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FACIAL LOCALISATION OF PAGET'S DISEASE IN AN ADOLESCENT: A CASE REPORT

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ABSTRACT

Paget's bone disease is rare. It is less common under the age of 40. Facial polyostotic involvement is exceptional. We report a case of a 17-years-old male with chronic right nasogenous swelling, with a right hemiprognathic facial deformity. A CT scan revealed bone thickening with osteocondensation of the right hemiface. The diagnosis of facial localization in Paget's disease was retained and clinical follow-up was initially undertaken. No signs of complications were detected.

KEYWORDS: CT-scan, Hemifacial disfigurement, Paget's bone disease, Polyostotic, Youth.

INTRODUCTION

Paget's bone disease is a rare in Africa.[1] Typically, it affects 3-4% of the population over the age of 45.[2] In rare cases, it occurs under the age of 40.[3] It is characterised by a focal bone metabolic disorder with a considerable and uncontrolled osteoclastic resorption and bone remodelling.[4] Diagnosis can be assessed by radiologic investigation. We report the case of Paget's disease with hemifacial localization discovered in an adolescent in order to describe the clinical and CT features of this disorder.

CASE REPORT

A young 17-year-old male is referred to our medical imaging centre for a chronic right nasogenous swelling. He had no substantial personal or family, medical or surgical history. The right nasogenous swelling would have progressed over the past year progressively, involving a facial disfigurement. There was no pain, no fever, and no change in overall status.

Clinically, there was a firm painless tumefaction in the right nasogenous region, with a right hemiprognathic facial deformity. There was no skin change associated to the swelling. Patient did not have a clinical or biological inflammatory syndrome.

The biology tests performed had normal results. A CT scan of the facial (Figure 1) was performed and revealed a right maxillary bone thickening extending to the right temporal bone and to the right half of the mandible, with homogeneous condensation of the affected bone structures. The right temporal bone was accompanied by a narrowing of the internal acoustic meatus. Clinical follow-up was undertaken initially, and the patient showed no signs of complication at that time.

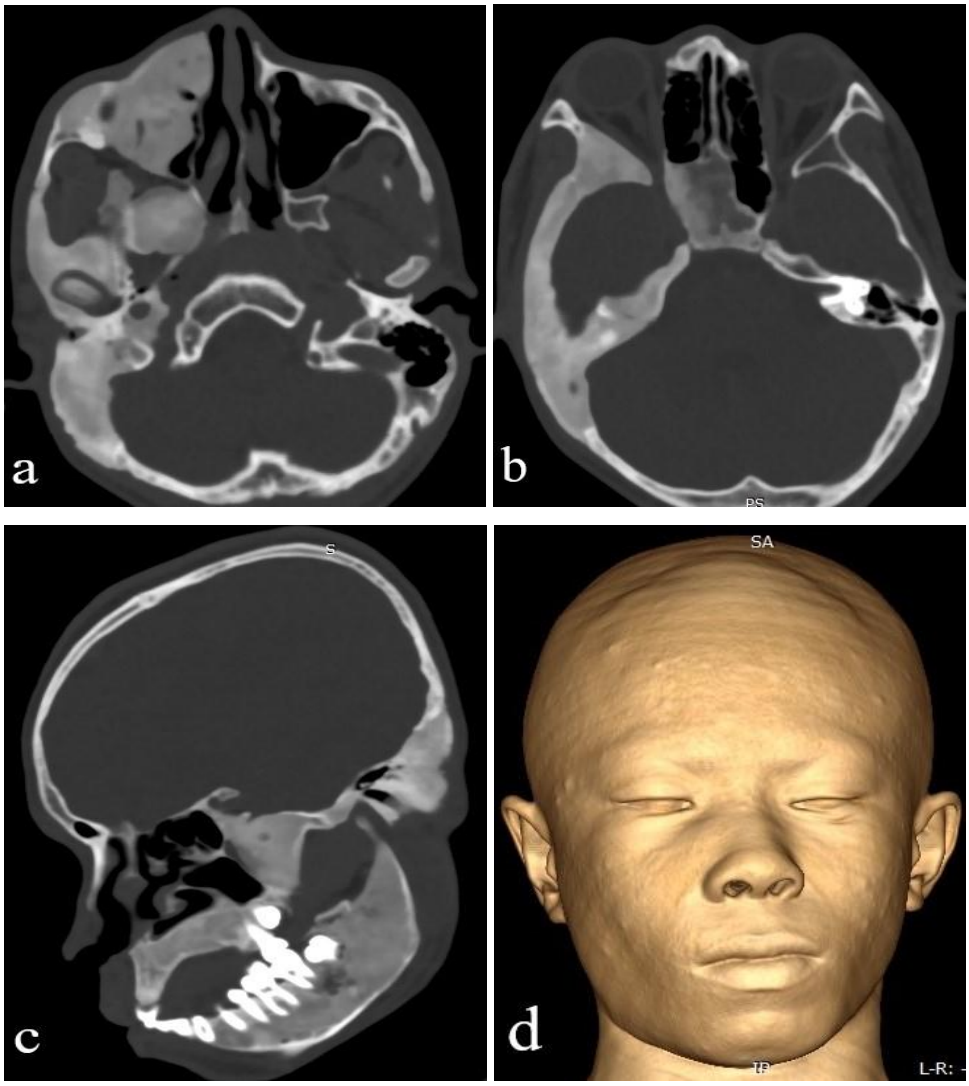

Figure 1: CT-scan of the skull showing thikening of the right maxillary, mandible, ethmoidal aned temporal bone (a, b, c). Facial deformity on Volume rendering reconstruction (d).

DISCUSSION

Paget's disease affects 3-4% of the population over the age of 45. [1]. Its incidence varies significantly between countries.[5] Incidence is higher in the United Kingdom than in other countries, also common in Australia, New Zealand, Western Europe, and the United States.[6] In this report, the concerned patient was a 17-year-old young male. According to the available literature, Paget's bone disease is more frequent in patients over the age of 50.[2] It is exceptional before 40 years old but our case confirms male predilection, resulting a sex ratio of 1:5.[6]

Its pathogenicity is not as yet well established.[7] Viral infection with a paramyxovirus in association with genetic predisposition has been evocated.[8] Classically, there are three chronological stages.[9]

- Early destructive stage with a osteoclastic activity
- Intermediate stage with a osteoblastic and an osteoclastic activity
- Late stage or inactive stage marked by sclerotic and osteoblastic activity

These stages are well correlate with the imaging findings. It is characterized by the gradual replacement of normal bone tissue with a rough, irregular and uncontrolled structure leading to an increasing bone density and a hypertrophy of the affected bone. The result of heightened osteoclastic activity and imperfect subsequent restoration, is a less-compact, deformable, fragile and fracturable bone.[7]

At the time of diagnosis, the majority of patients are asymptomatic. If present, symptoms include localized pain and tenderness, increased bone size involving deformation, bowing deformities, signs of complication as deafness, or sarcoma transformation.[2] In this patient, we found a facial deformation and a notion of deatness.

Two biochemical markers are useful for diagnosis: total plasma alkaline phosphatase and urinary excretion of hydroxyproline.[3]

The radiological findings will depend on the phase of the disease.[10,11] Plain radiography and CT scan show osteolytic lesions followed by rough trabeculae and bone enlargement. Sclerotic changes appear later in the disease process. Additional destructive features may become apparent if malignant transformation occurs.

Skull involvement shows initially a circumscribed osteoporosis leading into an large, well-defined lytic lesions involving the inner surface of the outer table of the skull with a preserved inner table**.** Then appears a mixed lytic and sclerotic lesions of the skull taking a cotton wool appearance. Latter**,** both inner and outer calvarial tables are involved. A pathognomonic sign is an enlargement of the frontal bone, with the appearance of the skull falling on the bones of the face, like a tam o'shanter's hat.

At least, the two most characteristic radiological features are bone hypertrophy and osteocondensation and fibrillar structure.[2]

In this observation, the location of the disease is the skull involving the maxilla, mandible and temporal bone in the right side with narrowing of the internal acoustic meatus.

Hemifacial involvement is uncommon. Only 11% of Paget's bone disease are localized in the facial bones.[7] Polyostotic disease is more prevalent than the monostotic type.[12] Affection is monostotic in 35% of cases.[3] The most frequent sites of involvement are spine, pelvis (often asymmetric), skull, proximal long bones.

The CT scan was able to confirm bone thickening with osteocondensation, strongly suggestive of the Paget disease. Radiology investigation can establish with certainty the diagnosis of Paget's disease. However, differential diagnosis is made with other lesions; the most frequent of which are diffuse hypertrophies as in haemolytic anaemia or renal osteodystrophy during which there may be heterogeneous osteopenia and much more frequently, local osteopenia such as frontal internal hyperostosis, which mainly affects the internal table of the frontal vault and preferably involves females, in a syndromic context.[4]

On MRI, signals of the lesion are variable. In the most common pattern, the signal intensity in Pagetic bone is similar to the intensity of fat as a high signal on T1 and T2 corresponding to à longstanding disease. In a mixed stage, it shows a low alteration of the T1 signal and high alteration of the T2 signal; corresponding to granulation tissue, hypervascularity, and edema. In sclerotic and ostoblastic stage, MRI show a low signal intensity on T1 and T2; correlating to the presence of compact bone or fibrous tissue. The marrow signal is preserved in all sequences except in the case of complications.

Scintiscan with 99Tc-MDP tracer is highly sensitive but not specific. It can be used to evaluate the activity of the disease, to assess the extension of the disease process and to

characterize high-risk locations.[3] Scintiscan show an increased uptake in all stages of the disease but on sclerotic and ostéoblastic stage, uptake may be normal.[13]

The treatment of Paget's bone disease is based on biphosphonates, which are strong anti-osteoclastic agents.[8] Symptomatic and high-risk forms are the main indications. Follow-up and subsequent treatment depends on clinical data and serum alkaline phosphatase values.

CONCLUSION

Paget's bone disease is a rare disorder. Prevalence varied between countries; It is uncommon before the age of 40. It is characterised by remodelling of affected bones. The facial polyostotic localisation is an unusual. Typically, radiological investigation is sufficient to establish the diagnosis by revealing bone thickening, plastic deformation, hypertrophy, and density abnormalities such as osteocondensation.

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