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Case Note

Idiopathic lobular panniculitis (Weber Christian disease): A case report

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Abstract
Weber Christian disease, an idiopathic relapsing febrile subcutaneous and visceral panniculitis is a rare disease in children. We report a case of Weber Christian disease for he first time from Kanti Children's Hospital.

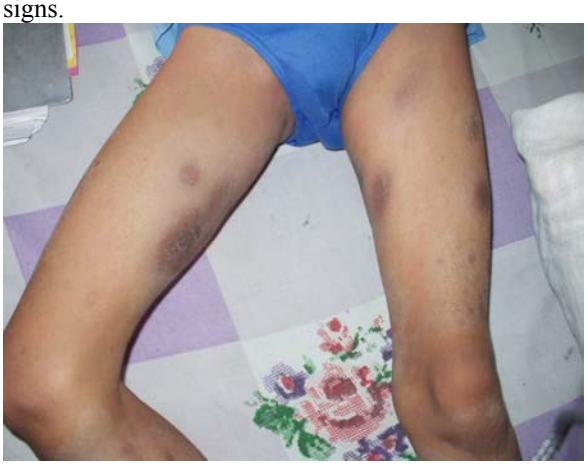
Pfeifer Weber Christian disease, a relapsing, febrile subcutaneous and visceral panniculitis is an extremely rare condition in children.[1-3] The involved areas in the skin manifest recurrent crops of erythematous, sometimes tender edematous subcutaneous nodules.[2,3] Although first described in 1892 AD, the aetiology is still unknown; therefore Weber Christian disease is often referred to as idiopathic lobular panniculitis.

Case report

A twelve year old girl presented with two and half month history of multiple nodular painful swelling in her legs and back associated with high grade fever with chills and rigors. She also complained of non disabling joint pains that have never been associated with features of arthritis. There was no history of suggestive viral infection, exposure to pets, travel, use of drugs prior to the onset of problems, loose or bloody stools and urinary problems. There was no known tubercular contact or a family history of collagen disorder. After the onset of the problems she had received multiple antibiotics without relief and without any change in her skin lesions. On examination she was febrile with a pulse rate of 130/min and blood pressure of 115/70 mm of Hg. She was not pale and had no jaundice, oedema of limbs or lymphadenopathy. There was no evident rash or sign of bleeding. There were multiple tender nodular subcutaneous swellings of various sizes in the legs bilaterally and on the back. The rest of the systemic examination was unremarkable.

Her investigations showed haemoglobin of 10.5gm/dl, white cell count of 6700/□l, N-62%, L-32%, E-04%, B-0%, ESR was 61mm/ 1st hr, Platelet 350000 /□l, PCV-33.6%, MCV-77.9fl, MCH-29.8pg and MCHC-31%. The serum transaminases, serum amylase and serum calcium were all normal. The renal functions and the electrolytes were also all normal. The urine was free of any sediments or protein. The chest radiograph showed no lung or mediastinal lesions and the abdominal sonogram also was reported as normal. The serum antinuclear antibody screen with ANA and anti dsDNA ELISA showed negative results for both. Biopsy of the skin nodule revealed a lobular panniculitis without significant septal involvement or vasculitis as seen in Weber Christian disease. Serum alpha 1 antitrypsin levels couldn't be done due to lack of investigative facilities.

A diagnosis of Weber Christian disease was made based on clinical and laboratory parameters. The patient was started on 2mg/kg of daily steroids which rapidly led to improvement of her symptoms and signs.



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Discussion

Idiopathic lobular panniculitis, originally described by Weber and Christian, is characterized by a systemic inflammatory process of adipose tissue of unknown aetiology. The term Weber-Christian disease is applied when cutaneous lesions are associated with systemic manifestations. [2-4] The course of the disease is characterized by acute onset with fever and malaise and the simultaneous appearance of painful subcutaneous nodules. [2-5] Some patients experience involvement of the intra-abdominal or retroperitoneal fat. The disease is most frequently reported in the fourth to seventh decades of life but is also seen in younger children. The disease occurs more often in women, who comprise approximately 75% of reported cases.[2,4,5]

Patients affected with Weber Christian disease describe crops of lesions that appear and resolve during a period of weeks to months. The lesions are often symmetric in distribution and the thighs and legs are most commonly involved sites. Individual nodules regress during the course of a few weeks.[2-6] Systemic symptoms include malaise, fever, nausea, vomiting, abdominal pain, weight loss, bone pain, myalgia, arthralgia and hepatosplenomegaly.[2-5] Weber-Christian disease may involve the lungs, heart, intestines, spleen, kidney, and adrenal glands. There have been few reports of pleural involvement, joint effusions, bowel perforation secondary to mesenteric disease, anaemia due to marrow infiltration with lipid laden macrophages and amyloidosis in Weber-Christian disease and other types of panniculitis.[7,8] Children with early onset disease have significant failure to thrive and recurrent infections. In patients with inflammation involving visceral organs, significant morbidity and mortality is usual.[7]

Routine laboratory evaluation is often noncontributory; changes in liver function tests, complete blood count and electrolytes usually reflect visceral involvement of organs including the lungs, heart, intestines, spleen, kidneys and adrenal gland. Patients may present with leucocytosis or leucopoenia, anaemia or hypocomplementemia. ESR is usually elevated.[2,3,9] The histopathological findings observed in Weber Christian disease are considered characteristic and include lobular panniculitis without septal involvement or vasculitis.[5] The diagnosis is therefore based on the characteristic lobular panniculitis seen on biopsy of inflammatory skin nodules after other types of panniculitis, such as those associated with pancreatitis or alpha1-antitrypsin deficiency, have been excluded.[5] Some pathologists also stage the disease histologically as [2,9]

Stage 1 characterized by an acute inflammatory reaction, in which lobules of fat are replaced by neutrophils, lymphocytes and histiocytes.

Stage 2 macrophages migrate and phagocytose degenerated fat, forming characteristic foam cells.

Stage 3 the foam cells are replaced by fibroblasts, and the inflammatory reaction is replaced by fibrotic tissue.

No uniformly effective therapy exists. Therapeutic responses have been reported with the use of fibrinolytic agents, hydroxychloroquin, azathioprine, thalidomide, cyclophosphamide, tetracycline, cyclosporin and mycophenolate mofetil.[2,3,10] Systemic steroids are usually effective in suppressing acute exacerbations but cause problems with prolonged use. Non-steroidal anti-inflammatory agents may reduce fever, arthralgias and malaise.

No effective methods of prevention are known. Prognosis is highly variable. The clinical course may be characterized by exacerbations and remissions of the cutaneous lesions for several years before the disorder resolves. Patients with severe systemic disease have high mortality.[6]

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