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Idiopathic Hypertrophic Osteoarthropathy

KEYWORDS: Idiopathic hypertrophic osteoarthropathy, Clubbing

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ABSTRACT *IDIOPATHIC HYPERTROPHIC OSTEOARTHROPATHY also known as IDIOPATHIC PACHYDERMOPERIOSTITIS is a rare syndrome. It is characterized by the presence of periosteal inflammation and new bone formation, arthritis and clubbing of digits. Thickening of skin of the hands, forearm and legs as well as marked accentuation of facial folds is typical. This is a rare condition often mimicking some other well known conditions like acromegaly and the leonine fancies seen in leprosy. This case is unusual because of the lack of family history and the late age of presentation. A case of idiopathic pachydermoperiostitis of unknown etiology is rare, is reported.*

CASE HISTORY

A 35 year old male patient, who is a non smoker and non alcoholic, came with complaints of multiple episodes of loose stools, watery in nature and continuous in nature for four days. History of fever which is intermittent and low grade was present for one day. History of dyspnoea on exertion was present which is relieved by rest since four years. Grade 2 type of breathlessness. Progressive enlargement of both hands and foot, accompanied by excessive sweating of face and extremities along with accentuation of facial folds and thickening of skins of the hands, forearm and legs were present, and grade four clubbing of all fingers and toes [Figure1-4] which started at the age of 15 years and was progressive in nature was found. No related family history or any other co-morbid illness in the past.

On examination patient had severe pallor and grade four clubbing along with severe dehydration. Routine investigations revealed low hemoglobin, low PCV and MCH level. RFT and LFT were within normal limits. Peripheral smear done showed Microcytic Hypo chromic, Pencil cell RBC'S. So Serum iron and TIBC was sent which revealed low serum iron level and normal iron binding capacity.

X-ray of both hands and foot done revealed symmetrical subperiosteal new bone formation with acroosteolysis of phalanges of both hand and foot [Figure 5-7]. UGI Scopy done showed Gastric Mucosal Prolapse and D1-D2 junction deformity with luminal narrowing in the duodenum. 2D-Echo done revealed a normal study.

1. accentuation of facial folds and thickening of skins of the hands, forearm and legs

2. thickening of skins of the hands, forearm

3. grade four clubbing of all fingers

4. grade four clubbing of all toes

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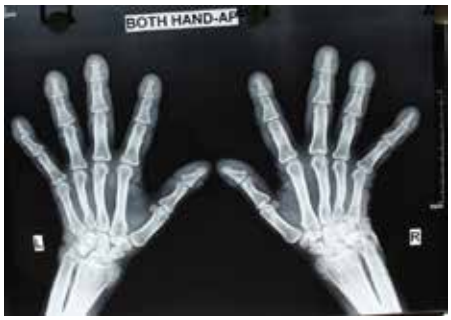
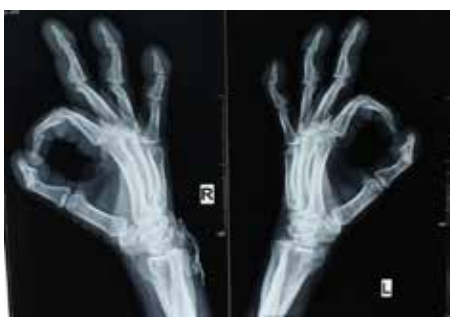
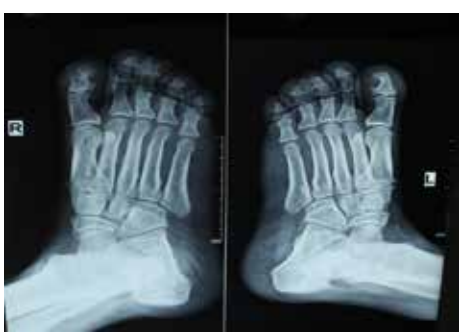


5. new bone formation with acroosteolysis of phalanges of both hand



6. new bone formation with acroosteolysis of phalanges of both hand



7. new bone formation with acroosteolysis of phalanges of both foot.

Discussion

Hypertrophic osteoarthropathy is divided into primary and secondary forms. Pachydermoperiostosis is the primary form which accounts for 5% of all cases of hypertrophic osteoarthropathy. Secondary hypertrophic osteoarthropathy is associated with underlying cardiopulmonary diseases and malignancies hence it is also called as pulmonary hypertrophic osteoarthropathy.[1]

Pachydermoperiostosis is a rare hereditary disorder. It was first described in 1868 by Friedreich.[2] Characteristic features of PDP are periostosis which is swelling of periarticular tissue and subperiosteal new bone formation, digital clubbing and pachydermia which is thickening of the facial skin and scalp. Pain, polyarthritis, cutis verticis gyrata, seborrhea, eyelid ptosis and hyperhidrosis are the characteristic features. [3,4]. PDP usually manifests in adolescence. It occurs exclusively in males and M: F ratio is 7:1. [9-10] PDP may remain undiagnosed, it may progress until there are significant deformities in face, joint and digits, until the patient seek medical attention.

Touraine solente and Gole individualized the three forms of pachydermoperiostosis in 1935 [5-7].

- The complete form which involves 40% of cases with pachydermia and periostitic changes. [1,5,6,7,8]
- The incomplete form lacking pachydermia but involving the bone changes occurring in 54% 0f cases. [1,5,6,7,8]
- A forme frusta which occurs in only 6% of cases, characterized by minimal or no bony involvement but has prominent pachydermia. [1,5,6,7,8]

The exact cause of these pathological differences is not known. *[1,5,6,7,8]*

CAUSES

PRIMARY	***Hereditary*** *[10,12,13,30]*	***Eg: X-linked disease autosomal dominance (IL)-6 and RANKL***
SECONDARY	cardiopulmonary diseases[21,29]	***Eg: bronchiectasis congenital heart diseases cystic fibrosis tuberculosis***
	hepatic diseases	Eg: Portal cirrhosis biliary cirrhosis
	gastrointestinal diseases [10,25-28]	Eg: inflammatory bowel disease polyposis Crohn disease peptic ulcer chronic gastritis Ménétrièr disease protein-losing enteropathy gastric carcinoma
	5.connective tissue and bony disorders[14-23]	Eg: Myelofibrosis Spondylolisthesis of the L5-S1 vertebrae Periodontal and alveolar bone abnormalities Osteoporosis Rheumatoid arthritis Ankylosing spondylitis Psoriatic onychopathy
	6.Nervous system disorders[19,24]	Eg: Compressive neuropathy Gynecomastia Atherothrombotic brain infarction
	7.Malignancies[11,28]	Eg: Hodgkin's disease carcinoma nasopharynx chronic myeloid leukemia En coup de sabre (a form of localized scleroderma)
	8.Others[7]	Eg: Hypoplastic internal genitalia inguinal hernia and peptic ulcer disease (rare association)

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[HOWEVER THE EXACT ETIOLOGY IN THIS CASE CANNOT BE FOUND OUT DUE TO LOGISTIC REASONS OF THE PATIENT. AND CONSIDERING THE HISTORY AND LAB INVESTIGATIONS THE POSSIBLE DIAGNOSIS OF IDIOPATHIC PACHYDERMOPERIOSTITIS IS MADE]

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