Fatal Progression of Gorham Disease: A Case Report and Review of the Literature

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Gorham disease is an idiopathic massive osteolysis, and maxillofacial involvement is rare. This report describes a case of a 12-year-old boy with severe progressive osteolysis in the mandible, hyoid bone, mastoid process, and cervical spine. Radiation therapy and interferon-α therapy were followed by bisphosphonate therapy. The patient died of respiratory failure. To describe the clinicopathologic features of Gorham disease of the jaws with an emphasis on the fatal types, 64 cases in the literature were reviewed (female-to-male ratio, 1:1.78; average age, 33.02 $\pm$ 19.38 yr). Most lesions were located only in the mandible or in other locations in combination with the mandible, except for 3 cases. During follow-up, there were 7 cases of disease-specific death, resulting in a mortality rate of 10.94%. The main causes of death were chylothorax, rib fractures secondary to osteolysis, or spinal fractures. Although most patients received surgical treatment (43.75%), the type of treatment was not related to prognosis.

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Massive osteolysis usually occurs in young patients and was reported in detail by Gorham and Stout.[1] It is characterized by painless progressive bone loss and sometimes is associated with trauma.[2] Its pathogenesis is largely unknown, and it mostly affects the long bones of the extremities.[3] The progression of this disease is unpredictable, but some cases regress spontaneously. Fatal progression of this disease is rather high (19.5%).[4] This article reports on a patient with fatal progression of Gorham disease.

Report of Case

A 12-year-old boy was seen on September 1, 2011 for persistent pain in his chin after being injured by a punch 1 week previously. He also said that his ability to open his mouth was limited. Otherwise, he was well. On examination, the teeth were mobile in the mandible and there was submental swelling. An initial clinical diagnosis of submental contusion was made. Computed tomography of the jaw depicted slight buccal cortex erosion involving the anterior mandibular area (Fig 1). The serum alkaline phosphatase level was abnormally increased (728 IU/L), and the serum phosphorus level was slightly elevated (5.1 mg/dL). However, the hematologic examination for hemoglobin, red blood cell indices, differential white blood cell count, serum calcium, glucose, urea, and creatinine were normal. In regard to the endocrine system, levels of growth hormone, thyroid-stimulating hormone, free thyroxin, parathyroid hormone, estradiol, and insulin-like growth factor were within normal limits. Surgical biopsy examination of the anterior mandibular area was performed (Fig 2). Grossly, the inside of the mandible was mostly emptied. The buccal cortex was thinned, resembling an eggshell, and loose connective

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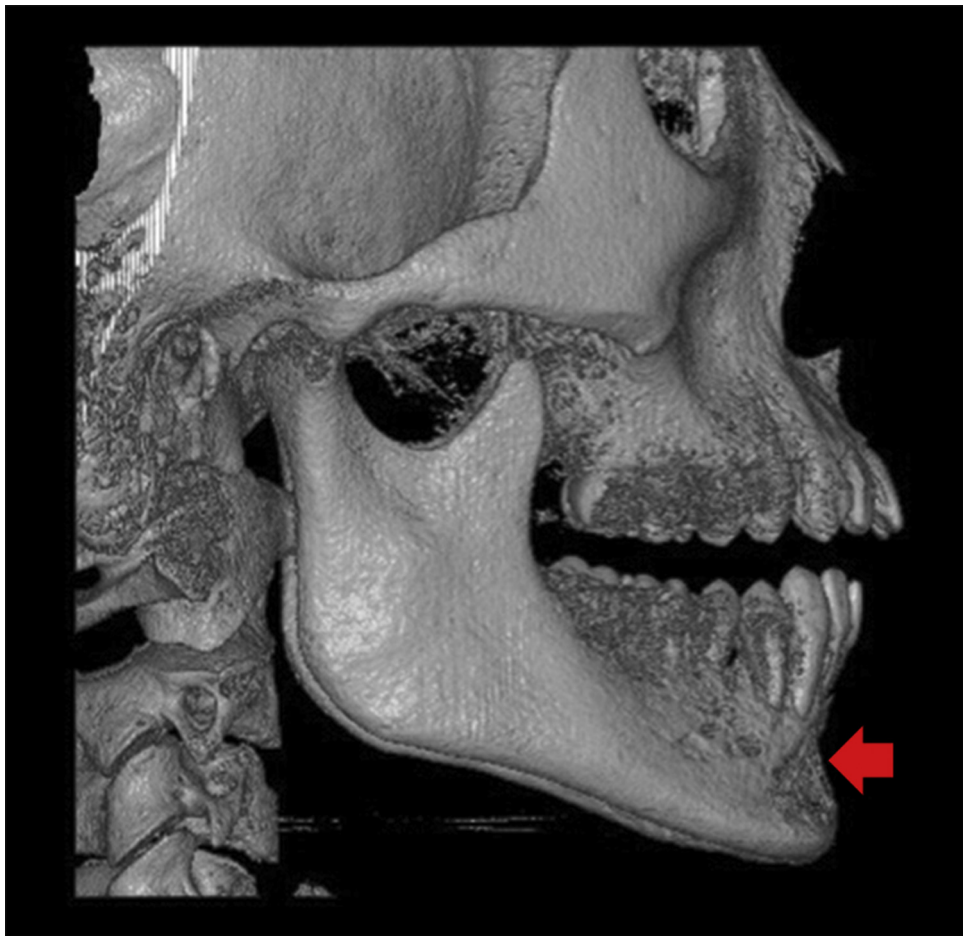

FIGURE 1. Computed tomogram obtained on September 1, 2011 depicts slight buccal cortex erosion involving the anterior mandibular area (*arrow*).

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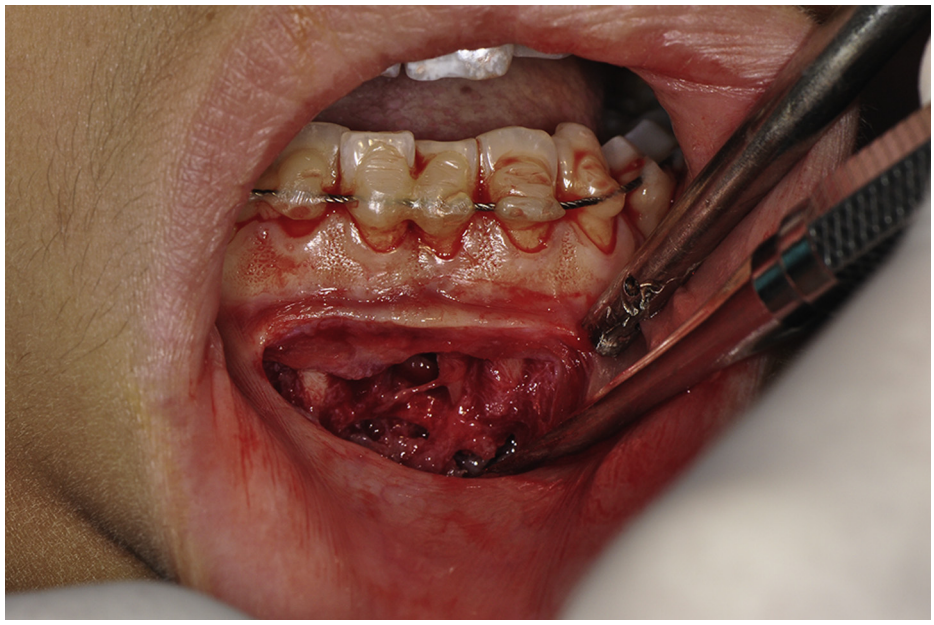

FIGURE 2. Surgical biopsy examination of the anterior mandibular area was performed. The inside of the mandible was mostly emptied.

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