

Thin atrophic mandible, a rarity in young adults Vanishing Bone Disease: A Case Report

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ABSTRACT

Loss of teeth following aging is a common physiological phenomenon, but the same in a young adult is something to think on. Vanishing or Disappearing bone disease (Phantom Bone, Gorham Syndrome, Massive Osteolysis) [2] is an unusual and uncommon disease characterized by spontaneous, progressive, resorption of bone leading to total disappearance of the same. It is of unknown etiology but appears to be related to an active hyperemia of bone and lymphatic proliferation. It must be appropriately differentiated from Osteolysis a due to infection (osteomyelitis, rheumatoid arthritis), Osteolysis associated with pre existing periodontal disease, Osteolysis because of disease of central nervous system (tabes dorsalis, syringomyelia, leprosy, myelodysplasia), osteogenesis imperfect, osteopetrosis. Most commonly one bone is affected but cases of multiple bone involvement too have been reported in the literature. The bones commonly affected are the clavicle, ribs, scapula, humerus, ilium, and sacrum, but less commonly the face and jaw bones. This is a case report of 39 years male who presented with a sinus opening and pus discharge from the right lower jaw region. The patient gave a history of wearing complete denture since last 8 yrs. A step deformity was palpable on the right posterior aspect of a thin and atrophic mandible indicating a pathological fracture of the right mandible. A panoramic radiograph was taken which correlated with the clinical findings. The patient was taken up for debridement of the infection under general anesthesia following which an inferior/lower border augmentation of the mandible was done using rib and iliac crest graft. The histopathological report came as fracture callus with no evidence of malignancy. The patient has been on periodic follow up and has been planned for prosthesis as there have not been any signs of infection. Atrophic mandible in a young adult is an uncommon finding and the reason for the cause is difficult to arrive at. Various differential diagnosis like vanishing bone disease, osteogenesis imperfect, osteopetrosis, osteolysis associated with infection/rheumatoid arthritis, pre existing periodontal disease can be given. A thorough workup including relevant investigation should be undertaken prior to planning a treatment and executing the plan on a priority basis or else the patient may not have a lower jaw to chew.

Keywords: thin atrophic mandible, young adult, vanishing bone disease, Gorham disease, massive osteolysis, uncontrolled proliferation of distended, thin-walled vascular or lymphatic channels within bone.

INTRODUCTION

Loss of teeth following aging is a common physiological phenomenon, but the same in a young adult is something to think about. Atrophic mandible in a young adult is an uncommon finding and the reason for the cause is difficult to arrive at.

Vanishing or Disappearing bone disease(Phantom Bone, Gorham Syndrome, Massive Osteolysis) [2] is an unusual and uncommon disease characterized by spontaneous, progressive, resorption of bone leading to total disappearance of the same.

It is of unknown etiology but appears to be related to an active hyperemia of bone and lymphatic proliferation. It must be appropriately differentiated from Osteolysis a due to infection (osteomyelitis, rheumatoid arthritis), Osteolysis associated with pre existing periodontal disease, Osteolysis because of disease of central nervous system (tabes dorsalis, syringomyelia, leprosy, myelodysplasia), osteogenesis imperfect, osteopetrosis.

Most commonly one bone is affected but cases of multiple bone involvement too have been reported in the literature. The bones commonly affected are the clavicle, ribs, scapula, humerus, ilium, and sacrum, but less commonly the face and jaw bones.^{[5][6][7][8]}

CASE REPORT

A 39 years male presented with a sinus opening and pus discharge from the right lower jaw region. The patient gave a history of wearing complete denture since last 8 yrs and a history of beetle leaf chewing with tobacco three times daily as well as regular alcohol consumption for the last decade. A step deformity was palpable on the right posterior aspect of a thin and atrophic mandible indicative of a pathological fracture.

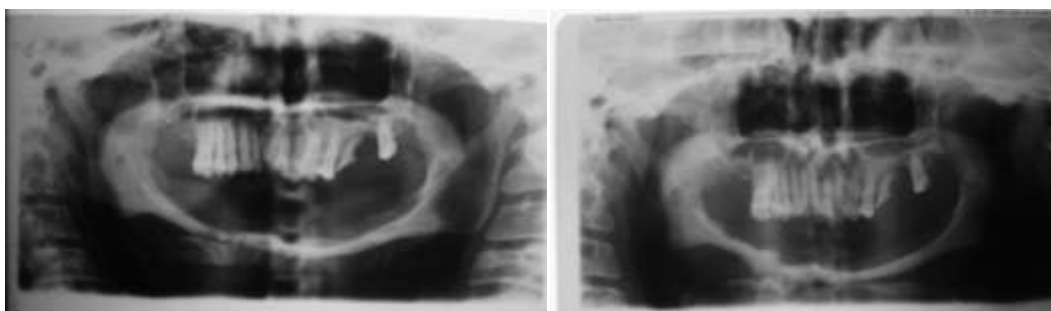


Pain was intermittent in character and aggravated on chewing food and relieved with jaw in resting position. Patient did not give any history of fever, difficulty in swallowing, speech, disturbance or weight loss. Patient gave a h/o **below knee amputation** for sepsis of left lower limb around 15 yrs back and **hearing loss (rt>lt)** Incisional biopsy from rt lower alveolus revealed granulation tissue and fibrosis but no malignancy. He was put on a course of tab clindamycin (600mg) twice daily for a week. Sinus tract subsequently healed. Mouth opening was normal with no restriction in movements.



Radiological evaluation:

A panoramic radiograph was taken which correlated with the clinical findings.



Investigations:

Complete blood profile, Complete urine analysis, Renal function test, Liver function test, Random blood sugar, Bleeding profile, Clotting profile, Alkaline phosphatase (ALP), Acid phosphatase, Serum calcium, phosphorous, HIV / HBsAG were all normal so were the chest x ray and ECG Based on the clinical and radiographic findings a clinical diagnosis of Generalized thin atrophic mandible (Vanishing bone disease) secondary to chronic bone infection was given.

Surgery:

The patient was taken up for debridement of the infection under general anesthesia following which an inferior/lower border augmentation of the mandible was done using rib and iliac crest bone graft.



Fig 1: Visor Incision marking



Fig 2: Defect on the lower border



Fig 3: Incision marking for harvesting rib graft



Fig 4: 5th intercostal rib opened



Fig 5: Bone graft harvested

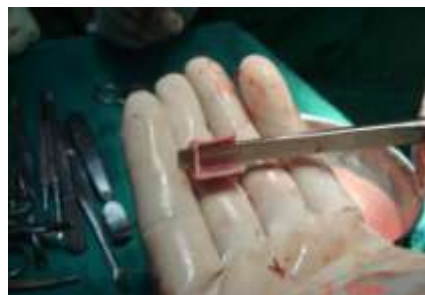


Fig 6: Rib graft splitted



Fig 7: Residual defect after rib graft secured with screws



Fig 8: Harvesting Iliac crest graft- medial aspect of thigh



Fig 9: defect obliterated after placement of graft



Fig 10: sutures and drain in place

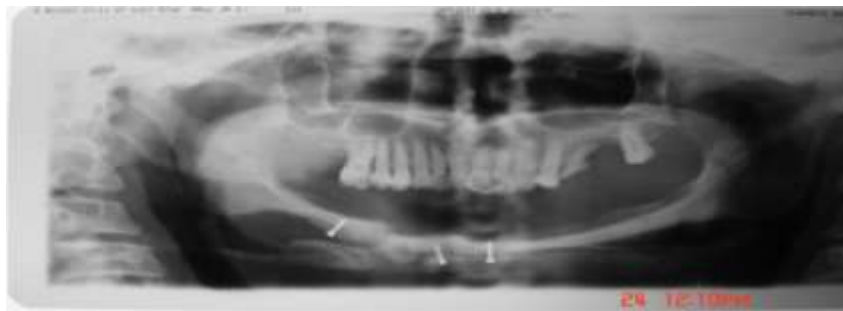


Fig 11: post op OPG, showing autogenous graft

The histopathological report : fracture callus with **no evidence of malignancy**. The patient has been on periodic follow up and has been planned for prosthesis as there has not been any sign of recurrence of infection.

DISCUSSION

Vanishing bone disease is seen in older children, and young and middle aged adults affecting males and females equally. The disease has a typical characteristic to begin suddenly and rapid advancing till the involved bone is replaced by a thin layer of fibrous tissue surrounded by a cavity. In many cases, the final result of Vanishing bone disease is severe deformity and functional disability.^{[5][6][9]}

The typical histologic feature is replacement of bone by connective tissue containing many thin walled blood vessels or anastomosing vascular spaces lined by endothelial cells. There have been very few case report of destruction of entire jaw. One of the consistent finding of the disease is pathological fracture of the involved bone.

Difficulty in breathing and chest pain may be present if the disease is present in the ribs, scapula, or thoracic vertebrae. These may indicate that the disease has spread from the bone into the chest cavity. The breathing problems may be confused to bronchial asthma.^[7] Extension of the lesions into the chest may lead to the development of chylous pleural and pericardial effusions.

These complications or their symptoms, such as difficulty breathing, chest pain, poor growth or generalised weakness leading to weight loss, and infection have sometimes been the first indications of the condition.^{[7][8][9]} It may or may not be painful, with swelling. However, all laboratory investigations are usually normal.

Differential Diagnosis:

Atrophic mandible in a young adult is an uncommon finding and the reason for the cause is difficult to arrive at

- **Mandibular atrophy**

Etiology:

- disuse
- pressure

- periodontal disease
- senility
- endocrine influences

Some of the disease which mimics the same or is associated with are:

- Vanishing bone disease/**Massive Osteolysis/Gorham syndrome**/phantom bone disease/disappearing bone disease/acute spontaneous resorption of bone/progressive atrophy of bone/progressive osteolysis/angiomas/haemangiomas/lymphangiectasis

In this disease there is resorption of bone which usually commences as a localised lesion, but which may progress to complete destruction, and this appears to be the main distinguishing feature of osteolysis

Points in favour:

- ❖ young adults of either sex
- ❖ laboratory investigation are normal
- ❖ most consistent h/p finding is the presence of an angiomatous tissue proliferation that replaces bone.

- **Osteogenesis imperfecta (brittle bone disease)**

points in favour: extreme fragility & porosity of bone proness to fracture deafness

points against: at birth blue sclera

- **Osteopetrosis**

points in favor: multiple pathologic fracture increased chances of infection of bone

points against: increased ALP, calcium, phosphorous increased acid phosphatase

- **Osteoporosis**

points against:

- ❖ **elderly females**

large degree of **maxillary atrophy** because BMC(bone mineral content) loss in the trabecular bone is relatively larger than in the cortical bone (large surface area/volume in the trabecular bone) osteolysis associated with infection/rheumatoid arthritis pre existing periodontal disease.

CONCLUSION

A thorough workup including relevant investigation should be undertaken prior to planning a treatment and executing the plan on a priority basis or else the patient may not have a lower jaw to chew on.

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