**ABSTRACT**

**Objectives:** To present a rare pediatric case of Gorham’s disease with extensive head and neck involvement.

**Study Design:** Case report and literature review

**Methods:** Literature review of Gorham’s disease in pediatric patients with head and neck manifestations with discussion of a representative case within our health system.

**Results:** We describe a rare and fatal pediatric case of Gorham’s disease originating in the mandible, and progressing over 5 years to involve the maxilla, sphenoid bone, temporal bone, occipital bone, and infratemporal fossa. A nine year old male initially presented with presumed mandibular trauma. An impressively progressive massive maxillofacial osteolysis ensued. The diagnosis of Gorham’s disease was established only after infectious, malignant, inflammatory and endocrinologic diseases were excluded. Facial photographs and computerized tomographic images demonstrate the dysmorphic facial features of this vanishing bone disease. The literature is reviewed and medical and surgical management is discussed. Gorham’s disease in a pediatric patient with extensive head and neck involvement is exceptionally rare.

**Conclusion:** To the best of our knowledge, since 1928, there have only been three other cases of Gorham’s disease in the literature involving the mandible of a male patient younger than ten years of age. No regeneration has ever been observed following the osteolysis. GD seems to progress through two phases. The first phase consists of massive osteolysis. During this quiescent phase, the haversian structure of the bone undergoes fibrosis. The duration of these stages is unpredictable and may last months to years. The lack of new bone formation is a trademark of GD. The diagnosis of GD is based on clinical, histologic, and molecular features. The etiology of the massive osteolysis is still unknown. The lack of a standard treatment regimen for patients suffering from GD correlates with its widely disputed pathogenesis. The range of treatment consists of either surgery and radiation therapy, or medical therapy. The latter therapy is more directed for the lack of a strong correlation with disease remission in affected patients. Early irradiation of the affected region has been observed to induce remission in some patients. GD seems to progress through two phases. Clinical and pathologic features are critical as these patients may require surgical stabilization. Proper protection such as helmets may be needed. Medical therapy has been used in refractory cases and consists of bisphosphonates, calciotin, calcium, vitamin D, and alpha-2b interferon therapy. Surgical treatment of disease involving the skull base and maxillofacial regions may not be an option given the potential post-surgical morbidity. According to Escande, treatment of GD in the maxillofacial location is local resection of the involved bone in an attempt to stop progression of the disease. Although GD is an insidious progressive disease, spontaneous regression has been described. Involvement of the cervical and thoracic spine resulting in persistent chylothorax is the usual cause of death in GD patients. Due to the rarity of pediatric GD, there is limited information on specific treatment options in pediatric patients.

**REFERENCES**