Management of craniofacial osteitis deformans

Access this article online
Website: www.amsjournal.com
DOI: 10.4103/2231-0746.147167
Quick Response Code:

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ABSTRACT

Paget's disease of bone (PDB) is a progressive chronic disease of unknown etiology, manifested as initial increase in bone resorption, followed by a disorganized and excessive formation of bone, resulting in pain and skeletal deformities. Treatment of the disease primarily aims at reducing the altered bone turnover by pharmacotherapy, along with surgical intervention for deformed bones, to reduce morbidity and improve the quality of living. We present a case of PDB with craniofacial involvement in a 55-year-old female, diagnosed on the basis of clinical features, radiological assessment along with biochemical findings and histopathological report. Our management included medical treatment in the form of intravenous bisphosphonates which alleviated symptoms by reducing the abnormal bone activity, and normalized serum alkaline phosphatase. Patient morale, self-confidence and social acceptance were all boosted due to the surgical correction of the facial deformity.

Keywords: Alkaline phosphatase, bisphosphonates, Paget's disease

INTRODUCTION

Paget's disease of bone (PDB) was first reported by Sir James Paget under the term osteitis deformans in 1877.[1] It is common in European descents, rare in Asians. This disease is found in 5th decade of life with more predilections for males. Cranium involvement is seen in 28% and jaw bones in 17% of cases.[2] Different treatments are advised in literature with varying success rates. The current mainstay for treatment of PDB is bisphosphonates which suppress the bone resorption by osteoclasts.[3] Present case report uniquely describes the successful management of craniofacial PDB by both medical and surgical means.

CASE REPORT

A 55-year-old female reported to Department of Oral and Maxillofacial Surgery with the chief complaint of a gradual increase in the size of the upper jaw since 20 years, along with headache and dizziness since 4 years. Weakness of the facial nerve was not elicited. No relevant medical history was recorded. On clinical examination, patient was short stature, hunched back with a slow waddling gait. Patient had an enlarged cranium with broad, prominent forehead and severely protrusive, expanded maxilla, caused obliteration of nasolabial sulci. The nasal bones were also increased in size with hypertelorism, and mandibular contour was normal [Figure 1]. On palpation, enlarged maxilla was bony hard and nontender. Rise in temperature over the swelling was felt, but overlying skin and mucosa appeared normal. Intraorally, maxilla presented as a multilobular swelling from the right maxillary tuberosity extending to the left tuberosity resulting in obliteration of the maxillary buccal and labial vestibule. Severe protrusion and spacing were seen with anterior maxillary teeth [Figure 2].

Radiographs showed "cotton wool appearance" in the skull and maxilla in postero-anterior (PA) view and lateral view of the skull [Figure 3] but mandible appeared normal. Orthopantomogram displayed "cotton wool appearance" of maxillary alveolus. Intraoral periapical radiograph revealed loss of lamina dura in the maxillary teeth. Computed tomogram (CT) scan illustrated mixed hyper and hypo dense lesion involving complete cranium, maxilla and nasal bone [Figure 4]. Three-dimensional CT scan demonstrated expansion of maxilla and cranium. PA and lateral spine view elucidated involvement of lumbar vertebra (radiolucent lesion) at L1, L2 with scoliosis at same

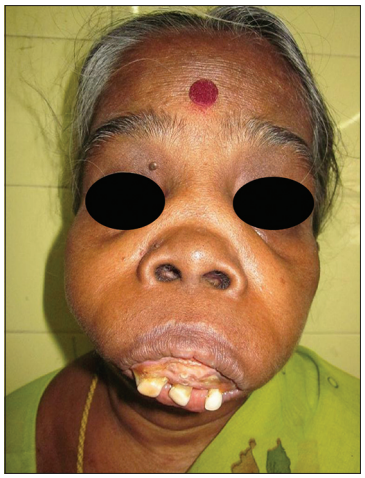

Figure 1: Extraoral view preoperative

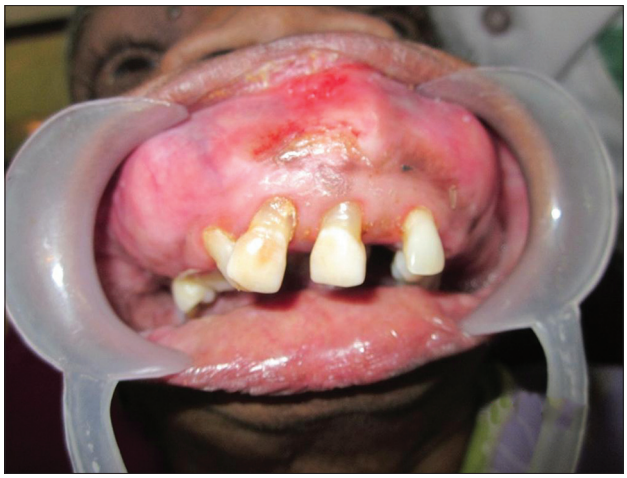

Figure 2: Intraoral view preoperative

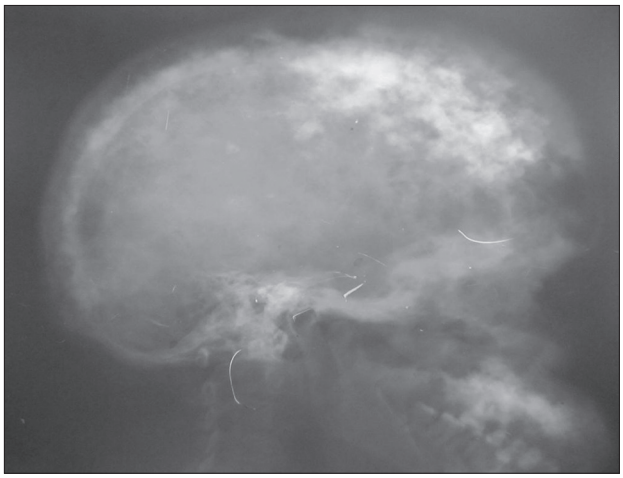

Figure 3: Lateral view of the skull shows "cotton wool appearance"

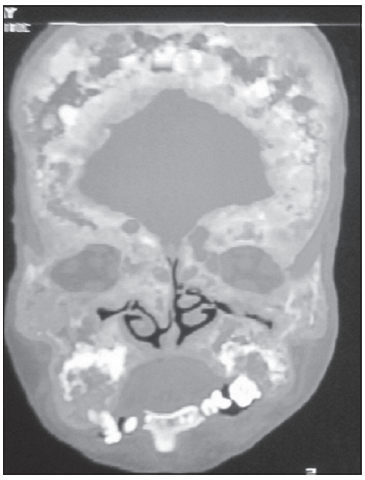

Figure 4: Computed tomogram scan shows involvement of cranium and maxilla

region. On studying the clinical features and radiological findings, a provisional diagnosis of PDB and a differential diagnosis of polyostotic fibrous dysplasia were given. Incisional biopsy and biochemical tests were performed to conclude final diagnosis.

Biochemical report showed routine blood parameters in normal range. Serum alkaline phosphatase (SAP) level was raised to 1020 IU/L (normal range 70-170 IU/L) which indicated approximately 10-fold increase in the SAP level. Serum calcium, serum phosphorus, and parathyroid hormones levels were in the normal range [Table 1].

Histopathologic examination revealed bony trabeculae with basophilic reversal lines in a mosaic pattern suggestive of Paget's disease. Numerous osteoclasts were found surrounding the irregular bony trabeculae.

Thus, a final diagnosis of PDB with craniomaxillary involvement was made on the history, clinical, and radiographic features along with biochemical and histopathological reports.

Patient was psychologically stressed due to facial deformity. Hence, active treatment in the form of surgery was mandatory.

Table 1: Biochemical report of the patient showing values within the reference range

Test	Patient's reported value	Reference range
Serum calcium (mg/dl)	7.8	8.5-10.0
Serum phosphorus (mg/dl)	4.2	4.5-6.5
Parathyroid hormone (pg/dl)	50.47	15-65

Neo-adjunctive therapy with bisphosphonates was planned due to extensive craniomaxillary involvement which resulted in a severe increase in the SAP levels. After explaining treatment to the patient, informed consent was obtained. To avoid osteonecrosis, prophylactic extractions of periodontally involved maxillary anterior teeth were performed under local anesthesia [Figure 5]. One week later, injection aclasta-5 mg (zoledronic acid, Novartis India) single dose was administered by intravenous route. Patient observed for 1 month after infusion for any immediate reactions or adverse effects of the drug.

Surgical contouring of maxilla was done under general anesthesia. Intraoral maxillary vestibular incision was given bilaterally to reflect mucoperiosteal flap which exposed the underlying bone [Figure 6].

Infraorbital nerves were identified on both sides and secured. Bone reshaping was done by hand and motor driven instruments to achieve bilateral symmetry [Figures 7 and 8]. Remaining maxillary teeth were extracted to facilitate bone recontouring. Water tight closure of mucosa was strictly followed with 3-0 vicryl sutures to avoid bone exposure and subsequent osteonecrosis. Postoperative medications and instructions were given. No postoperative complications encountered apart from transient paresthesia of both infra-orbital nerve which gradually resolved in 4 weeks. Biochemical test done after 2 months of surgical intervention,

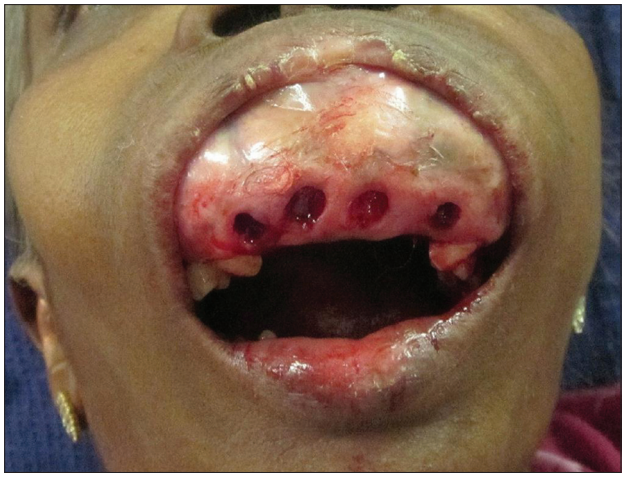

Figure 5: Extraction done of 12, 11, 21, 22

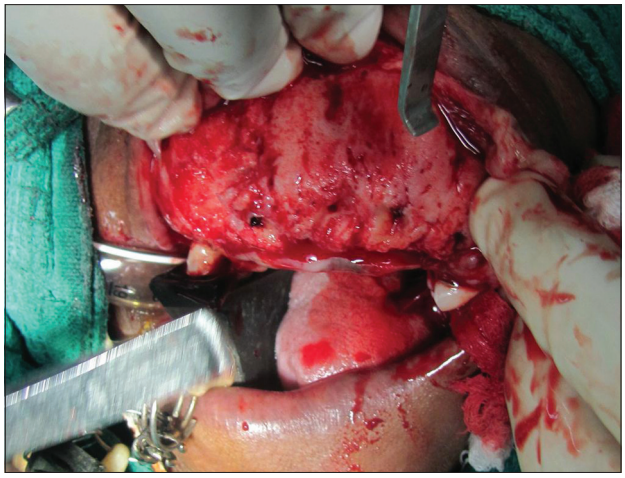

Figure 6: Intraoperative view-exposure of enlarged maxilla

Figure 7: Bone removed after recountering

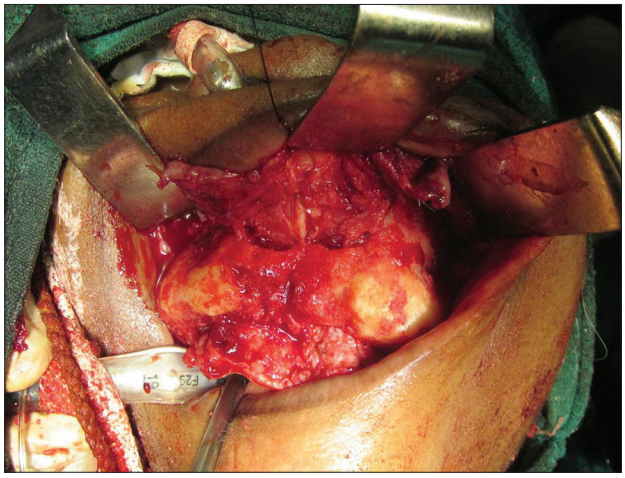

Figure 8: Intraoperative view-maxilla after reshaping

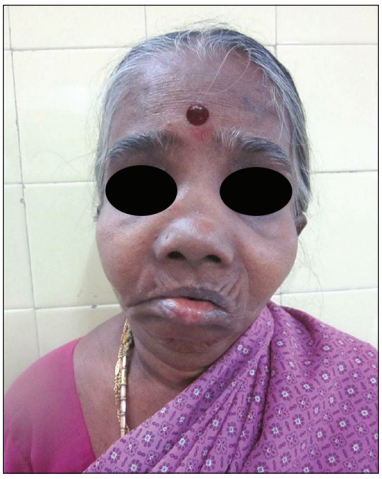

Figure 9: Extraoral view-postoperative 1 year

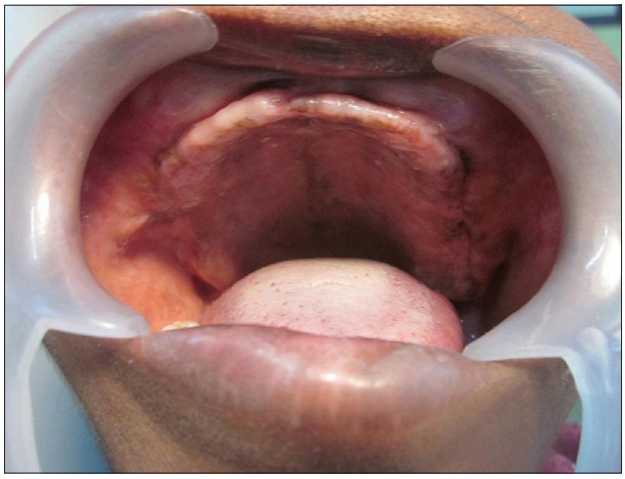

Figure 10: Intraoral view-postoperative 1 year

showed decreased SAP level from 1020 IU/L to 97 IU/L. Patient was regularly followed-up for 1 year at every 1-month interval for first 6 months and every 3 months, thereafter [Figures 9 and 10].

In the 1-year follow-up period, neither regrowth was observed postoperatively nor evidence of osteonecrosis in both jaws. Patient was symptom-free with a substantial reduction in headache and backache attributable to medical treatment.

DISCUSSION

Paget's disease of bone is a chronic bone remodeling disorder characterized by increased osteoclast-mediated bone resorption, with subsequent compensatory increases in new bone formation, resulting in a disorganized mosaic of woven and lamellar bone at affected skeletal sites.[3] Etiology is uncertain, but both environmental and genetic factors are thought to be involved in the pathogenesis.[1]

Rationale for treatment in PDB is (1) symptom control (2) Reduce long-term complications and (3) surgery for bone deformity or pathologic fracture.[4] Specific available medical treatment for PDB is (i) calcitonin in form of salmon calcitonin was used to treat PDB in past. It reduces bone pain and improves SAP level. Efficacy is low and resistance to treatment can develop. (ii) Second generation bisphosphonates are widely used as these are potent drugs with fewer side effects (approved by Food and Drug Administration 2001).[5] Bisphosphonates promote osteoclastic apoptosis and reduces bone turnover by reducing osteoclastic activity. The disadvantage of these drugs is a risk of osteonecrosis when used for long-term and as high dosage intravenous therapy.[6] Gallium nitrate (mithramycin) and estrogen have been tried but were not found to be effective.[7] Intravenous bisphosphonates are good neoadjuvant, and surgical correction is useful to improve the quality-of-life. Similar treatment protocol was followed in our case. Remarkable improvement in the facial esthetics encouraged participation in social activities which the patient had ceased since past 15 years.

This rare and successfully treated case concludes that bisphosphonates if used precisely can be a potent drug for normalizing SAP level in PDB patients and provide symptomatic relief. Consideration for surgical intervention is necessary to reduce deformity, morbidity, and improve the quality of living.

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Cite this article as: Umamaheswari G, Pangarikar AB, Urade VB, Parab PG. Management of craniofacial osteitis deformans. Ann Maxillofac Surg 2014;4:243-6.

Source of Support: Nil, **Conflict of Interest**: None declared.