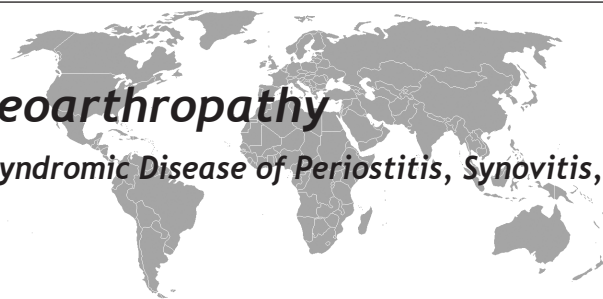


Idiopathic Hypertrophic Osteoarthropathy (Pachydermoperiostitis); A Syndromic Disease of Periostitis, Synovitis, Clubbing and Skin Furrowing

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İdiopatik Hipertrofik Osteoartropati (Pakidermoperiostoz)

ABSTRACT

Idiopathic Hypertrophic osteoarthropathy is a syndromic disease which is characterized by periostitis and thickening of long bones, clubbing of toes and fingers, synovitis of joints, hyperhydrosis, thickening and furrowing of skin. However, to say an idiopathic HOA, secondary causes of clubbing should be investigated and ruled out. The disease is usually self limiting and most patients enjoy healthy lives except moderate discomforts. We report a case of 24 years male patient diagnosed as idiopathic HOA who responded partially with low dose of steroid (prednisolone).

Key words: Hypertrophic osteoarthropathy, idiopathic, periostitis, synovitis.

INTRODUCTION

Hypertrophic osteoarthropathy (HOA) is a syndromic disorder characterized by periostitis of the long bones, clubbing of finger or toes, synovitis of the joints, and skin changes¹. HOA usually occurs secondary to the pulmonary neoplasms or other extra or intra-thoracic disorders which is called secondary HOA (2,3). In 3 to 5% of cases when no obvious causes are found, the condition is termed as idiopathic HOA (4). Idiopathic HOA occurs more commonly in adolescent and adults (Adulthood variety) and less commonly in children (childhood variety) (2).

ÖZET

İdiopatik hipertrofik osteoartropati periostit, uzun kemiklerin kalınlaşması, ayak ve parmaklarda çomaklaşma, eklemlerin sinoviti, hiperhidroz ve deride kalınlaşma/buruşma ile karakterize bir sendromik bir hastalıktır. Ancak bir idiyopatik HOA söylemek için çomaklaşma yapan diğer nedenleri dışlamak ve araştırmak gerekir. Hastalık genellikle kendi kendini sınırlar ve hastaların çoğu orta düzeyde rahatsızlıkları dışında sağlıklı bir hayat tadını çıkarır. Biz düşük doz steroid (prednisolon) ile kısmen cevap alınan idiyopatik HOA tanısı almış 24 yaşında bir erkek hasta olgusunu sunduk.

Anahtar kelimeler: Hipertrofik osteoartropati, idiyopatik, periostit, snovit

CASE

A 24 year male patient presented to our hospital with complains of gradual swelling of both ankle and wrist joints, thickening of both legs and fingers, coarsening of skin of bilateral hands, feet and face for past 5 years. There was no history of redness and stiffness of joint, hemoptysis, chest pain, purulent expectorant, and fever. No history of cyanosis and dyspnea in the childhood. There was no history of jaundice, pruritus, ascites, diarrhea, mucus or blood in stool, and no family history as well. Physical examination revealed swelling of bilateral wrist, ankle, knee joints, marked thickening of the bilateral legs, painless clubbing of both hand and feet, coarsening and oily appearance of skin, and profuse sweating of both hands. Investigations including complete blood count, blood sugar, rheumatoid factor, CRP, liver function test,

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renal function test, thyroid function test, electrolytes, calcium, phosphorus, autoantibody screen, chest xray and echocardiography were normal except ESR which was 34 mm/ first hour. X-ray of both legs showed the periosteal thickening of bilateral tibia and fibula (Figure 1). Diagnosis of idiopathic HOA was established after exclusion of secondary causes of HOA, acromegaly, rheumatoid arthritis, engleman's disease. Patient was prescribed oral prednisolone 20 mg once a day for 1 month followed by tapering dose for another one month. Two months after medication his signs and symptoms were relieved partially. melanoma metastasis) were excluded since their number was inadequate for statistical analysis.

DISCUSSION

When a patient presented with clubbing, various causes needs to be excluded like heart diseases (congenital cyanotic heart disease, acute bacterial endocarditis), lung diseases (bronchiectasis, lung abscess, cystic fibrosis), gastrointestinal and hepatobiliary disease (chronic inflammatory bowel disease, cirrhosis, chronic active hepatitis) and miscellaneous conditions (thyrotoxicosis, hodgkins lymphoma) (2). When no underlying causes are found the clinical condition with triad of periostitis, arthritis, and clubbing is diagnosed as idiopathic HOA (4). Most patients with idiopathic HOA do not show any symptoms until adolescence when they begin to present with clubbing, swelling of joint, and thickening of bones. These features progress for number of years and finally become static (1). Childhood variety of idiopathic HOA usually begins around the age of 1 year and is associated with delayed closure of sutures in skull, and presence of wormian bones (5). Bony changes of patients consist of symmetric and irregular periosteal hypertrophy with cylindrical thickening of usually long bones. Although these changes may involve any bones, skull and vertebral column are usually spared. Articular surface of joints are not involved, however there may be the moderately painful or asymptomatic intermittent swelling and effusion of joints (6). Xray of hands shows resorption of distal phalanges of hands and feet called as acro-osteolysis (7). Painless clubbing in these patients is due to proliferation of soft tissues of distal phalanx which gives bulbous appearance of the digits (2). Skin changes involve generalized thickening or pachyderma, seborrheic overactivity, hyperhidrosis, and features of eczema. Excessive thickening and redundancy of skin of scalp and forehead gives the coarse 'leonine

like' appearance or cutis verticis gyrate (1).

Most patients enjoy healthy and normal lives except moderate discomforts and cosmetic problem because of the disease. Although their main concern is related to the cosmesis and hyperhidrosis, serious problems sometimes may develop. Thickening of skin eyelids occasionally causes ptosis which requires surgical correction. There have been reported facial nerve palsy due to thickening of skull bones and extra-medullary hematopoiesis with bone marrow failures due to destruction of marrow by idiopathic HOA (8). In 40 to 50 percents of all reported cases, family history is present and the disease is more common in males and transmitted as an autosomal dominant trait with variable penetrance (2,6). Secondary or pulmonary HOA is differentiated from idiopathic type by less pronounced anatomic changes, more severe discomforts, simultaneous development of features along with the primary disease process, and absence of family history (1). The exact pathogenesis of the disease remains unknown, however, various mechanisms like growth factors, immune mechanism, vagal stimulation, platelet endothelial interactions have been attributed (4). The disease is usually asymptomatic but short course of prednisolone relieved the clinical signs and symptoms noticeably in our case.

The main problem in this type of disease is lack of diagnosis and patients are wandering to different physician, der-



Figure 1. X-ray of both legs; periosteal thickening

matologist, and orthopedic surgeon wasting the time and money. Even though the disease is rare, attending physicians should have the sound knowledge regarding the diagnosis of the disease and they should properly counsel the patients about the causes, courses, pathogenesis, future outcomes, and possible treatment modalities of the diseases.

In conclusion, idiopathic HOA is not so uncommon disease which should be diagnosed only after exclusion of secondary HOA in any patients presenting with features of clubbing, swelling of joints, coarsening of skin, periostitis and thickening of long bones in X-ray, and positive family history in some cases. Proper counseling regarding the prognosis of the disease is mandatory for the patient's satisfaction.

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