Case Report

Hypertrophic Osteoarthropathy With Acro-osteolysis in a Patient With Primary Pulmonary Hypertension

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A B S T R A C T

Hypertrophic osteoarthropathy is an entity characterized by a triad of periostitis of long bones, clubbing and arthritis. Radiologically there are two patterns, one characterized by new bone formation which predominates in patients with pulmonary disease, and another by acro-osteolysis that is most frequently associated with congenital heart disease. We report the case of a 30-year-old man diagnosed with primary pulmonary hypertension for two years, developing hypertrophic osteoarthropathy with a mixed radiological pattern.

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Osteoartropatía hipertrófica con acro-osteolisis y neoformación ósea en un paciente con hipertensión pulmonar primaria

R E S U M E N

La osteoartropatía hipertrófica es una entidad caracterizada por la tríada de periostitis de huesos largos, acropaquias y artritis. Radiológicamente se distinguen 2 patrones; uno caracterizado por neoformación ósea que predomina en pacientes con patología pulmonar, y otro por acro-osteolysis que se asocia más frecuentemente con cardiopatías congénitas. Presentamos el caso de un varón de 30 años diagnosticado de hipertensión arterial pulmonar primaria desde los 2 años, que desarrolló una osteoartropatía hipertrófica con un patrón radiológico mixto.

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Introduction

Hypertrophic osteopathy (HOA) is a disease characterized by chronic proliferative periostitis of long bones, acropaquies and arthritis. It can be divided according to its etiology in primary HOA, with no apparent underlying cause, familial aggregation and chronic progression; and secondary, associated with lung, cardiac, hepatic or intestinal diseases, with a tendency to bilateralism, symmetric and rapidly progressive. There are two distinct radiological patterns of HOA, one characterized by hypertrophy or bone neoformation, predominant in patients with lung disease (pneumic HOA) and which has its onset after puberty; and another with acroosteolysis, associated frequently with cyanotic congenital heart disease and which has its onset during childhood. Two variants have been seen in this last pattern, one in which the reabsorption of the distal phalanges makes them adopt a pyramidal form and another in which its massive destruction leads to the formation of flat surfaces. We present the case of a patient with primary pulmonary hypertension who developed HOA with a mixed radiological pattern.

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Fig. 1. Hand X-ray (A) in which there is a periosteal reaction on the radius. There is distal widening with hypertrophic changes and areas of osteolysis, with a good example on the right fourth finger. Juxtaarticular demineralization. Feet X-ray (B) observing marked destruction of the distal phalanges, some with flattened morphology and distal hypertrophy with bone proliferation. There is marked deformity of the fingers.

Fig. 2. Comparative image of the distal phalange of the hand (A) and foot (B); the first has proliferative changes and the second destructive changes with flattened morphology.
Clinical Presentation

The case is a 30-year-old male patient with a history of primary pulmonary hypertension without a shunt and followed by pediatric cardiology ever since. He had no other history of importance, including rheumatic disease or psoriasis. At age 29 he started presenting episodes of symmetric, additive polyarthritis which affected knees, ankles, shoulders, wrists and metacarpophalangeal joints, with important functional limitation. Physical examination found, in addition to generalized cyanosis and acropachy, no skin or nail lesions suggestive of psoriasis. Laboratory analysis found normal acute phase reactants, negative rheumatoid factor and positive low titer anti-CCP antibodies in a single determination. X-rays showedacroosteolysis of the distal phalanges of the hands and feet, severest in the latter, in which some of the phalanges adopted the form of a pyramid or had a flattened end, with important bone neoformation in both locations, especially the hands (Figs. 1 and 2). Both wrists observed, in addition to radiocarpal and intercarpal joint impingement, especially of the left side, no erosions. Long bones (tibia, peroneal, radial) had a cottony cortex and periosteal separation compatible with periostitis. A bone scintigraphy showed moderate uptake in the distal phalanges of both hands, as well as right wrist and radius with a theoretical zone in the left semilunar.

The patient was diagnosed with pneumic HOA and treated with low dose steroid (deflazacort 6 mg/day) and calcium and vitamin D supplements. He responded rapidly to treatment with remission of arthritis and steroids were suspended after a few months; he only presented a single relapse which responded after 2 weeks of treatment with no further relapses.

Discussion

This case involved a patient with underlying lung pathology of pediatric onset, who developed a late polyarthritis that led to the diagnosis of pneumic HOA. However, changes such as acroosteolysis, new bone formation and periostitis probably had a longer evolution, but because these events often have a more insidious course and patients are often asymptomatic, it is usually detected in an incidental radiological study.

Previous studies have reported that foot affection is more frequent and more severe than that of the hand, as in our patient. Solid and continuous periosteal reaction in the long bones is also a common finding, such as the one our patient presented on the tibial and radial bones.

The mixed pattern combining acroosteolysis and bone neoformation in the same patient was what called our attention, but because these events often have a more insidious course and patients are often asymptomatic, it is usually detected in an incidental radiological study.

The authors have no conflicts of interest to declare.

References


