Panniculitis-polyarthritis-pancreatitis syndrome

Sir,

Pancreatic panniculitis is a rare variant of panniculitis characterized by subcutaneous fat necrosis that affects 0.3-3% of patients across a range of different pancreatic disorders. Skin lesions are the presenting feature in about 40% of pancreatic panniculitis and precede the abdominal symptoms by 1-7 months.[1] Clinically, they present with painful, tender, ill-defined, erythematous to violaceous nodules that may undergo spontaneous ulceration and discharge of an oily brown, viscous material resulting from liquefactive necrosis of adipocytes. These lesions usually involve the lower extremities although they may also appear on the buttocks, trunk, arms and scalp. In addition to the skin, fat necrosis may involve peri-articular, abdominal and intramedullary adipose tissue. The

triad of pancreatitis, panniculitis and polyarthritis is known as pancreatitis, panniculitis and polyarthritis (PPP)-syndrome.[2] It is a rare syndrome with high morbidity and mortality so a high index of suspicion is required to diagnose it and initiate timely and appropriate treatment.

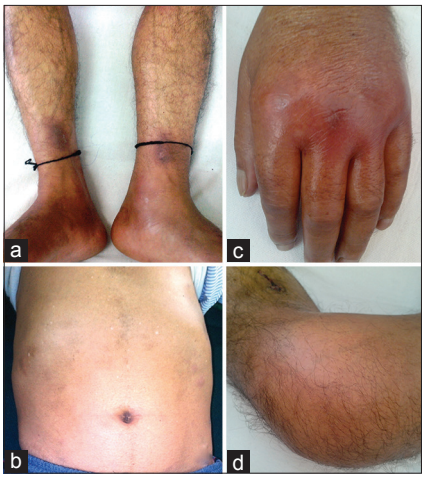

Figure 1: Multiple tender subcutaneous nodules over the lower legs and abdomen (a and b) and swelling of the joints of hand and knee (c and d)

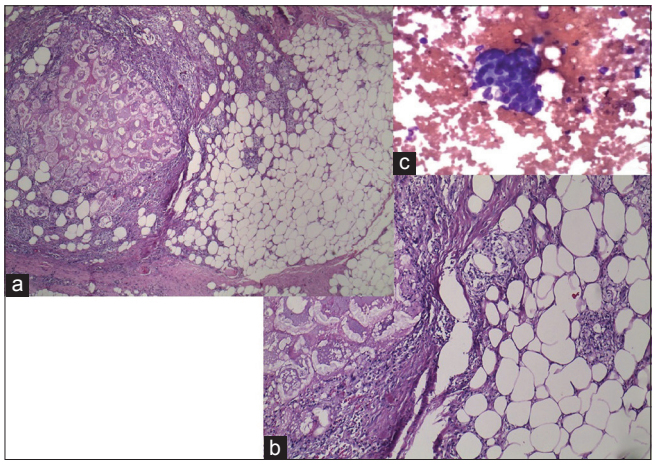

Figure 2: (a) Lobular coagulative panniculitis involving several fat lobules (H and E, ×10) and (b) fat lobules are surrounded by inflammatory infiltrate of lipophages, lymphocytes and eosinophils with thickening of fat septa (H and E, ×40). (c) Atypical cells on cytology (×10)

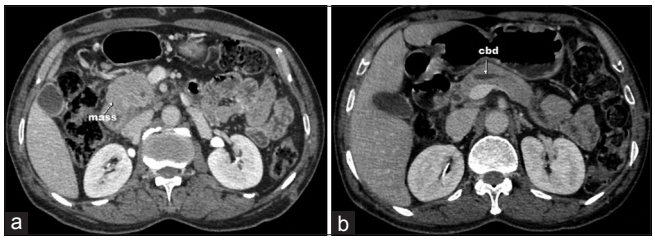

Figure 3: Computed tomography scan showing (a) mass in the head of pancreas and (b) common bile duct

A 57-year-old man with a 20-year history of heavy alcohol abuse presented with painful subcutaneous nodules on shins, buttocks, abdomen and shoulder accompanied by pain and swelling of both knees, ankles and elbows for the last two weeks. One month back, he had an episode of abdominal pain, nausea and vomiting. Records revealed that he was then diagnosed with acute pancreatitis and improved with symptomatic treatment. Physical examination revealed multiple tender subcutaneous nodules over the ankles, shins, thighs, abdomen and shoulders [Figure 1a and b]. There was no ulceration and oozing from the lesions. He had pallor and swelling and restricted mobility of the involved joints [Figure 1c and d]. The rest of the systemic examination was normal.

Laboratory investigations revealed that the erythrocyte sedimentation rate was raised, C-reactive protein was positive by latex agglutination method, gamma-glutamyl transpeptidase was raised three times the normal level, serum lipase was twice the normal and serum amylase was grossly raised to about five times normal. Screening for hepatitis B and C was negative and the tumor marker, carcinoembryonic antigen was within the normal range. Biopsy of the skin nodule showed lobular panniculitis and intense necrosis of adipocytes surrounded by an inflammatory infiltrate of lymphocytes and eosinophils, features characteristic of pancreatic panniculitis [Figure 2a and b]. Ultrasonography guided fine-needle aspiration cytology revealed atypical cells [Figure 2c]. Joint X-rays were normal. The abdominal computed tomography scan showed an ill-defined hypodense mass, approximately 2.2 × 2.7 cm with a craniocaudal extent of 2.6 cm in the head of pancreas with the pancreatic duct dilated to 4 mm throughout its course [Figure 3a and b]. There were also multiple soft tissue nodules in the subcutaneous plane. These findings pointed towards the possibilities of chronic pancreatitis or adenocarcinoma of pancreas. With confirmation of panniculitis on histopathology and investigations confirming pancreatic disease and associated joint involvement, a diagnosis of PPP-syndrome was made and the patient was referred to the department of surgery for further management. A triple bypass surgery for chronic pancreatitis was performed. There was good improvement in skin lesions and joint symptoms.

In 80-100% of cases of pancreatic panniculitis, the underlying condition is pancreatitis or pancreatic carcinoma, most commonly pancreatic

islet cell tumors, ductal adenocarcinoma and acinar-cell carcinoma.[1] Other causes include pancreatic pseudocyst, post-traumatic pancreatitis, pancreatic divisum or hemophogocytic syndrome. Pancreatic lipase and amylase are known to have a pathogenic role causing subcutaneous fat necrosis leading to panniculitis.[3] Ghost-like anucleated cells with shadowy walls are pathognomonic findings of pancreatic panniculitis on skin biopsy.[4]

PPP-syndrome is a rare condition with about 25 well-documented cases reported in the literature so far.[2] Early recognition of this triad is critical due to the high mortality from pancreatic disease when the diagnosis is delayed. Appearance of panniculitis in such cases and its detection can be life-saving.[5]

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