Weber Christian disease or relapsing febrile nodular non suppurrative panniculitis was described separately by Weber and Christian in 1925 and 1928 respectively. It is associated with a skin condition that features recurring inflammation in the fat layer of skin (panniculitis). The involved areas of skin become reddish, tender and raised (inflamed). It usually involves both sides of the body and the thighs and lower legs are the most frequently affected areas. The inflamed areas can lose their blood supply and the skin can actually die in the area, a yellowish discharge results and it become infected. Systemic nodular panniculitis may exist by itself or may accompany systemic lupus erythematosus (SLE), diabetes mellitus, infections like tuberculosis, multiple myeloma, lymphoproliferative neoplasia and Hashimoto’s thyroiditis.

We are reporting here a case of Weber-Christian disease associated with type 2 diabetes.

Case Report

Mr. SPC, a 62 year old man, was admitted to our centre with complaints of swelling both legs, hands and difficulty in breathing. He was diagnosed to have impotaneous edema and was treated with parenteral steroids and antibiotics. He had diabetes of 25 years duration and was treated with twice daily insulin and oral hypoglycaemic drugs. He had multiple trophic ulcers and peripheral neuropathy due to Hansen’s disease. Patient had exfoliation of skin all over the body with small scales with mild itching. He had subcutaneous erythematous nodules over the chest, both hands and abdomen of three weeks duration. The lesions started following a bout of fever. A clinical diagnosis of erythema nodosum was made as he had a past history of Hansen’s disease and also had thickening of the ulnar nerves.

Mr. SPC had a fasting plasma glucose of 226 mg/dl and the glycosylated haemoglobin (HbA1c) was 8.6%. Renal function test was normal. Liver function test showed evidence of albumin : globulin reversal.

Weber Christian Disease in a Type 2 Diabetic Patient

G Premalatha, L Rajmohan, B Srinivas, V Mohan

Total bilirubin : 0.5 mg/dl (0.5 - 1.0 mg/dl);
Total protein : 6.5 (6.5 - 8.0 g/dl);
Albumin : 3.1 (3.0 - 5.0 g/dl);
Globulin : 3.4
SGOT : 15 IU/L (15-35 IU/L);
SGPT : 12 IU/L (10-40 IU/L);
Alkaline phosphatase : 254 IU/L (90-230 IU/L).

Haemogram showed evidence of mild anaemia and elevated ESR.

Total count : 10,600 cells/mm³
Differential count
Polymorphism - 70%. Lymphocytes - 24%, Eosinophilia - 6%
ESR 1 hr : 40 mm
Hb% : 10.6 g/dl
RBC : 3.5 million cells/mm³
PCV : 32%

He was seen by a dermatologist (BS) who carried out a biopsy on the erythematous nodule. The biopsy report showed that the epidermis and dermis were normal. The subcutis showed ingestion of fat cells by macrophages and foam cells. A dense infiltrate consisting of numerous neutrophils were seen around the fat cells and peri-vascularly. Large giant cells with ingested fat (foam cells) were also seen (Fig. 1). A diagnosis of Weber Christian (W-C) disease was made. Patient was treated with parenteral steroids for a week followed by oral steroids to which he responded. Two months later the patient returned for a diabetic checkup and the lesions had completely disappeared.

Discussion

Systemic nodular panniculitis also called Weber-Christian disease is a syndrome that may exist by itself or may accompany SLE or pancreatic disease. The syndrome is characterized by recurrent attacks of crops of painful, red, tender, subcutaneous nodules, most commonly located on the legs and buttocks. The outbreaks are accompanied by fever, leukocytosis and eosinophilia. Attacks are self-limiting, lasting two to three weeks. In severe cases, any area of the body may be involved except the face; there may also be extraneous sites of involvement, such as the mesentery. Localisation of the inflammatory nodules in periarticular areas leads to acute arthritis of one or more joints.

Differential diagnosis of Weber Christian disease
include erythema nodosum, subcutaneous nodules in rheumatoid arthritis, erythema induratum, cold panniculitis, post-steroid panniculitis and subcutaneous nodular fat necrosis in pancreatitis. Occasionally inflammation occurs in other organs of the body to cause heart, lung, kidney, liver or spleen problems.

Khan et al in 1996\(^5\) described a white woman with fever and recurrent episodes of painful nodules of the lower extremities and the excisional biopsy confirmed panniculitis.

Proptosis as the manifesting sign of Weber-Christian disease was described by Cook et al.\(^4\) Orbital inflammation associated with cutaneous nodules, myalgia, nausea and malaise may be present.

An immense proliferation of benign cytophagic histiocytes can be observed in the bone narrow, spleen, lymph nodes and in the fatty and interstitial tissues of the visceral organs. A 49 years woman died in a septic shock, consequent to the pancytopenia and clotting disorder due to Weber-Christian panniculitis and this association with systemic histiocytosis was described by Steininger and Missmahl in 1988.\(^5\)

A review of 30 cases with Weber-Christian panniculitis was done by White and Winkelmann.\(^6\) Fat necrosis is also found in following diseases like pancreatic disease, alpha-1 anti-trypsin panniculitis, lupus and connective tissue disease panniculitis, lipomembranous panniculitis, lipoatrophy and cytophagic panniculitis.

In our case, although a clinical diagnosis of erythema nodosum was made, tissue diagnosis revealed typical features like infiltration of neutrophils around the fat cells and perivasularly and large giant cells ingested with fat and foam cells. This clinched the diagnosis of Weber-Christian disease and the patient showed response to corticosteroids. The case is being reported for its rarity and its association with type 2 diabetes.

References