



CLINICAL EVOLUTION OF PRIMARY HYPERTROPHIC OSTEOARTHROPATHY (PACHYDERMOPERIOSTOSIS): CASE REPORT

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BACKGROUND

Pachydermoperiostosis is a genetic syndrome characterized by the progressive increase of the thickness of the long bones, arthritis, skin thickening and digital clubbing.

CASE REPORT

A 15-years-old male patient, without comorbidities, sought clinical care in 2006 complaining of arthralgia and edema in the lower limbs with normal laboratory tests. Physical and radiological exams revealed proliferation of the cortical layer of the long bones, proliferation and thickening of the face and scalp, and joint effusion in the knees. Prednisone (5mg/day), Chloroquine Sulfate (150mg/day) and Alendronate sodium (70mg/week) were prescribed. In 2010, he returned with painful complaints on knees and wrists to efforts, drowsiness, in the use of the medications. It was prescribed Meloxicam (15mg/day) and started weaning of Prednisone (3mg/day). In 2011, he presented diarrhea and epigastric pain. Physical examination revealed seborrheic nodules in the frontotemporal region, joint effusion in the knees, and increased bone diameter in the distal third of the tibiae. Raloxifene (1 tablet/day for 2 months) and Omeprazole (20mg/day) were prescribed, Meloxicam was maintained and Alendronate and Prednisone were suspended. There was a referral to the Dermatologist who prescribed Isotretinoin. After 2 months, through improvement of the dyspeptic symptoms, he prescribed Risedronate (5mg/day) and Diacerein (50mg/day). Patient returned in January/2016, and three years ago he suspended Risedronate on his own, complaining of fatigue, palpitation and numbness in the feet. On physical examination, bradycardic, with sparse extrasystoles, skin without seborrhea or sebaceous cysts, with bone diameter stabilized in relation to the second evaluation. Risedronate (5mg/day) was reintroduced and the dose of Meloxicam was reduced (7.5mg/day). In April/2016, he reported a change in his own dose of Meloxicam (15mg/day) and Diacerein (100mg/day), due to knee arthritis, showing improvement. Echocardiogram and Exercise Test were normal. Pachydermoperiostosis was compensated for bone and cutaneous proliferation, with intermittent articular manifestations, in response to Meloxicam. Risedronate (35 mg/week) was prescribed and Meloxicam (15 mg/day) and Diacerein (50 mg/day) were maintained. Between 2017/2018, on physical examination, there were stabilized cutaneous and bone manifestations. On cardiac auscultation, isolated extrasystoles. Holter was performed with a Sporadic Ectopic Activity report, and Atenolol (25mg/day) was prescribed to control nocturnal tachycardia.

CONCLUSION

Although there was little evidence of adequate treatment, there were symptom compensation and good results with oral bisphosphonates. The studies are scarce and show empirical use of Pamidronate, unavailable in Brazil.