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Pachydermoperiostosis and Menetrier's Disease in a Young Male

KEYWORDS: pachydermoperiostosis, menetrier's disease, hypertrophic osteoarthropathy

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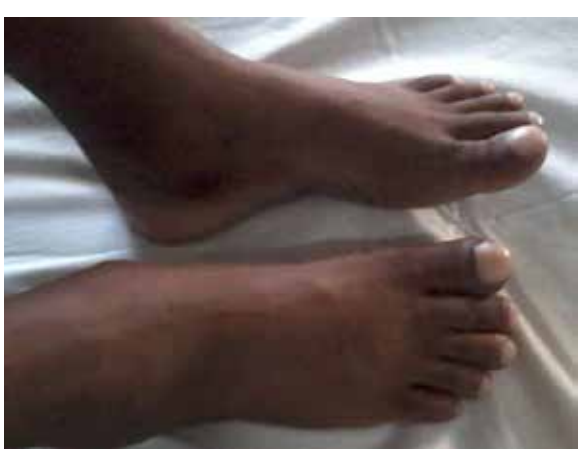
ABSTRACT *Primary pachydermoperiostosis is a rare inherited disorder that manifests clinically by digital clubbing, extremely enlarged, painful and swollen joints, hypertrophic skin changes and periosteal new bone formation. The pathological mechanisms are still debated and proposed etiological factors include genetic influences, anomalies in fibroblast acitivity or alteration in peripheral blood flow. Menetrier's disease is a form of hypertrophic gastropathy occurring primarily in middle aged males. It is characterized by thickening of the gastric mucosa due to hyperplasia of gastric glands. Clinically, they present with dyspepsia, upper abdominal discomfort, loss of appetite, anemia, hypoproteinemia and its associated features, apart from weight loss. While the etiology of Menetrier's diseas is still unknown, association of cytomegalovirus and Helicobactor pylori infection of gastric mucosa has been reported. [1],[2] Some consider it as a precancerous condition.[2] Here we report a case of primary pachydermoperiostosis with associated Menetrier's disease.*

CASE HISTORY

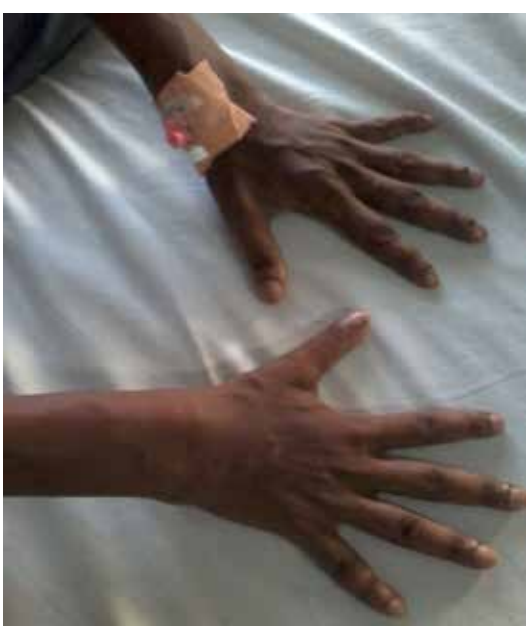
A 26year old male came to our general medicine OPD with chief complaints of loose stools for 15 days which was watery in consistency, not blood stained. History of painful, swelling of joints in both hands and legs for 6 months. On general examination, skin of scalp and cheeks were thrown into folds with thickening of eyelids giving a leonine facies appearance.The skin was also coarse and thickened over the hands and feet. Spade like enlargement of hands and feet was present.

FIGURE 1 : SPADE LIKE ENLARGEMENT OF FEET



Pan digital clubbing was present in both hands.

FIGURE 2 : PAN DIGITAL CLUBBING OF FINGERS



There was no macroglossia. Examination of the other systems were normal. Patient's vital data were with in normal limits. ESR was elevated moderately. Patient's routine urine examination, serum proteins, routine blood biochemistry, serum T3, T4, and TSH and ECG were with in normal limits. Elisa for HIV & II was negative. His other lab parameters were normal. Barium meal follow through revealed hypertrophied gastric rugal folds with rapid transit time.

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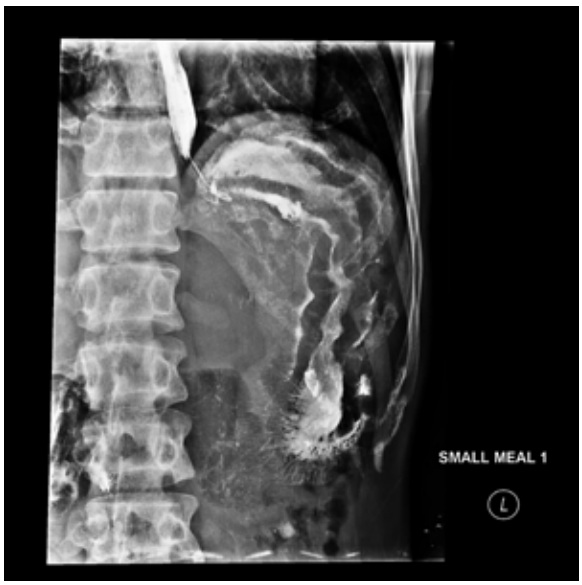
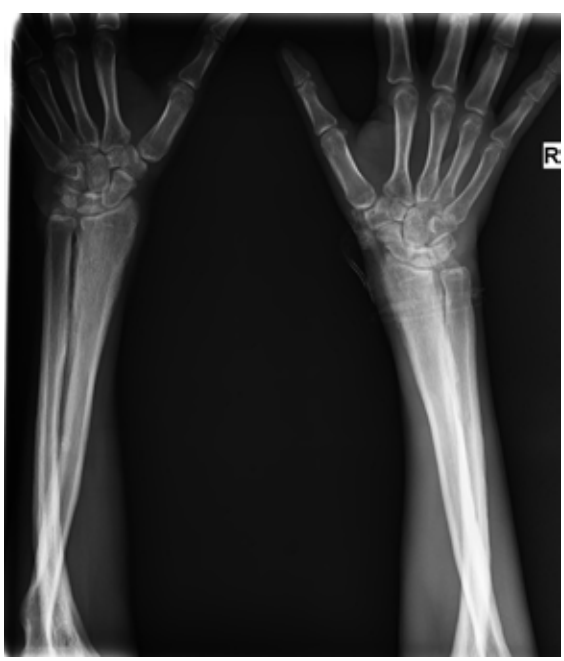
FIGURE 3 : BARIUM MEAL SHOWING HYPERTROPHIED GASTRIC RUGAL FOLDS



X-ray forearm showed cortical breach with periosteal reaction in the medial aspect of ulnar bone.

FIGURE 4 : XRAY FOREARM SHOWING CORTICAL BREACH WITH PERIOSTEAL REACTION OF ULNAR BONE



DISCUSSION

Pachydermoperiostosis(PDP) or Primary hypertrophic osteoarthropathy is a rare genetic disorder that affects both bones and skin characterized by periostosis, arthralgia and pachyderma and finger clubbing. Dermatological features usually found are coarse facial features, cutis vertices gyrata, hyperhydrosis of palms and soles, hyperplasia of sebaceous glands and seborrhoea. Associated features described are carpal and tarsal tunnel syndromes, chronic leg ulcerations, large calcifications of Achilles tendon, gynecomastia, opthalmological abnormalities, Crohn's disease and mental retardation.[3]

The pathophysiology of this condition is not clear. Alteration of fibroblast biosynthetic activity and increased sensitivity of sex hormone steroid receptor and epidermal growth factor receptor system in the affected tissues were reported.[4]Usual age of onset of this condition is after puberty, though occasional cases are reported in children. Skin and bone changes usually progress for the first 5 to 10 years, although may continue to progress indefinitely.

Menetrier's disease is a rare entity characterized by large tortuous gastric mucosal folds. The etiology of this condition is unknown. Menetrier's disease was first described in 1880, but in spite of more than 300 reported cases in the literature, the disease is almost as mysterious now as when it was first described. The existing data suggests that Menetrier's disease might represent a common response to a number of endogenous and exogenous stimuli. The onset is insidious with peak incidence in fourth to sixth decade and the common clinical features associated are epigastric pain, vomiting, weight loss and peripheral edema. Most striking abnormality observed during endoscopy is the thick, tortuous gastric rugae which do not flatten with luminal air with occasional gastrict erosions and ulcerations. Menetrier's disease has been previously reported in a family in association with primary pachydermoperiostosis.[3]

Defects in the synthesis and degradation of regulatory macro molecules were observed in both these conditions. Abnormalities in the production of transforming growth factors (TGF) have especially been reported. Increased TGF - alpha immunoreactivity has been shown in gastric mucosa of patients of Menetrier's disease,[5] whereas TGF-beta was observed to stimulate fibroblasts in patients of pachydermoperiostosis,[6] apart from the altered processing of plasminogen activator inhibitor I (PAI-I).

Not all patients of pachydermoperiostosis have gastrointestinal symptoms, hence are not investigated for its involvement. As both pachydermoperiostosis and Menetrier's disease are hypertrophic conditions of the skin and gastric mucous membranes respectively and have at least some similarities in the abnormalities of growth factors, their association could be more frequent than reported.

This case is presented for its rare features. So far there has been only one case reported in India.[7]

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