AN INTERESTING CASE OF GRADE FOUR CLUBBING

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Abstract:
Hypertrophic Osteoarthropathy is a syndrome characterised by abnormal proliferation of the skin and osseous tissue at the distal parts of extremities. Hypertrophic Osteoarthropathy is either primary or most often secondary due to neoplastic or infectious cause. Here we report an interesting case of Primary Hypertrophic Osteoarthropathy with hypertrophic gastritis and hypoproliferative anemia.

Keyword: PRIMARY HYPERTROPHIC OSTEOARTHROPATHY, PACHYDERMOPERIOSTITIS, GRADE 4 CLUBBING

INVESTIGATIONS: Complete blood count: Hemoglobin-8 gm/dl, Total count 6000/cu.mm. Packed cell volume: 30% ESR 40/80mm/hr. Platelet count – 50,000cells/cu.mm. Renal function test and Liver function test were normal. Peripheral smear study showed normocytic normochromic anaemia with decreased platelets. Reticulocyte count -1%. Chest x-ray, ECG and Ultrasound abdomen were normal. Xray forearm and legs showed predominant sclerosis of cortex and medulla of entire long bones. Upper gastrointestinal endoscopy showed pan-gastric mucosal inflammation, edematous hypertrophic giant gastric folds and cobblestone appearance of gastric mucosa with erosions in the fundus and pyloric antrum. Gastric Biopsy findings were consistent with menetriers disease. Bone marrow biopsy revealed hypocellular marrow.

TREATMENT: Patient was managed conservatively for primary disease. Proton pump inhibitor was given. Analgesics were administered for pain control.

DISCUSSION: PRIMARY HYPERTROPHIC OSTEOARTHROPATHY:
Hypertrophic Osteoarthropathy (HOA) is a syndrome of chronic periostitis of bones, clubbing, and synovitis. Primary form is also called as Pachydermoperiostitis or Touraine –Solente -Gole syndrome. This disorder is commonly inherited as autosomal dominant trait and common in boys. Primary HOA is characterized by the insidious development of clubbing with “spade-like” enlargement of hands and feet. Complaints include cosmetic unsightliness and decreasing dexterity or awkwardness in using the fingers. There may be vague bone pain, joint pain and swelling which may be exacerbated by alcohol.
There will be prominent clubbing with cylindrical thickening of Forearms and legs—these often correspond with radiographic periosteal thickening. Recurrent, mildly symptomatic joint effusions may be accompanied by acro-osteolysis particularly with resorption of distal phalanges of hands and feet. Facial skin may be thickened and furrowed with deep nasolabial folds and a corrugated scalp, known as "leonine" facies. The skin of the face, scalp, hands, and feet may show excessive sweating and a "greasy" feel. Other features include gynecomastia, female hair distribution, striae, acne vulgaris, and cranial suture defects. The skin overlying the scalp becomes very thick and corrugated commonly referred to as cutis verticis gyrata. Associated feature of hypertrophic osteoarthropathy include bone marrow failure and hypertrophic gastropathy. The genetic abnormality in Primary Hypertrophic Osteoarthropathy involves a mutation in the hpgd gene that encodes 15- hydroxyprostaglandin dehydrogenase, which is primary enzyme involved in prostaglandin degradation. No definitive treatment is available for the disease so far. Only anecdotal success with tamoxifen is reported. Analgesics are used for pain control. Our case of Primary Hypertrophic Osteo-arthropathy had many manifestation of the disease including cutis verticis gyrata, hypertrophic gastropathy, hypoproliferative anemia which satisfy the criteria for the diagnosis. In all cases of grade four clubbing we need to rule out systemic cause of clubbing particularly the underlying malignancy.

REFERENCES:
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