Gorham Disease an Enigma

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Gorham Disease an Enigma

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Abstract

Vanishing bone disease, or Gorham disease, was first defined as a specific entity by Gorham rare disorder characterized by proliferation of vascular channels, which results in destruction and resorption of osseous matrix. Only a few cases have been reported in the jaws. Vanishing bone disease or massive osteolysis of the lower jaw will initially affect the mandibular basal and alveolar bone, which subsequently involves the rami and the condyles.

Introduction

Gorham disease is a rare entity of unknown etiology, uncertain pathogenesis unpredictable prognosis. First described by Jackson in 1838. Reviewed extensively by Gorham in 1955, hence known as Gorham Disease. Also known as Vanishing Bone Disease, Massive Osteolysis, Gorham- Stout Syndrome or Phantom Bone Disease.

Case Report(s)

A 17 year old girl reported to Department of Oral Medicine and Radiology, G.D.C.H. Nagpur with a chief complain of pain and facial deformity in lower left posterior region of face since 6 months.

Intraoral Examination revealed a deformity in between 33, 34 with segmental mobility of 34, 35, 36, 37, 38. The collapsed arch was below the occlusal plane. No ulceration or growth were noted.

Radiographic Examination

The left lateral oblique radiograph taken 3 month back revealed severe alveolar bone loss with 35, 36, 37, 38. The body and ramus were resorbed. With only the inferior border of Mandible visible. Root resorption was seen with 37,38. 35,36,37,38 were pushed towards inferior border of mandible.

Recent Orthopantomograph revealed slight mesial drifting of teeth without any further bony destruction.

Computed tomography presented complete destruction of body and ramus of left side of mandible. The Head of condyle was spared from destruction.

MPR and surface rendering revealed vanishing of body and ramus of mandible and tapering of bone.

Differential Diagnosis

Eosinophilic granuloma.

Osteomyelitis.

On the basis of clinico

1. Radiographic features provisional diagnosis of Gorham disease was made.

Investigations

Differential leukocyte count and serum chemistry were within normal limits.

1. FNAC was inconclusive.

2. Biopsy was advised.

Histopathological Findings

Histopathological examination revealed highly vascular connective tissue with numerous dilated and engorged thin walled blood vessels. A cellular infiltrate was present. Large amount of extravasated blood was also evident.

Final Diagnosis

On the basis of clinico-radiographic and histopathologic findings, final diagnosis of Gorham disease was made.

Discussion

Gorham disease is a rare disease characterized by - Spontaneous and usually progressive destruction of one or more bones. The destroyed bone is replaced by vascular proliferation which is eventually replaced by dense fibrous tissue. Usually only one bone is affected.

The most commonly affected bones are Clavicle, Scapula, Humerous, Ribs and Sacrum. Involvement of Maxillofacial bones is very rare. Only about 39 cases of maxillofacial involvement have been reported till date.

Eight Clinical and Histopathological Criteria

1. Biopsy with angiomatous tissue.

2. Absence of cellular atypia.

3. Minimal or no osteoblastic response and absence of dystrophic calcification.

4. Evidence of local, progressive osseous resorption.

5. Non expansive, Non ulcerative lesion.

6. Absence of visceral involvement.

7. Osteolytic radiographic pattern.
8. Negative immunologic and infectious etiology.

**CLINICAL FEATURES**

Mostly affects all ages, ranging from 1 month to 66 yrs.

1. The average presenting age is 29.6 years in patients with facial bone involvement.

There is 2:1 male to female ratio.

The number of reported cases is insufficient to estimate a racial prevalence.

No family history of bone disease or developmental abnormality.

Mandible is more commonly involved as compared to maxilla, individual maxillary bone involvement has not been reported.

Clinically two sequential phases of the disease have been detected. In the first phase there is active bone destruction and lysis associated with mild to moderate pain, soft tissue swelling, tenderness and varying degree of erythema followed by radiographic evidence of progressive bone destruction. In the second phase there is absence of swelling, pain, bone destruction or radiographic evidence of bone repair.

1. Clinical course of the disease is usually unpredictable, passing at undetermined intervals from active to quiescent phases.

2. In the presence of secondary infection there is reactive lymphadenopathy.

3. The mortality rate is relatively low, most deaths are due to the involvement of Clavicle and Ribs.

**RADIOLOGICAL CHANGES**

Early intraosseous destruction appears as small intramedullary radiolucencies.

Late extraosseous lysis when medullary and cortical destruction occurs simultaneously.

Absence of periosteal reaction or osteosclerosis.

In tubular bones a “tapering” effect, attributed to the collapse of unsupported cortical plates.

**TREATMENT**

Attempts of bone grafting have been made. Normally the grafted autogeneous bone undergoes necrosis and heals by “Creeping substitution”.

1. Chemotherapy and radiotherapy have been attempted but not widely accepted as standard treatment.

2. A regimen consisting of Vit. D 50,000 units/ week, Calcium glycerophosphate 6 gm/day and Sodium flouride 50 mg/day had been tried successfully.

**Conclusion**

Even though Gorham disease is rare it should be included in differential diagnosis of any obscure osteolytic disease.
Illustrations

Illustration 1

Intraoral view of floating teeth

Illustration 2

Lateral oblique view of mandible
Illustration 3

Orthopantomogral showing floating teeth

Illustration 4

Three dimensional reconstruction on CT scan showing massive osteolysis
Illustration 5

Histopathological picture of tissues
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