Massive osteolysis of hemimandible: a case report

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Abstract Massive osteolysis or Gorham's stout disease or phantom bone is a rare disorder which normally appears in adulthood with no specific predilection for any sex. Work to find out exact etiology of disorder has been done. But actual cause has not yet been found. Any of the bone could be involved in this disease. A clinical, radiographic and histological evaluation of massive osteolysis of mandible is presented and discussed.

Keywords Gorham's stout disease · Massive osteolysis · Mandible

Introduction

Massive osteolysis or GORHAM STOUT disease is also known as disappearing bone disease or vanishing bone disease or phantom bone [3]. It is an insidious, slowly progressive and extremely destructive disease of bone.

It was first described by Jackson [8] in 1838 and Gorham, stout in 1955 established the association between massive osteolysis and presence of hemangiomas and lymphangiomatous tissue [5]. It is exceedingly rare with fewer than 100 cases reported in the literature. Partial or complete resorption of normal appearing bone occurs which may continue to progress for years. In some cases, resorption stops spontaneously, but regeneration fails to occur. Etiopathophysiology are unknown but can be discrete or multifocal with most commonly affected sites being shoulder [16], upper arm [16], pelvis [11], jaw, spine [15] and thorax [15].

Basically bone is apparently resorbed and may be replaced by fibrous tissue. But no other peculiar features are present. Literature suggest that bone resorption in GSD patients is due to enhanced osteoclastic activity and IL-6 plays an active role in increased bone resorption [14]. Gorham stout disease appears to be due to increase in number of osteoclast but due to increase in their activity [7].

This disorder is usually asymptomatic at the initial stages and is discovered on

routine radiographs in some patients whereas some patients may complain of deformity, disturbed occlusion, pain, impaired function or may have pathological fracture [15].

Case report

A patient aged 25 years presented to the department of oral and maxillofacial surgery of Punjab Government Dental College and Hospital, Amritsar with a chief complaint of pain and slight deformity of face (Fig. 1). History revealed nothing significant apart from continuous slow progressive bone resorption for about two years which has become static for the last six months.

There was no history of trauma, infection, neoplasm or previous jaw surgery. Family history was non contributive.

On clinical examination patient was found in pain with slight asymmetry of face and slight depression of right ramus. No bony structure was found distal to canine on the right side to the condylar region. Remaining mandible was freely mobile with slight pain.

No other skeletal abnormality to remainder of the skeleton was found on clinical survey. Lateral oblique view and CT scan for facial bones was advised to the patient.

Lateral oblique view and scan showed no bone distal to the canine. The coronoid process and condyle were also missing with only floating teeth.

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Scan revealed precise zone of osteolysis which showed complete resorption of right side of mandible including condyle and coronoid process up to canine region along with floating teeth (Figs. 2 and 3)

Repeated complete lab assessments were all normal. Endocrine assessments for (thyroid, diabetes, parathyroid) hematological findings (complete blood count, ESR etc.) and biochemical assessment (urea, creatinine, alkali and acid phosphatase) were all normal.

FNAC (Fig. 4) and histopathological (Fig. 5) findings showed high concentration of thin walled vascular channels with some active fibroblast and chronic inflammatory cell infiltration. There was no evidence of malignancy.

Differential diagnosis

The diagnostic work up eliminated any infectious disorder (osteomyelitis) endocrine disorder (diabetes, thyroid) vascular disorder (hemangiomas), malignant tumour (sarcoma).

Since patient's condition did not reveal any of the CNS system disorder, osteolysis associated with diseases of CNS such as tabes dorsalis, syringomelia, leprosy or myelodysplasia could be ruled out.

Surgical exploration of the resorbed area was done and residual bone was submitted for histopathology. Reconstruction of the mandible was done using reconstruction plate (Figs. 6 and 7).

Extensive radiolucent lesion could correspond to either an odontogenic or non-odontogenic tumor but radiographic appearance was demarcated by radiolucency reflecting osteolysis.

Final diagnosis of massive osteolysis of right body and ramus of mandible was made.

Treatment was done which included surgical exploration of area of bone resorption. Biopsy of residual bone was done. Reconstruction was done by reconstruction plate (Figs. 7 and 8). As the higher risk of bone infection a bone graft was not done.

Regular follow up at frequent intervals has been done till date, patient has no complaints. There was no pain on movement. Function and occlusion were maintained. No further resorption or regeneration of bone was evident (Figs. 8 and 9).

Discussion

Multiple osteolytic states affect the axial and appendicular skeleton, among them include occupational acro-osteolysis, post-traumatic osteolysis and idiopathic (sometimes hereditary) multicentric osteolysis [13].

Gorham's stout disease is an unusual and uncommon disease characterized by spontaneous, progressive resorption of bone with ultimate total disappearance of the bone [15]. It is of unknown etiology but appears to be related to an active hyperemia of bone. Clinical case reported here did not present any ulcerative or clinically invasive lesion, cellular atypia, osteoblastic response, calcification, visceral disease or hereditary, metabolic, neoplastic, immunological or infectious etiology.

Several publications in literature have reported difficulties in establishing a diagnosis. Histological examination do not provide an actual diagnosis but remain essential to exclude differential diagnosis [6]. The positive diagnosis is difficult for number of reasons. The disease has insidious onset. It evolves by successive phases of bone destruction, over a period of several months to years. Moreover cases of spontaneous arrest [15] has also been described.

There seems to be no specific treatment of the lesion. External beam radiotherapy has been noted to have a marked effect on progression of disease [10]. Various other treatment modalities [9] have been proposed with marked variability in the result. Treatment based on calcium, vitamin

Fig. 1 Preoperative photograph

Fig. 2 Intraoral

Fig. 3 CT scan

Fig. 4 CT scan

Fig. 5 FNAC

Fig. 6 Histopathology

Fig. 7 Intraoperative reconstruction plate

Fig. 8 Postoperative radiograph

C and D, calcitonin [4], bisphosphonates and estrogens [12] have been described in the literature.

Surgical resection along with reconstruction offered the esthetic,

functional and psychological benefits to our patient.

Prognosis of the disease is unpredictable with one reported case of spontaneous arrest of the osteolytic process

Fig. 9 Postoperative frontal view

Fig. 10 Postoperative lateral view

and at the other end of spectrum cases of rapid and aggressive deterioration have been reported that may be life threatening (pleural effusion due to costal involvement) [2].

Positive diagnosis of this rare little known disease is very difficult to establish [1]. Diagnosis cannot be confirmed by histological examination therefore the term bone osteolysis should be reserved for a particular clinical entity and should not be used to designate any histologic changes.

No evidence of etiology of the disease was obtained from history, biochemical investigations or from histological studies. Because of the fear of resorption, assessment of any method of treatment is difficult. The results of this mode of therapy are impossible to evaluate at the present time.

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