

Vanishing Mandible: Gorhamstout the Culprit

Kishan Ashok Bhagwat¹, Vikram M. Patil², Siva bharani³, Pruthvi Maliki⁴

¹Associate Professor, ^{2,4}Post graduate residents, ³Professor.

^{1,2,4}Department of Radiology, SSIMS & RC, Davangere, Karnataka, India.

³Department of oral surgery, College of dental sciences, Davangere, Karnataka, India.

Abstract: Gorham's stout disease is rare skeletal condition presenting with massive osteolysis of the involved bone due to intraosseous vasculolymphatic tissue. This entity is rather misdiagnosed as aggressive lytic lesion, which results in patient being subjected to painful biopsy procedures. However this entity is seen to progress rapidly after any form of traumatic procedures according to some literature. Trauma is one of the important events that precedes this condition, However few cases had no antecedent trauma and were considered idiopathic. We report a 19 year old male who presented with left sided facial swelling with no antecedent trauma. Clinical features were not directive to any specific diagnosis. Various laboratory and radiological investigations were performed. Imaging was rather nonspecific other than mandibular osteolysis with no periosteal reaction or soft tissue calcification, but our experience with previous such entity, negative laboratory findings & retrospective clinical correlation, helped us diagnose this entity. Imaging was followed by biopsy (on clinicians demand), which confirmed our interpretation of facioskeletalgorham's.

I. Introduction:

Gorham's disease, also known as massive osteolysis, is a rare disorder characterized by spontaneous and progressive osteolysis of one or more skeletal bones.¹ Jackson² was the first person to describe a case of vanishing bone disease in 1838 while Romer³ was the first to report a case in the jaws of a 31-year-old female in 1924. It was Gorham⁴ in 1954 and Gorham and Stout¹ in 1955 who presented a case series and defined the condition as a specific pathological process. To date, approximately 200 cases have been described in the literature, with 41 cases involving the maxillofacial region, especially the mandible.⁵

This entity is characterized by proliferation of vascular or lymphatic channels in an uncontrollable manner within the bone, leading to resorption of bone with subsequent replacement with fibrous tissue. This is basically an aggressive entity of unknown etiology seen in many flat bones, that causes massive osteolysis in an unpredictable manner, thus also called vanishing bone disease. We report a rare case of facioskeletal Gorham's disease involving mandible.

II. Case Report

We report a case of 19 year old male with 3 months prior history of left sided facial swelling. The patient complained of transient regression of the swelling. The swelling appeared to increase slowly, with slow but significant progression of the swelling. However little tenderness could be elicited at the swollen area.

Routine investigations like complete blood count, complete metabolic panel, alkaline phosphatase, C-reactive protein, erythrocyte sedimentation rate turned negative. This ruled out infectious, endocrine, vascular disorder.

Further evaluation with OPG showed large lytic lesion involving the left ramus of mandible with thinned out cortex, with adjacent soft tissue opacity. Further evaluation with CT scan revealed osteolysis in the mandible at the body, ramus and coronoid process on the left side with cortical thinning. Post contrast arterial and venous phases show insignificant enhancement. No soft tissue calcification or periosteal reaction was seen. Additionally, screening with MRI did not reveal diffusion restriction ruling out infective/inflammatory/neoplastic lesion. USG image revealed abnormal soft tissue with minimal vascularity.

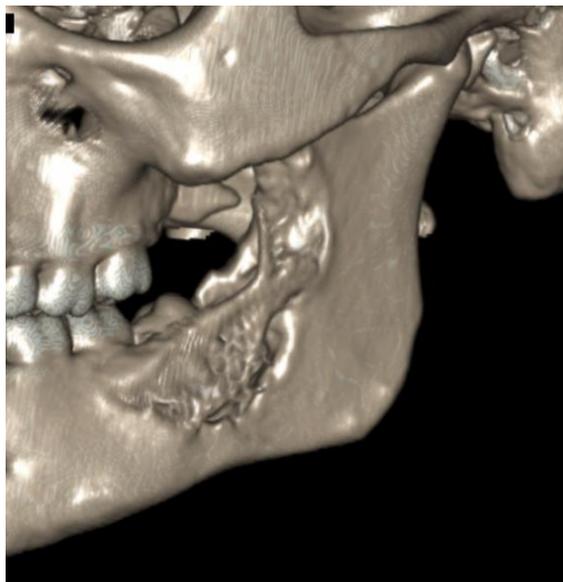
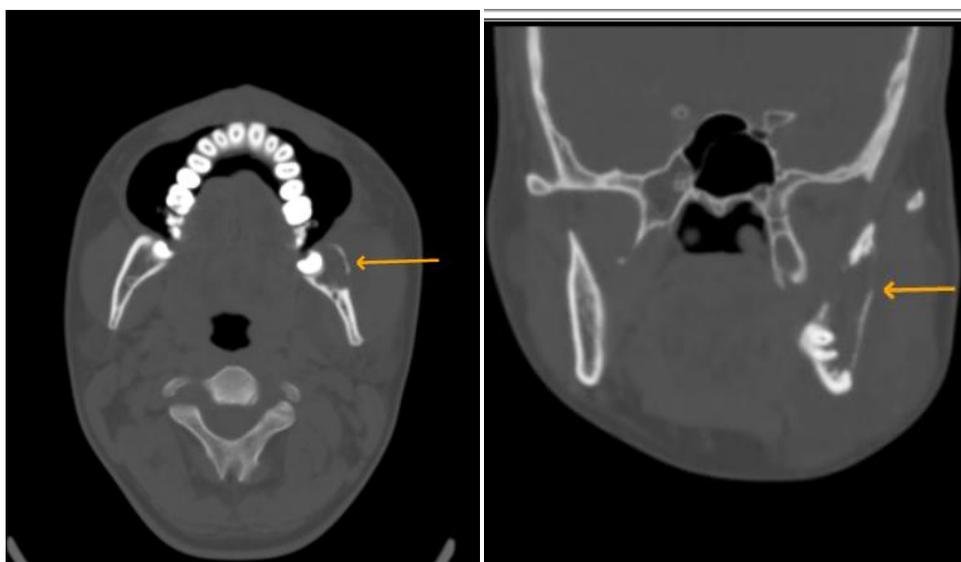


Figure1. Reformatted SSD image of the eroded mandible



Figure2. Erosive change seen along anterior margin of left ramus of mandible and in retromolar region.



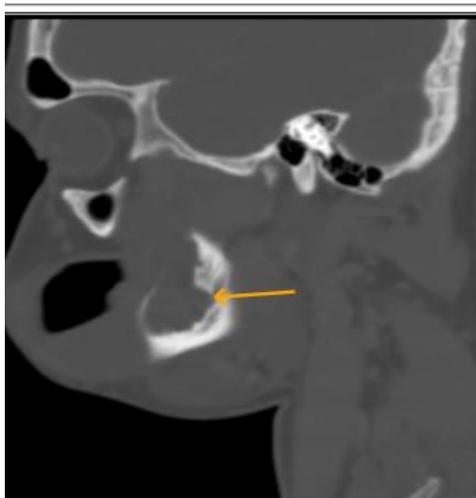


Figure 3. Axial, reformatted coronal (upper right) and sagittal CT (lower left) sections in bone window, showing destructive lesion along anterior margin of left ramus of mandible and in retromolar region.

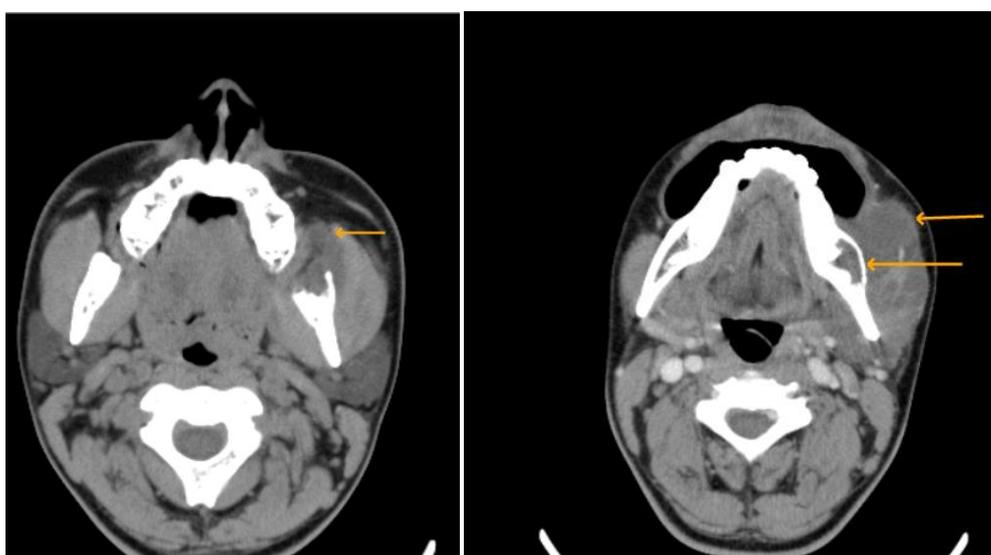


Figure 4. Axial Plain (left) and contrast CT (right) scan images revealed destructive changes along the anterior margin of left ramus of the mandible with insignificant enhancing soft tissue in the left cheek/submandibular region.

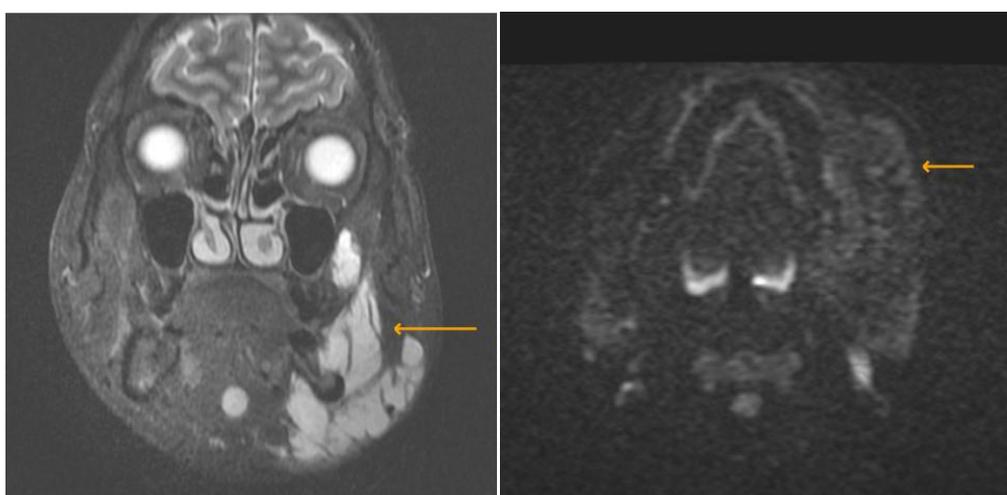


Figure 5. Coronal STIR (left) and Axial DWI (right) showing proliferative inflammatory tissue and absent restriction respectively.

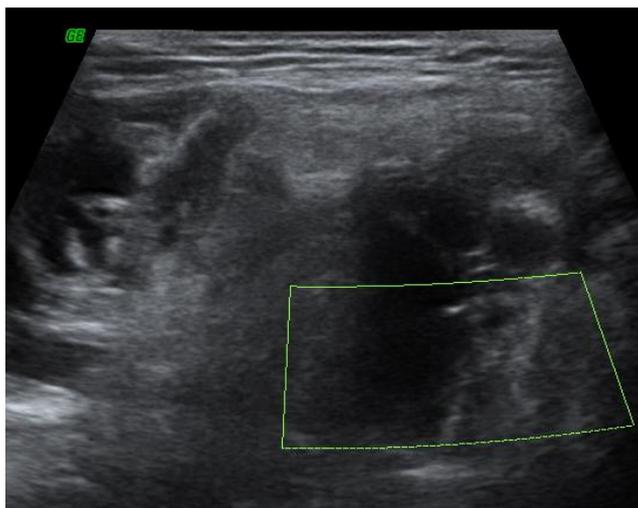


Figure 6: USG revealed abnormal soft tissue with minimal vascularity.

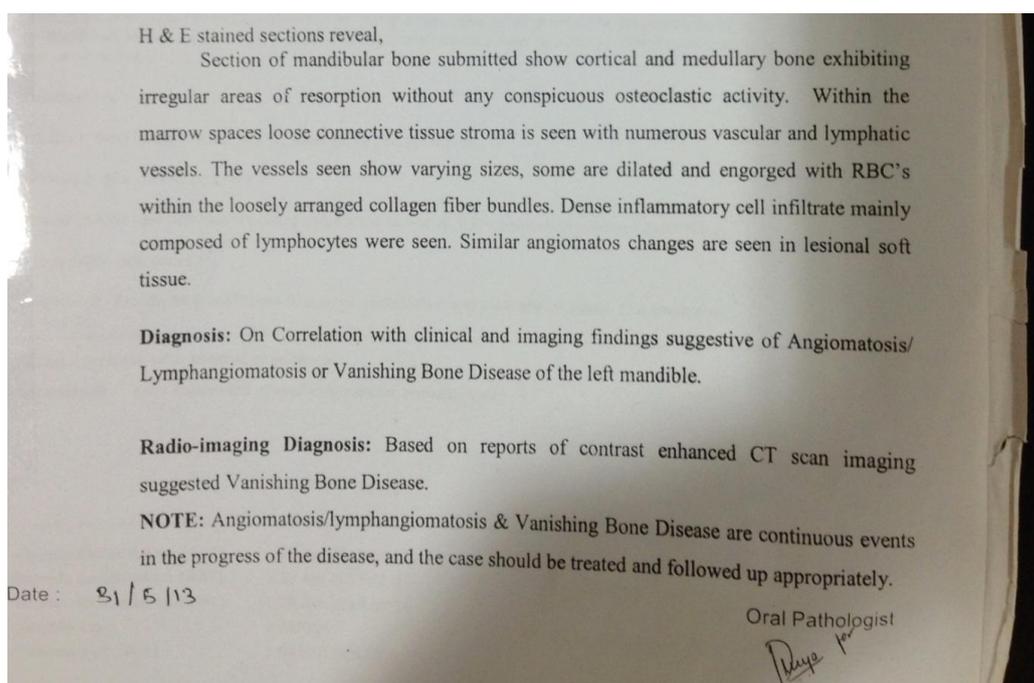


Figure 7. Biopsy report

Following imaging, biopsy was performed. Biopsy revealed high concentration of thin walled vascular channels with some active fibroblast and chronic inflammatory cell infiltration. There was no evidence of any malignant cells.

III. Discussion & Conclusion:

Disappearing bone disease or Gorham's disease is a rare disorder characterized by osteolysis with associated proliferation of vascular or lymphatic channels within bone and the surrounding soft tissues⁶. These cases usually are within age group of 40yrs⁷. The etiology of Gorham's disease remains largely unclear. However Gorham and Stout attributed the etiology to uncontrolled proliferation of vascular and lymphatic tissue with local P^h changes that causes bone resorption with subsequent replacement by fibrotic tissue⁷. The disease may affect any bone, however its predilection to affect mandible or skull is more classical and makes diagnosis relatively easy and regular follow up should suffice as the disease may regress in some patients⁸.

Treatment involves complete resection and reconstruction of hemimandible. Other treatment options include Radiation therapy, bisphosphonate therapy, sclerotherapy, percutaneous bone cement, bone graft, prosthesis, surgical stabilization and amputation^{9,10,11,12}.

Knowledge of this entity is essential for correct diagnosis as imaging plays a key role in evaluation of such rare entities, thus helping further management and timely intervention if needed.

References

- [1]. Gorham L, Stout A. Massive osteolysis (acute spontaneous absorption of bone, disappearing bone): its relation to hemangiomas. *J Bone Joint Surg Am* 1955;37:985–1004
- [2]. Jackson JBS. A boneless arm. *Boston Med Surg J* 1838;10:368–369
- [3]. Romer O. Die Pathologie der Zahne. In: von Henke F, Lubarsch O, editors. *Handbook of pathology, anatomy and histology*. Vol 4 Berlin, Germany: Springer Verlag; 1924. pp 135–499
- [4]. Gorham LW, Wright AW, Shultz HH, Maxon FC. Disappearing bones. A rare form of massive osteolysis: report of two cases, one with autopsy findings. *Am J Med* 1954;17:674–682
- [5]. Escande C, Schouman T, Françoise G, Haroche J, Ménard P, Piette JC, et al. Histological features and management of a mandibular Gorham disease: a case report and review of maxillofacial cases in the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2008;106:e30–e37
- [6]. L. W. Gorham and A. P. Stout. "Massive osteolysis (acute spontaneous absorption of bone, phantom bone, disappearing bone): its relation to hemangiomas," *Journal of Bone and Joint Surgery. American*, vol. 37, pp. 985–1004, 1955.
- [7]. D. V. Patel, "Gorham's disease or massive osteolysis," *Clinical Medicine & Research*, vol. 3, no. 2, pp. 65–74, 2005.
- [8]. SJ Reddy and DS Jatti. Gorham's disease: a report of a case with mandibular involvement in a 10-year follow-up study. *Dentomaxillofac Radiol*. Sep 2012; 41(6): 520–524.
- [9]. Branco F, Da Horta JS. Notes on a rare case of essential osteolysis. *J Bone Joint Surg Br* 1958;40:519–527
- [10]. Mendez AA, Keret D, Robertson W, MacEwen GD. Massive osteolysis of the femur (Gorham's disease): a case report and review of the literature. *J Pediatr Orthop* 1989;9:604–608
- [11]. Ohya T, Shibata S, Takeda Y. Massive osteolysis of maxillofacial bones: report of two cases. *Oral Surg Oral Med Oral Pathol* 1990;70:698–703
- [12]. Booth DF, Burke CH. Massive osteolysis of the mandible: an attempt at reconstruction. *J Oral Surg* 1974;32:787–791