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Case Report
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A Case of Pachydermoperiostosis

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Case Report

The Tourain-Solente-Gole syndrome or pachydermoperiostosisis a rare genetic form of primitive hypertrophic osteoarthritis; It is characterized by pachyderma (thickened facial skin), skeletal changes (periostosis), excessive sweating (hyperhydrosis) and acropachia (digital clubbing).

There is a complete form with pachydermia, clubbing and periostosis, the incomplete form with isolatedperiostosis and limited skin changes and a whips form with predominant pachydermia and minimal periostosis [1].

Adolescent males are predominantly affected with male-to-female ratio of approximately 7:1. Typically, men are affected more severely than women. It begins during childhood or adolescence and progresses gradually over the next 5–20 years before stabilizing. Life expectancy may be normal [2].

Mutations at hydroxy-prostaglandin dehydrogenase enzyme which is responsible for PGE2 degradation or at SLCO2A1 which is PGE2 transporter result in increase of serum levels of PGE2 that stimulate various intracellular signaling pathways including vascular endothelial growth factor expression (VEGF). The latter stimulates various cellular components of fibrous and bony tissues such as osteoblast, osteoclast, and fibroblast that lead to various bony and dermatological manifestations of hypertrophic ossteoarthropathy [3,4].

Diagnosis of pachydermoperiostosisis made clinically based on the combination of digital clubbing and periostosis of tubular bones. We report the case of a male patient of 49 years old who came to our outpatient clinic, for pain at the tibial tarsal joints and mild fever. The patient's history begins at the age of 15 with the appearance of ankle pains with functional impotence. In the suspect of rheumatic fever, the patient was treated with penicillin G 1200.000 U for about 6 months without any benefit. In the next months worsening of painful articular symptoms.

In later months, onset of joint effusion of the knees for which arthrocentesis occurred and pharmacological treatment with piroxicam 20 mg/day had been necessary for long months.

Atage 30, he noted the appearance of major paresthesia in the hands that over time had increased in volume with the appearance of skin patches on the fingers probably related to the existence of severe obesity.

For two years he referred accentuation of numbness in the fingers and reported recurrent episodes of sciatica with good improvement after use of ibuprofen 600 mg.

The physical examination documented a constitution characterized by brachitipo, thick neck,

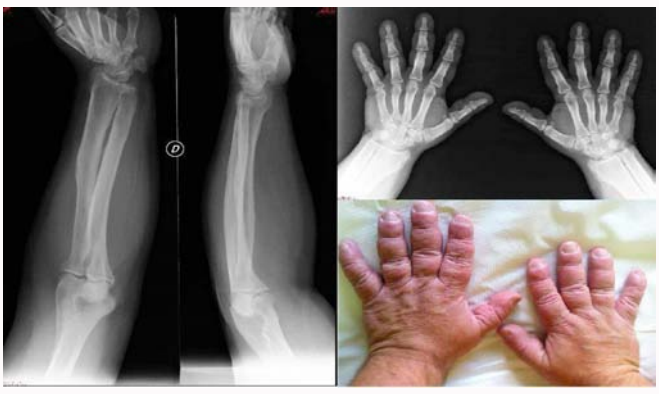

Figure 1: The radiographs of the hands showing periostosis at the distal ulna and radius.

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severe obesity Height 152 cm; Weight 92,800 kg; BMI 40.1; Altered symmetry between the forearm and arms with the presence of swelling along the ulnar face of the forearm on both sides, pachydermia on the upper and lower limbs, hyperhidrosis; Joint sex amination were characterized by swelling and pain in the ankles and on the sacral lumbar spine and cervical column but without no functional impairment.

At labs investigation, inflammatory markers, rheumatoid factor and citrullinated antibodies were negative, hemocrome were also normal. The radiographs of the hands showed periostosis at the distal ulna and radius (Figure1). Based on radiographic findings, clinical and laboratory data adiagnosis of Pachydermoperiostosis was made. This rare disease must be considered and placed in differential diagnosis with secondary forms of secondary osteoarthritis and with chronic rheumatic diseases.

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