http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v8i12.18

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Eruptive Xanthoma with Acute Pancreatitis: A Case Report

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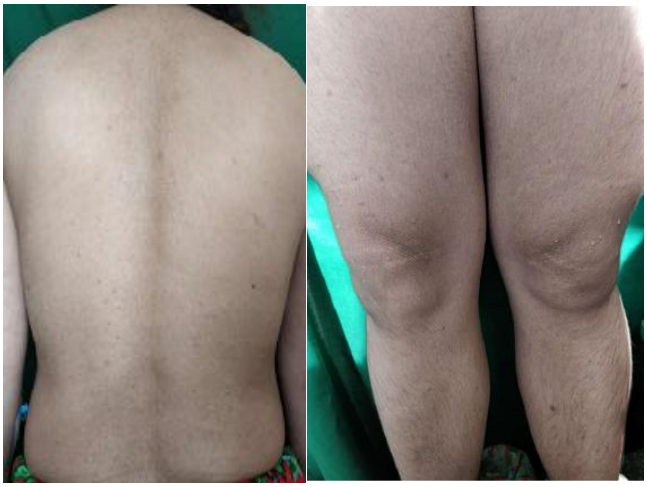
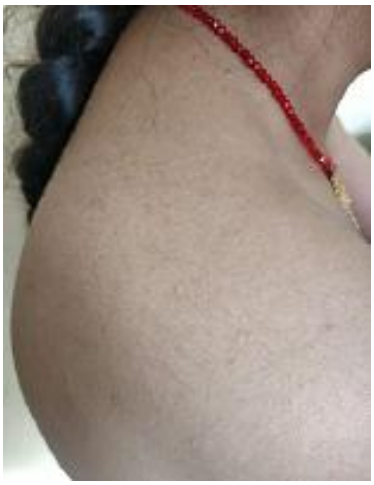
Introduction

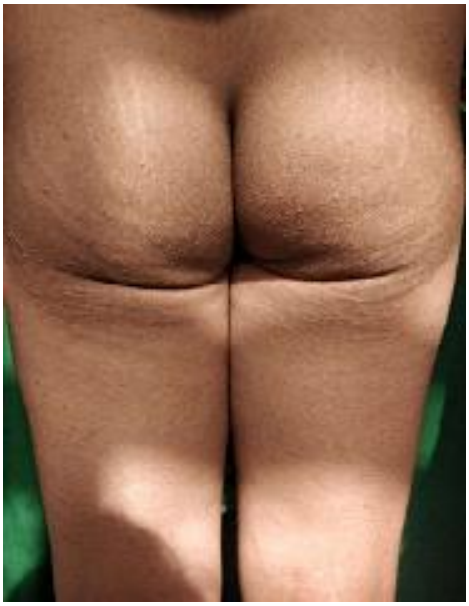
Xanthomas are benign lesions caused by localized lipid deposits in the skin, tendons and subcutaneous tissue. Clinically they are yellowish papules, nodules or plaques. They can present as early manifestations of some systemic disorders such as coronary artery disease, cerebrovascular accidents, hepatic steatosis and acute pancreatitis. The major forms of xanthomas associated with hyperlipidemia are eruptive, tuberous, tendinous and plain xanthomas. Early recognition and treatment of the underlying condition associated with xanthomas can decrease the morbidity and mortality.[2] Here we report a case of admitted patient of eruptive xanthomas with acute pancreatitis.

Case Presentation

A 24-year-old female patient presented to the outpatient department of surgery with sudden onset epigastric pain of 2 days duration which was radiating to back and associated with vomiting. On examination, she was found to have multiple scattered yellow papules over the upper and lower limb mainly over the extensor surfaces, buttocks and back. Historically, lesions were 10-15 days old and clinically asymptomatic. Ultrasound abdomen revealed bulky pancreas with heterogenous echogenicity and minimum peripancreatic fluid. Serum amylase and lipase levels were elevated to the level of 286u/l and 457u/l respectively. Diagnosis of mild acute pancreatits was made as there was no organ failure on laboratory investigations. There was no history alcohol and intake of any drug. All other possible causes of acute pancreatitis were sought but showed no relation. Her fasting lipid profile showed cholesterol levels of 247mg/dl and serum triglycerides levels were ….. Biopsy from the lesion revealed dense, diffuse infiltrate of sheets of foamy histiocytes without any accompanying inflammatory infiltrate in the dermis. The patient was started on fenofibrate 160mg and managed for mild pancreatitis as per protocol.







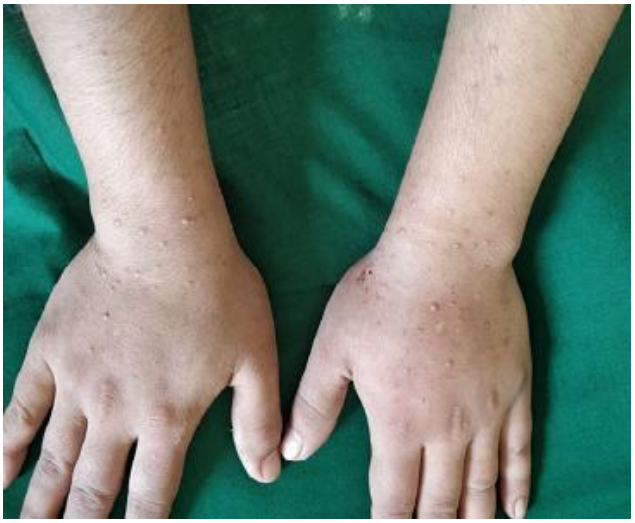


Figure 1-5: Showing yellowish papules over the back, lower limbs, buttocks and hands.

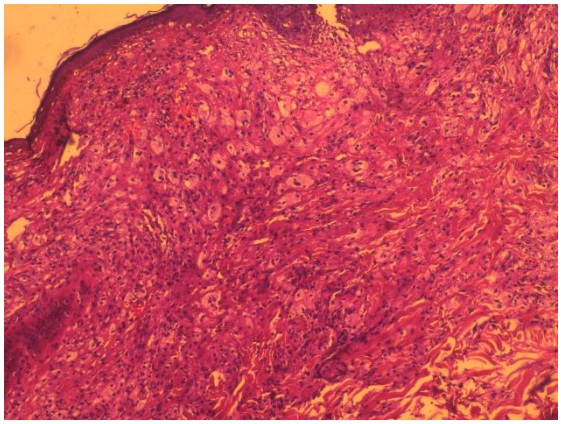


Figure 6: histopathology showing foamy macrophages in the dermis (10x)

Discussion

Xanthomas are fatty deposits in the skin. Five different forms have been described, including eruptive xanthomas, tuberous/ tendinous xanthoma, flat xanthomas, verrucous xanthomas, and xanthelasmas.[1]

Eruptive xanthomas present as crops of pinkish yellow shiny firm papules ranging from about 1 to 5mm in diameter. They appear usually on the extensor surface of the extremities, buttocks, back and hands.[1,2,3] The lesions are usually asymptomatic however, variable amount of itching and pain has been described.[1,2]In addition, Koebner phenomenon has also been described in some cases in the literature.[1]

The disease is a typical manifestation of extreme hypertriglyceridemia. Eruptive xanthomas have been retrospectively reported in 10% of patients

with severe hypertriglyceridemia.[2] Triglyceride levels in patients with eruptive xanthomas are often over 80mmol/L and can be seen in hypertriglyceridaemia type 1 (chylomicron increased), type 4 (increased very low density lipoprotein (VLDL)) and type 5 according to the Frederickson hyperlipidaemia classification.[1,4] The increased levels of triglyceride levels can be due to inability to clear circulating lipids or due to liver overproduction of triglyceride-rich lipoproteins. Increased hepatic production can be either familial or secondary to obesity, diabetes, alcohol or estrogen administration. Decreased lipolysis of VLDL and chylomicrons can also be genetic or acquired as in hypothyroidism, beta blockade or diabetes mellitus. Secondary hypertriglyceridemia can also be seen with end-stage renal disease and HIV infection.[1,2] Treatment with several medications (isotretinoin, sodium valproate, protease inhibitors, sertraline, thiazide diuretics, cyclosporine and tacrolimus) may also result in hypertriglyceridemia. Eruptive xanthomas have also been described in subjects with normal serum cholesterol and triglyceride levels as occurring in xanthomas consisting of phytosterols as well as in the cases of endemic injuries.[3]

Diagnosis is usually clinical and skin biopsy is indicated for confirmation. Characteristic histological findings of eruptive xanthomas are foamy macrophages containing lipids in their cytoplasm. Extracellular lipid in the dermis is found.

The main differential diagnosis is Xanthoma disseminatum which is a rare disease consisting of numerous, small, reddish-brown papules that are evenly spread over the face, skin-folds, trunk, and proximal extremities. Other differentials includes Langerhans cell histiocytosis, disseminated granuloma annulare, non-Langerhans cell histiocytosis, generalized eruptive histiocytoma and juvenile xanthogranuloma.[1,2]

The therapy always refers to the primary disease. Adequate treatment involves controlling the underlying hyperlipidemia with strict dietary therapy and HMG-CoA reductase inhibitors. Weight reduction and dietary carbohydrate restriction are helpful in cases associated with insulin resistance. Gradual resolution of cutaneous lesions is typically observed after normalization of serum lipids.[2] Lesions not resolving after medical management may require surgery, laser, or cryosurgery.[3]

Hypertriglyceridemia is a rare cause of acute pancreatitis. Its pathogenic mechanism is yet unknown, although it has been suggested to be due to the toxic effect of free fatty acids and of lysolecithin on cell membranes. These free fatty acids would be generated inside the pancreas due to the effect of pancreatic lipase on triglycerides.[4] In our case, the onset of acute pancreatitis could be due to the increased serum lipid levels.

To conclude, the appearance of eruptive xanthomas can herald the onset of serious complications due to severe hypertriglyceridemia. Prompt recognition of eruptive xanthomas and knowledge of its association with hypertriglyceridemia can help to decrease the time between a patient presentation to the physician and initiation of treatment for a serious medical condition.

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