Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology 28 (2016) 147–151

Contents lists available at ScienceDirect

Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: www.elsevier.com/locate/jomsmp

Case Report

Gorham-Stout disease: Progressive massive osteolysis of the mandible

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ARTICLE INFO

Article history:
Received 4 September 2014
Received in revised form 4 September 2015
Accepted 24 September 2015
Available online 1 November 2015

Keywords:
Massive osteolysis
Lymphangioma
Gorham-Stout disease

ABSTRACT

Gorham-Stout disease (GSD) is an extremely rare massive osteolytic bone disease, and the pathogenesis and etiology are unknown. It is characterized by the uncontrolled proliferation of distended, thin-walled vascular and lymphatic channels within bone, which leads to resorption and replacement of the bone with angiomas and/or fibrosis.

A 35-year-old man had complained of masticatory disturbance caused by mandibular teeth mobility and mandibular swelling for several years. He suffered from an eating disorder because of the severe mobility of his mandibular teeth, and he presented at our hospital for consultation. He had swelling and grade III mobility without pain; his skin was normal in color with a 70 mm × 50 mm lesion of elastic and firm texture at the submental region. The ill-defined mandibular bone destruction was seen radiographically. Magnetic resonance imaging (MRI) showed that the lesion occupied the submental region and inside the mandibular bone, and these lesions were isolated. The biopsy results suggested a diagnosis of lymphangioma. The patient received radical surgery, and histological examination suggested lymphangioma. We diagnosed GSD because massive osteolysis of the mandible associated with lymphangioma was seen in the lesion. The postoperative course was uneventful, with no local recurrence 36 months after surgery. Given the rarity of this disease entity, there is no standard therapy. The details of this case are presented, and diagnostic and management considerations are described.

1. Introduction

Lymphangiomas are benign tumors that grow proportionally with patients' body growth by the proliferation of lymphatic vessels. The incidence of lymphangioma is estimated to amount to 6% of all benign tumors in children, but lymphangioma in adults is very rare. In the head and neck region, lymphangioma appears most commonly on the tongue, buccal mucosa, lips, and neck [1].

Mandibular osteolysis is usually the result of a well-defined pathogenic process. Usually the destruction is associated with some underlying disease, such as periodontal or periapical infection, osteomyelitis, or odontogenic cyst, or tumor.

Gorham-Stout disease (GSD) is an extremely rare massive osteolytic bone disease, but the pathogenesis and etiology are unknown. The lesion typically occurs in the shoulder, skull, pelvic girdle, jaw, ribs, or spine. It is characterized by the uncontrolled proliferation of distended, thin-walled vascular and lymphatic vessels within bone, which leads to resorption and replacement of the bone with angiomas and/or fibrosis. Treatment of the massive osteolysis is, for the most part, palliative and limited to symptom management [2–4].

Here, we describe a case of a massive osteolysis in the mandible and severely mobile teeth with lymphangioma. We performed radical surgery for this lesion. The lesion was finally diagnosed as GSD. The etiology, diagnosis, and management considerations of GSD are described in this literature.

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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http://dx.doi.org/10.1016/j.ajoms.2015.09.007

2. Case report

A 35-year-old man presented at our department with masticatory disturbance caused by mandibular teeth mobility and mandibular swelling for several years. He had been injured in a motorbike accident with laceration at the mental region, and he underwent debridement and suture treatment 15 years ago. Extraoral examination revealed swelling at the submental region, which was 70 mm × 50 mm in diameter, painless, non-tender, elastic, and non-mobile (Fig. 1A). The lesion contained serous liquid, and blood oozed from part of the left lower molar when the mass was compressed. Intraoral examination revealed significant periodontal bleeding, increased probing depth, remarkable teeth mobility, and gingival crevicular fluid (Fig. 1B).

Panoramic radiography revealed horizontal alveolar bone loss at the mandible (Fig. 2). Magnetic resonance imaging (MRI) showed that the lesion occupied the submental region and inside the mandibular bone, and these lesions were isolated. Contrast-enhanced MRI of the mandibular alveolar bone region showed multiple high signals in the lesion (Fig. 3A), and the submental region showed swelling deep in the platysma muscle and an unclear accumulation image (Fig. 3B). 99mTc scintigraphy showed accumulation in the right mandibular bone, the right mandibular condyle, and the left temporal bone (Fig. 4), but gallium scintigraphy did not show accumulation. There were no abnormal findings in any other regions.

A needle biopsy was performed, and the specimen was taken from the submental lesion. Histopathological diagnosis suggested lymphangioma. We performed a radical surgery for the submental lesion and alveolar lesion that includes extraction of all lower teeth under general anesthesia. The submental lesion was resected with submental approach, and the alveolar lesion was resected with intra-oral approach. Intraoperative findings showed that the tumor

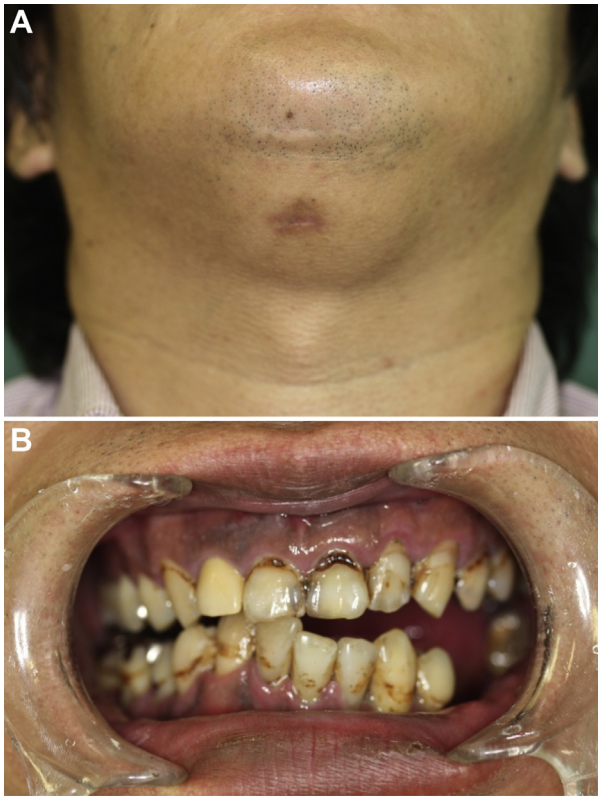

Fig. 1. (A) Extraoral examination revealed swelling of the submental region. (B) Intraoral examination revealed significant periodontal bleeding, increased probing depth, remarkable teeth mobility, and gingival crevicular fluid.

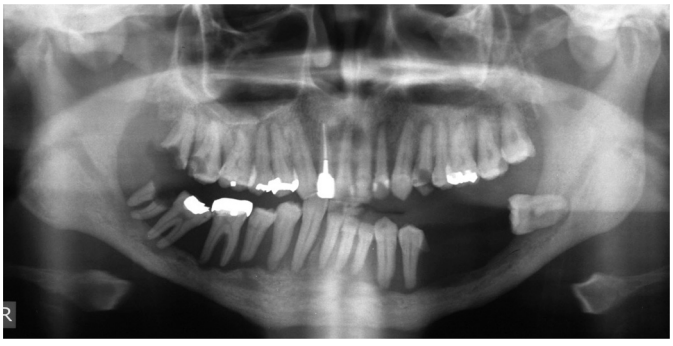

Fig. 2. Panoramic radiography shows resorption and decreased vertical height of the mandibular body with resorption extending toward the basal bone.

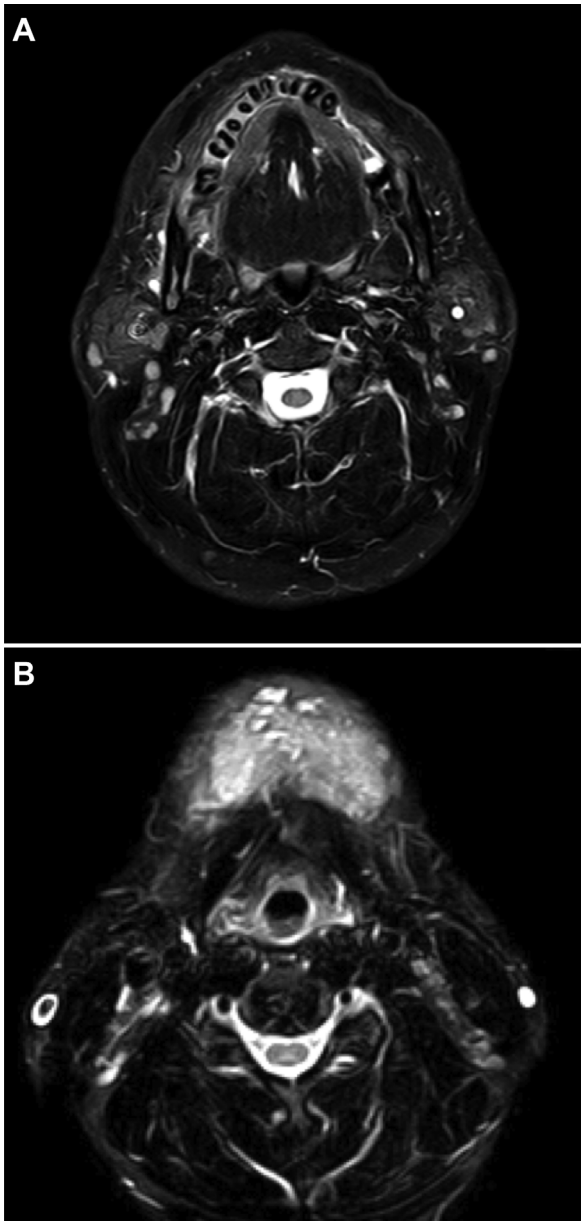

Fig. 3. Magnetic resonance imaging (MRI). (A) Mandibular alveolar region showed multiple high signals in the lesion. (B) Submental region showed multiple high signals in the lesion; its border was indistinct under the mentum.