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Case Report

Paget's disease of maxilla: A case report

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ABSTRACT

Paget disease of bone (PDB) is a metabolic bone disease characterized by increased and disorganized bone turnover. Its etiology is not clear, but includes endocrine, genetics and inflammatory factors. Some patients are asymptomatic, whereas others develop complications such as pain, osteoarthritis, fracture, deformity, deafness, and nerve compression syndromes. Diagnosis of PDB is usually made incidentally by radiographic examinations obtained for other reasons or during evaluation of high serum alkaline phosphatase levels. In this study, we present a case of a 57-year-old female patient with complaint of swelling in the maxilla. Panoramic radiography accompanied by laboratory studies confirmed the diagnosis which was very important for choosing the proper treatment.

1. Introduction

Paget's disease of bone (PDB), first described by Sir James Paget in 1877, is a metabolic bone disease which can affect one or more bones and is characterized by abnormal bone deposition and resorption.[1,2] PDB, also known historically as "osteitis deformans", is usually seen after the fourth decade of life and males are affected more frequently.[3] In the geriatric population it is the second most frequent metabolic bone disease after osteoporosis.[4] Diagnosis of PDB is usually made incidentally by radiographic examinations obtained for other reasons or during evaluation of high serum alkaline phosphatase levels (ALP).[3]

Herein we report a case who applied to our clinic because of an expansion in the alveolar crest of the maxilla and was diagnosed with PDB and discuss the case in the light of literature.

2. Case report

A 57-year-old female patient was admitted to Department of Oral and Maxillofacial Radiology with complaint of swelling in the maxilla. Physical examination revealed maxillary expansion at an advanced stage (Fig. 1). She had a slow-progressive expansion for the last 2 years at maxillary alveolar crest, had

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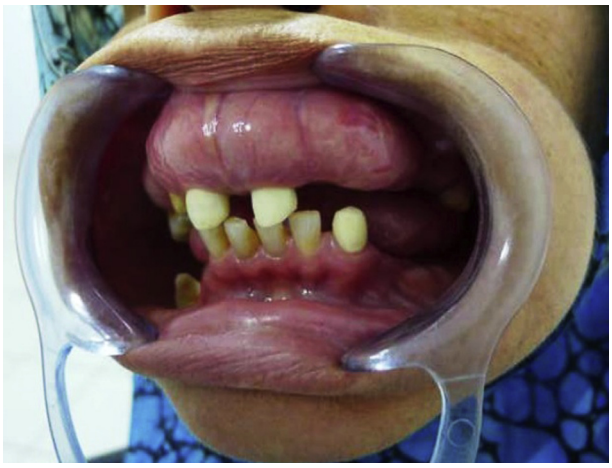

Fig. 1 – Intraoral photo of the patient.

no pain, and diastemas had occurred by the time in the maxillary incisors. Panoramic radiography showed presence of both radiolucent and radiopaque areas with the hypercementosis of right maxillary second molar (Fig. 2). She was otherwise healthy and had no complaint of pain. Laboratory studies showed elevated serum ALP level (629 U/L, N: 35–105 U/L) with normal serum calcium (9.1 mg/dL N: 8.4–10.5 mg/dL), phosphorus (3.5 mg/dL N: 2.3–4.7 mg/dL) and parathormon levels (52.48 pg/mL, N: 12–72 pg/mL). Bone biopsy confirmed the diagnosis of Paget's disease so the patient was referred to Endocrinology-Metabolism Out-patient Clinic, Cerrahpasa Medical Faculty, University of Istanbul.

Additional laboratory studies revealed low level of 25 dihydroxyvitamin D level (9 ng/ml, N: 20–120 ng/ml) with high levels of bone formation and resorption markers [bone specific ALP was 591 U/L (N: 35–105 U/L) and β-CTX was 1.95 ng/ml (N: 0–1 ng/ml)]. Bone scintigraphy revealed polyostatic paget's disease with increased uptake in the right occipital region of the cranium; T1, T3, T12, L1, L2 vertebraes; posterior of the right sixth rib; left first rib; maxilla and pelvis (Figs. 3 and 4). Lateral and posteroanterior radiographs of the skull demonstrated a cotton wool appearance (Figs. 5 and 6).

After the replacement therapy with vitamin D, the level of 25 (OH) 2 D Vitamin increased to 51 ug/L (N: 20–120 ug/L). Bisphosphonate therapy with 5 mg intravenous infusion of zolendronic acid was considered. All necessary dental

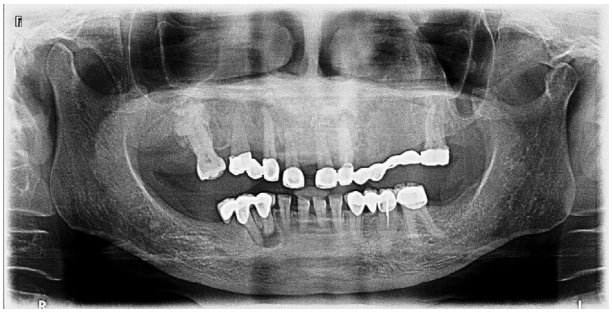

Fig. 2 – Panoramic radiography of the patient.

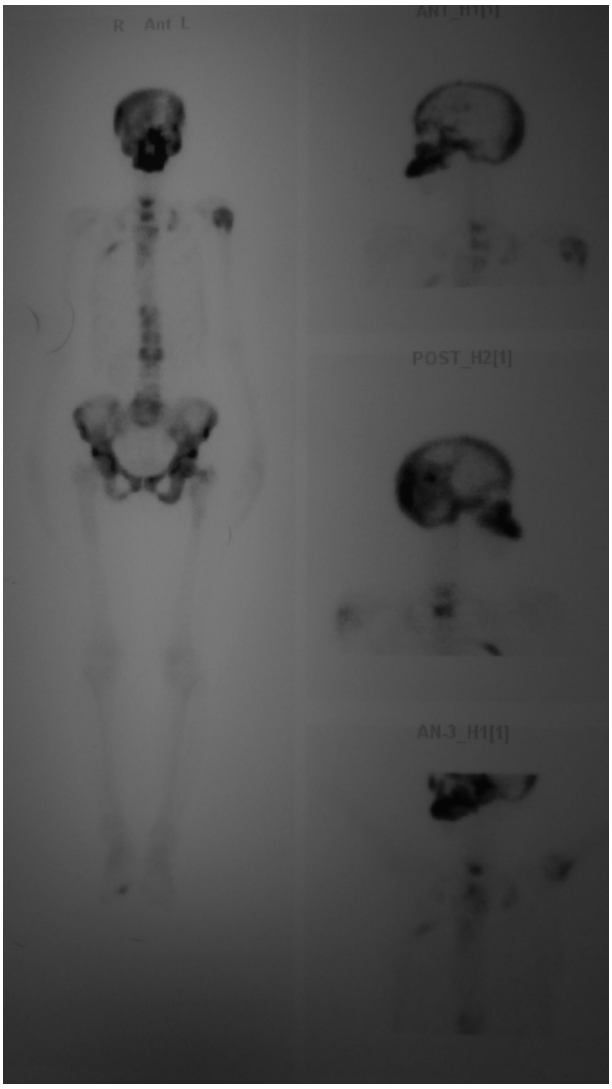

Fig. 3 – Scintigraphy of the patient (front).

treatments were completed before the bisphosphonate therapy against the risk of osteonecrosis.

3. Discussion

Prevalence of the PDB differs in various regions of the world. PDB, being frequent in United Kingdom (5%), Australia (3.6%) and North America (3.9%), has low prevalence in Scandinavia, Italy and Spain (0.5–1.1%).[5] Although the etiology of PDB is uncertain, it is thought that endocrine, genetics and inflammatory factors are influential. Paramyxoviruses might have a role in etiology.[6] (15–40%) of the cases have family history of PDB and there is a positive correlation between PDB and mutations of the SQSTM1 gene.[2,7] It is also hypothesized that vitamin D deficiency in childhood has an effect on PDB in the coming years as well.[1] PDB has monostatic and polyostatic types. It most frequently effects pelvis, femur, tibia, vertebras and cranium.[2] Although majority of the patients are asymptomatic, PDB may also cause pain, deformities, and fractures of the involved