Weber Christian Disease

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Abstract
Weber Christian disease or idiopathic lobular panniculitis is a skin condition that features recurring inflammation in the fat layer of the skin. We report a case of a 17 year old boy with Weber Christian disease as its occurrence is extremely rare with only sparse case reports available, who presented with eighteen-month history off fever with chills off and on, arthralgia predominantly of the knees, ankles, wrists and elbows without swelling or gross limitation of movement. There was also swelling around the eyes and puffiness of the face with lumpy swellings on the jaw, cheeks and chin and painful lumps on the anterior abdominal wall. On examination a febrile patient with pallor, pitting edema of the legs, periorbital edema and multiple non-matted firm to hard tender swellings of the face, jaw and chin and multiple firm tender lumps in the anterior abdominal wall was noted. Investigations revealed anemia, leucopenia, raised sedimentation rate and mild rise of liver enzymes. Biopsy of the abdominal nodule demonstrated a lobular panniculitis without vasculitis as seen in Weber Christian disease.

INTRODUCTION
Weber Christian disease or idiopathic lobular panniculitis is a skin condition that features recurring inflammation in the fat layer of the skin. The involved areas of the skin manifest as recurrent crops of erythematous, sometimes tender edematous subcutaneous nodules. Because its etiology is unknown Weber Christian disease is often referred to as idiopathic lobular panniculitis.

CASE REPORT
A seventeen-year-old boy presented with an eighteen-month history of fever with chills off and on. He also had arthralgia of all joints predominantly knees, ankles, wrists and elbows although there was no swelling or gross limitation of movement at the joints. He had periorbital edema, swollen face around the cheek, jaw and chin and painful lumps on the abdominal wall. His mother had SLE and she was on azathioprine. On examination he was febrile, temp 100o F, pulse 100/min, BP 100/80,marked pallor, pitting edema on the legs and periorbital edema. There were multiple, non-matted firm to hard tender swellings on the face, cheek jaw and chin regions, and multiple firm tender lumps in the abdominal wall around the umbilicus.

Except for a mild hepatomegaly, the other systemic examination was normal. Complete blood count revealed Hb-10gm/dl, WBC-2800/cu mm, N-65%, E-01%, L-29%, M-05%. Platelet count 4.05 lakhs/cu mm,PCV-37.6%, MCV-67.9fl, MCH-23.8pg and MCHC-35%. There was mild rise of liver enzymes, AST-63U, ALT-49U, bleeding time-3mins, clotting time-3mins 30 secs, prothrombin time 11.8secs. control being 10.5 secs. Sedimentation rate 75mm/hr, 24 hr urinary sample showed no protein and serum calcium was 10.7mg/dl with alkaline phosphatase-120IU/ml. The antinuclear antibody was negative being 0.42 od ratio (+ve >1) by Eliza test, serum complement levels C3-161 mg%(N-83-177mg%), C4levels 30mg%(N- 15-60mg%), Brucella IgG-negative, IgM negative, alpha1 anti-trypsin level-2.25gm/dl (n-9.2gm/dl) and TSH-1.9IU/ml. Serum amylase and serum lipase were normal. Biopsy of the nodule revealed a lobular panniculitis without vasculitis as seen in Weber Christian disease. Biopsy with immunochemistry CD1, CD A and B cell markers, leucocyte common antigen were normal. Immunohistochemistry profile showed T lymphocyte cell markers. The patient was put on azathioprine 75mg, which showed marked improvement and regression of the edema and size of the nodules. Below are the patient’s photographs before and after therapy. Follow up investigations showed Hb 12.5gm/dl, WBC 6700/cu mm, N-62%, L-32%, E-04%, B-0%. ESR-11mm/hr, AST-37U, ALT-22U.

DISCUSSION
In this case other conditions considered for differential diagnosis were a cryptogenic histiocytic panniculitis or cutaneous lymphoma, or Epstein Barr virus infection or a subcutaneous T cell lymphoma causing panniculitis, but immunohistochemistry CD1, CDA and B cell markers and leucocyte common antigen were normal. Other differential
diagnosis like alpha1 antitrypsin deficiency panniculitis, cutaneous polyarteritis nodosa, eosinophilic fasciculitis, scleroderma panniculitis, lupus panniculitis, panniculitis associated with pancreatic disease and erythema nodosum were all ruled out.

Weber-Christian disease may occur in young children, but has been reported most frequently in the fourth to seventh decades of life. 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 involved more commonly. Individual nodules regress during the course of a few weeks. Occasionally the epidermis overlying the nodule breaks down and the lesion discharges a brown liquid oil called liquefying panniculitis. Systemic symptoms include malaise, fever, nausea, vomiting, abdominal pain, weight loss, bone pain, myalgia and arthralgia. In individuals with visceral involvement, hepatomegaly or splenomegaly may be present. Changes in liver function tests, complete blood count and electrolytes reflect visceral involvement of organs including the lungs, heart, intestines, spleen, kidneys and adrenal gland. Patients may present with leucocytosis or leucopenia, anemia or hypocomplementemia. ESR is usually elevated.

Three histopathological stages are observed in Weber-Christian disease:

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. Systemic steroids may be effective in suppressing acute exacerbations. Non-steroidal anti-inflammatory agents may reduce fever, arthralgias and other signs of malaise. Involvement of specific organs may require specific supportive drugs. No surgical treatment is indicated. When the condition subsides prophylaxis may be unnecessary. No effective methods of prevention have been discovered. 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 visceral inflammation of lungs, heart, spleen, kidney, intestines or adrenal glands may not survive.

REFERENCES

2. Lazarus GS: Panniculitis and Disorders of the Subcutaneous Fat.