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Weber-Christian Disease Presenting with Proptosis : A Case Report

Weber-Christian disease (WCD) is a rare inflammatory disease of adipose tissue, which is characterized by painful cutaneous nodules and constitutional symptoms. Although any area of the body containing fat can be affected by WCD, the involvement of retrobulbar fat is uncommon and proptosis is a rare presenting manifestation. We report a case who presented with proptosis of the right eye which is accompanied by painful subcutaneous nodules, high fever and myalgia. Biopsies of retrobulbar tissue and suprapubic nodule showed lobular panniculitis with mixed cellular infiltration, mainly composed of histiocytes and lymphocytes. He responded well to high-dose glucocorticoid.

Key Words: *Panniculitis, Nodular Nonsuppurative; Exophthalmos, Proptosis*

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INTRODUCTION

Weber-Christian disease (WCD) was first described by Pfeifer in 1892, and more clearly defined by Weber and Christian in the 1920s (1). It is a relapsing, febrile nodular nonsuppurative panniculitis which is characterized by painful subcutaneous nodules and constitutional symptoms such as fever, malaise, arthralgia and myalgia (1). The ocular manifestations of WCD include anterior and posterior uveitis (2), macular hemorrhage (3), and episcleral and conjunctival nodules (4). Proptosis, which is caused by the panniculitis of orbital fat, is a rare manifestation of WCD (5). We report a patient with WCD who had proptosis as the presenting sign and was treated successfully with high-dose glucocorticoid.

CASE REPORT

A 34-year-old man presented with a 2-month period of high fever and ocular pain with proptosis of the right eye. His family history was not significant. He had a history of high fever and painful subcutaneous nodules about two years ago which were improved after taking medicine for a few weeks. He had myalgia and painful subcutaneous nodules in the suprapubic and epigastric areas. He was treated with antibiotics for presumed orbital cellulitis without improvement of symptoms at an ophthalmologic clinic. Physical examination showed that body temperature was 40℃, pulse rate 100/min and blood pressure 120/80 mmHg. The right eye was protruded with chemosis and injection of conjunctiva, and with periorbital swelling (Fig. 1). Two tender nodules were palpated over the epigastric and suprapubic areas,

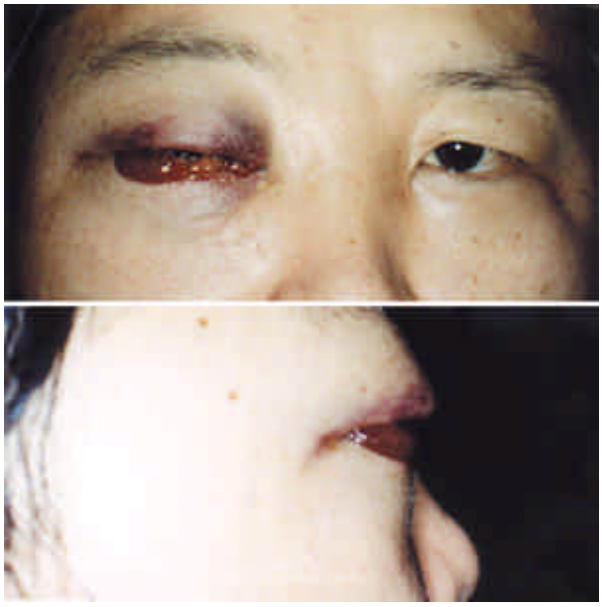

Fig. 1. Photographs of the patient show severe proptosis and chemosis of the right eye and periorbital facial swelling.

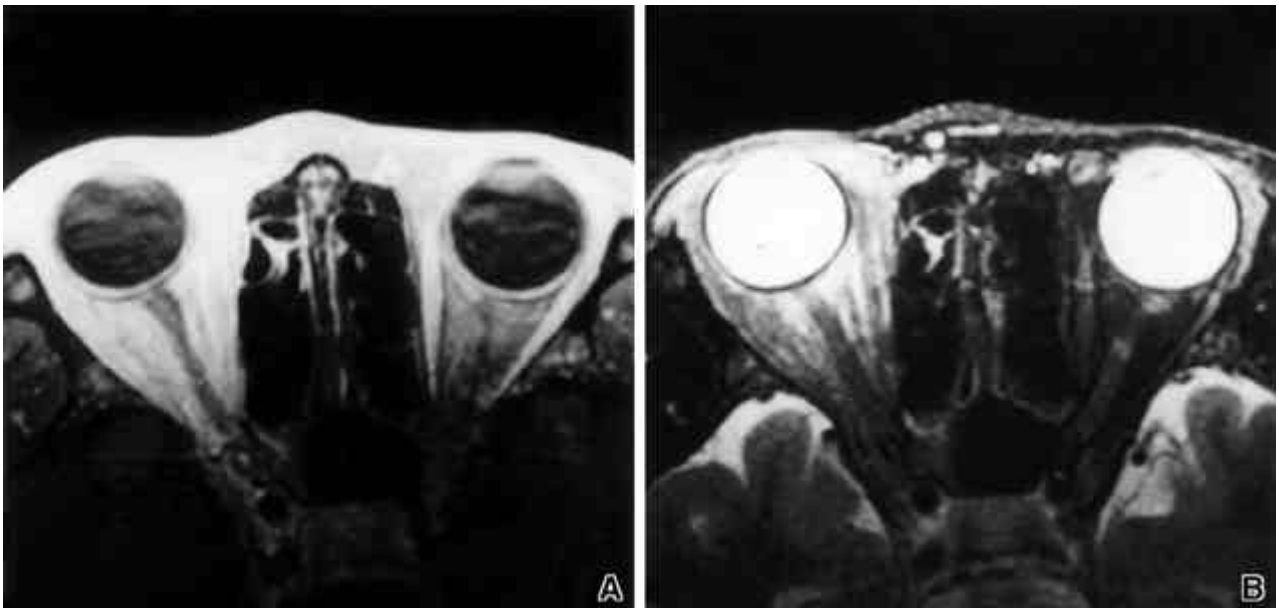

Fig. 2. Orbital MRI shows high signal intensity lesions in retrobulbar space with proptosis of the right eye on enhanced T1WI (A) and T2WI (B).

of which the latter was an aggregated nodule forming a mass. Lung and heart were normal. Liver and spleen were slightly enlarged and multiple enlarged lymph nodes were palpated on inguinal areas. Anterior and posterior chambers were unremarkable on ophthalmoscopic examination.

Results of laboratory studies were as follows: hemoglobin 13.0 g/dL, white blood cell 2,730/μL (neutrophil 66.7%, lymphocyte 19.9%, monocyte 7.1%, eosinophil 1.2%), platelet 177,000/μL, ESR (Westergren method) 12 mm/hr, C-reactive protein 2.45 mg/dL and cholesterol 115 mg/dL. Among lymphocytes, T cell (CD3+) was 87%, B cell (CD19+) 5%, NK cell (CD56+) 5%, and CD4+/CD8+ T cell ratio 1.5:1. Serum AST was 223 mg/dL, ALT 73 mg/dL, LDH 2,176 IU/L, and creatine phosphokinase 196 mg/dL. Serum amylase and lipase were within normal range. Serum complements and immunoglobulins were normal. Antinuclear antibody and rheumatoid factor were negative. Hepatitis C viral antibody was positive. Hepatitis B surface antigen and HIV antibody were negative. No organisms were cultured in blood, retrobulbar and suprapubic subcutaneous tissues. Orbital magnetic resonance imaging revealed proptosis of the right eye, which was caused by soft tissue inflammation of the retrobulbar tissue sparing extraocular muscles and optic nerve (Fig. 2). Biopsies of retrobulbar and suprapubic tissues showed lobular panniculitis with mixed cellular infiltration, mainly composed of histiocytes and lymphocytes (Fig. 3). He was treated with high-dose glucocorticoid (prednisolone 1 mg/kg/day) followed by gradual tapering. There was an improvement of constitutional symptoms, disappearance of subcutaneous nodules and proptosis. After treatment, the right eye was depressed.

DISCUSSION

Weber-Christian disease, known as recurrent febrile nodular panniculitis, is an infiltrative, inflammatory disease of fat characterized by multiple recurrent subcutaneous nodules with accompanying fever. Other constitutional symptoms include malaise, arthralgia, hepatosplenomegaly, anorexia and weight loss (1, 6). Although nodules usually appear on the lower extremities, trunk, and upper extremities, any area of the body containing fat such as mesentery (7), heart, lung (8) and liver can be affected by WCD. Panniculitis of orbital fat, especially accompanied by proptosis, has rarely been reported. Cook and Kikkawa described a case of WCD manifesting proptosis in whom orbital computed tomographic scan showed diffuse thickening of Tenon capsule, orbital fat shadowing and soft tissue inflammation in the orbit (5). In their case, orbital panniculitis was not confirmed by biopsy. In our case, magnetic resonance imaging showed soft tissue inflammation of orbital tissue with intact extra-ocular muscles, and Tenon capsule, but it was indistinguishable from those of other inflammatory diseases such as orbital cellulitis (10). There are no specific findings of lobular panniculitis on orbital computed tomography or magnetic resonance imaging and histopathologic confirmation is essential for the diagnosis of

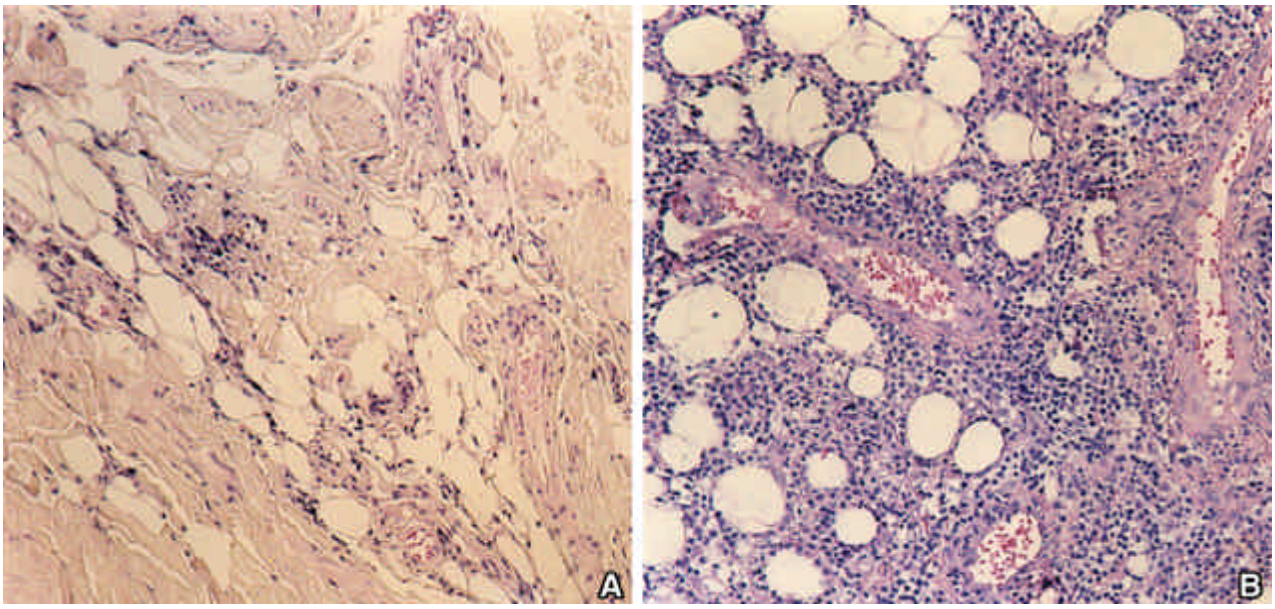

Fig. 3. Biopsy specimens of retrobulbar (A) and suprapubic (B) subcutaneous tissue show lobular panniculitis characterized by infiltration of inflammatory cells, mainly composed of lymphocytes and histiocytes (H&E; ×100).

lobular panniculitis. In our case, retrobulbar tissue biopsy showed typical findings of lobular panniculitis. In respect to the large proportion of fat in the retrobulbar tissue, the rarity of involvement of orbital fat in comparison with subcutaneous tissue needs further research in the pathogenesis of WCD. There have been reports describing ocular involvement such as episcleral and conjunctival nodules (4), recurrent uveitis (2), and retinal vascular abnormalities (3). This case showed no intraocular abnormalities except for marked conjunctival injection and chemosis which was thought to be caused by proptosis and retrobulbar inflammation. Histopathologic findings of lobular panniculitis change with the stages of disease (11). Early infiltration of inflammatory cells predominantly consisted of lymphocytes and monocytes is eventually replaced by fibrotic tissue often leaving an atrophic scar which may be the possible cause of the depression of proptotic eye after treatment with corticosteroid in this case.

The cause of Weber-Christian disease is not known but there are some evidence that immune mechanisms may play a pathogenic role. Immunologic abnormalities such as increased circulating immune complexes (12, 13), impaired cellular immunity (14) and concomitant occurrence of autoimmune disorders (15) were suggested as evidence of immune mechanism as a cause. Therapeutic response to high-dose glucocorticoid and immunosuppressives such as azathioprine (16) also support the immune mechanism. In our patient, there were no detectable abnormalities in immunologic tests, including autoantibodies, complements, and immunoglobulin levels, but the response to high-dose glucocorticoid treatment has been good.

In our case, proptosis caused by lobular panniculitis was confirmed histologically. We recommend that WCD should be included in differential diagnosis of proptosis and considered especially in the presence of constitutional symptoms and cutaneous nodules.

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