemic lupus erytematosus and cytophagic histiocytic panniculitis (2). Histologic findings include areas of fat necrosis with an inflamatory infiltrate showing a lobuler pattern and usual presence of macrophages with foamy cytoplasma (2,3). We present a case of WCD presenting with reccurent pancytopenia and anticardiolipin antibody positivity.

Yazışma Adresi:
Dr.Mehmet YILMAZ Gaziantep Üniversitesi Tıp Fakültesi
Hematoloji ABD
Adres: Gaziantep Üniversitesi Tıp Fakültesi
Şehitkamil/Gaziantep
Tel: 0 342 360 6060 / 76128
Fax: 0 342 360 13 14
E-mail: mmyilmaz@gantep.edu.tr

CASE REPORT

The patient was 50 year-old man who had pallor, fever, fatique and artralgia. In physical examination fever was 39.3 ºC, 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 streptomycine. Laboratory studies showed a hemoglobin level 3.3 gr/dL, White Blood cell count of 1.410 x109/L, neutrophil count of 0.144x109/L, and platelet count of 19.800 x109/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 policlonal gammapaty in serum protein electrophoresis. ANA and anti dsDNA were negative. Brucella aglitunation was 1/80 positive. Antibodies including to CMV, EBV, HSV and HIV were negative. Parvo virus IgG and HbsAg were positive but, HBV DNA was negative. Bone marrow aspiration and biopsy disclosed mild hypercellularitiy with concomitant mild increasing lympocytes and plasma cells. It was thought reactive changes. In thorax CT, fibrotic bands was seen in apical regions. Neutropenic fever was treated with meropenem, amicasin, teicoplanin and amphoterisin B. Pancytopenia resolved and fever declined 25 th days after starting therapy.

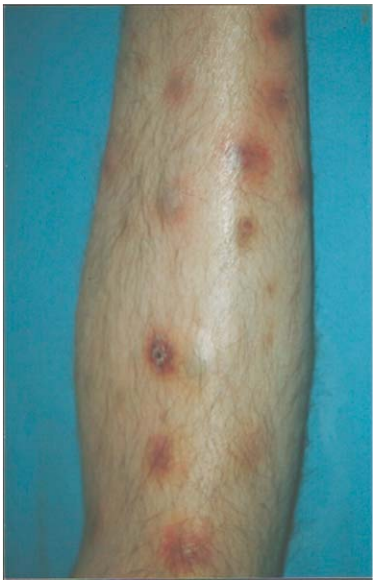

Figure 1. Redish, tender and painful subcutaneous nodules at lower extremity.

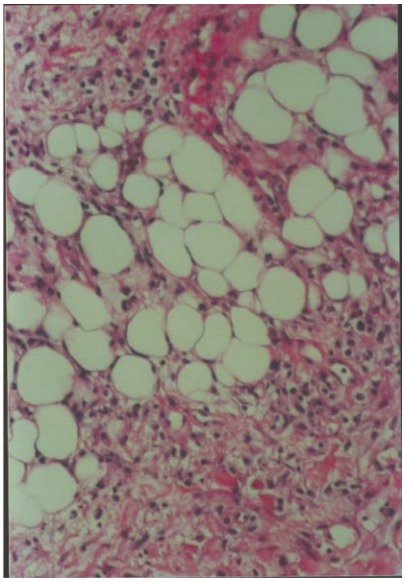

Figure 2. Skin biopsy from the subcutaneus nodules at the lower extremity. Adipose tissue is infiltrated with inflammatory cells and fat-laden macrophages.Fat degeneration and fibrosis is seen (HE stain x 200).

Three months after resolving of pancytopenia attack, he suffered from tender, swelling, redish subcutaneous nodules at lower extremitias (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 ELİSA 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 microbiolgically and cytologically was normal. The patient was treated with steroid for six months period and than dosage tapered and stopped. For neutropenic fever; meropenem, amicasin, 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 dermathomyositis, systemic lupus erythematosus, rheumatoid arthritis, auto immun chronic hepatitis and haemolytic anemia (4). The etiolgy of WCD remains unknown. However, ıt 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.

Inreased 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, antimithocondrial antibodies, elevated immunglobulin levels, hypergammaglobulinemia (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 diversre 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 searced for and treated if found. Treatment is symptomatic, systemic corticosteroids are helpful but, reccurences 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 supression of T-cell reactions (14). In conclusion, patients with pancytopenia, cutaneous lesions and ACA positivity should be searched for WCD.

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