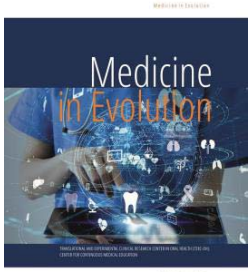
Giant cell granuloma of the floor of the mouth in a patient with glycogen storage disease type Ib. Case report



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Abstract

Central giant cell granuloma (central giant cell granuloma) is an uncommon benign bony lesion that occurs in the mandible and maxilla and accounts for approximately 7% of all benign tumours of the jaws. Central giant cell granuloma occurs predominantly in children or young adults, with approximately 75%of cases presenting before 30 years of age although presentation can occur at any age. The majority of these lesions were asymptomatic and relatively innocuous. However, some displayed a more aggressive clinical course characterized by root resorption, pain or paresthesia, and cortical perforation. Females are affected more frequently than males, with a ratio of 2:1.

Keywords: Central giant cell granuloma, bony lesion, mandible, tongue, glycogen storage disease

INTRODUCTION

Giant cell-rich lesions of bone represent a heterogeneous group of multinucleated giant cell proliferation of osteoclastic type. In the maxillomandibular region, some are commonly found, such as central or peripheral giant cell granulomas, cherubism, aneurysmal bone cyst, and brown hyperparathyroidism tumor, while others are more rarely found, such as giant cell tumors of the bone and giant cell tenosynovial tumor of the temporomandibular joint [1,2]. Giant cell tumor of the bone was described in 1818 by Cooper and Travers and is listed in two forms in the general classification of bone and soft tissue tumors that was modified in 2020: the bone form classified as osteoclastic giant cell-rich tumors, malignant or of intermediate malignancy (locally aggressive, rarely metastatic), and the form in "soft tissue" classified as so-called fibrohistiocytic tumors of intermediate malignancy [3,4]. Bone giant cell tumors are either benign or malignant primary tumors. In the Glycogen Storage Disease (GSD) type 1b, the enzyme glucose-6-phosphatase (G6P) cannot be transported across the microsomal membrane in the liver, and the glycogen cannot be metabolized [5]. The glycogen accumulates in several organs, such as liver and kidneys [6,7], and the disease leads to systemic and intraoral manifestations. The main systemic manifestations are hypoglycemia, hyperlipidemia, hyperuricemia, hepatomegaly, short stature, growth retardation, neutropenia, and neutrophilic dysfunction with recurrent infections [8, 9]. Patients with GSD 1b show poor prognosis and high mortality rate [10]. The incidence of glycogen storage disease type I (both GSD Ia and GSD Ib) is 1 in 100,000 live births. Approximately 20% of glycogen storage disease type I is GSD Ib.

MATERIAL AND METHOD

We present a case of a 19-year-old patient known with a glycogen storage disease type I b. He presented to the Emergency County Hospital Bihor with difficulty breathing, feeding and sialorrhea. Intraorally there was a large hyperplasic purplish lesion on the floor of the mandible (Figure 1) measuring approximately 6,5 cm long and 4 cm width, asymptomatic with no numbness of the area, lower lip and mental area.

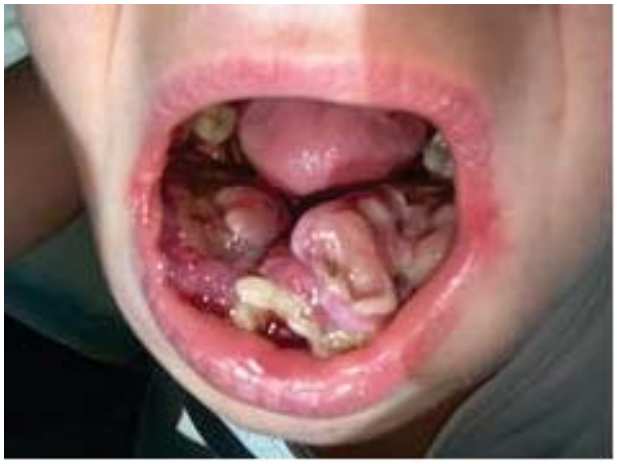

Figure 1. Clinical aspect: polylobulated parenchymal and osteolytic lesion

The CT examination demonstrated parenchymal and osteolytic polylobulated lesion centered on the body of mandible and oral floor, with differential iodophilia and expansion in

the subcutaneous soft tissue and perilingual groove. It causes osteolysis in the superior one third of the body of the mandible and alveolar bone (Figure 2).

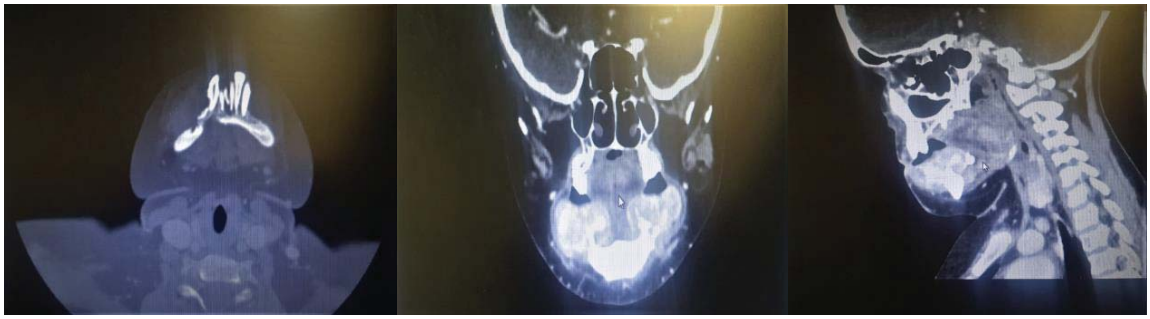

Figure 2. Lesion centered on the body of the mandible and the floor of the mouth; osteolysis of the upper third of the body of the mandible and the alveolar process

The patient understands the necessity of surgery, is informed about the steps involving the therapy and prognosis. The complete excision of the tumor is performed under general anesthesia (Figure 3).

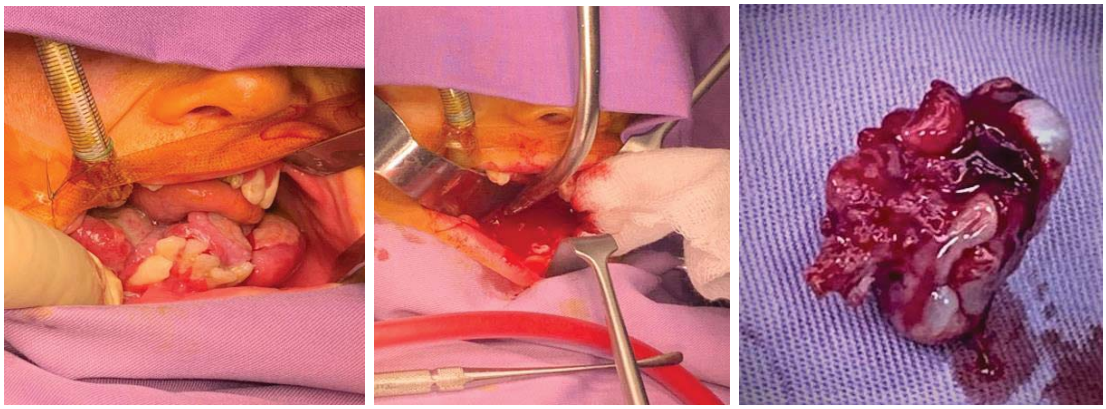

Figure 3. Excision of the tumor and extraction of the frontal teeth

After performing the surgery under general anesthesia, the excised tissue was taken for histopathology examination (Figure 4).

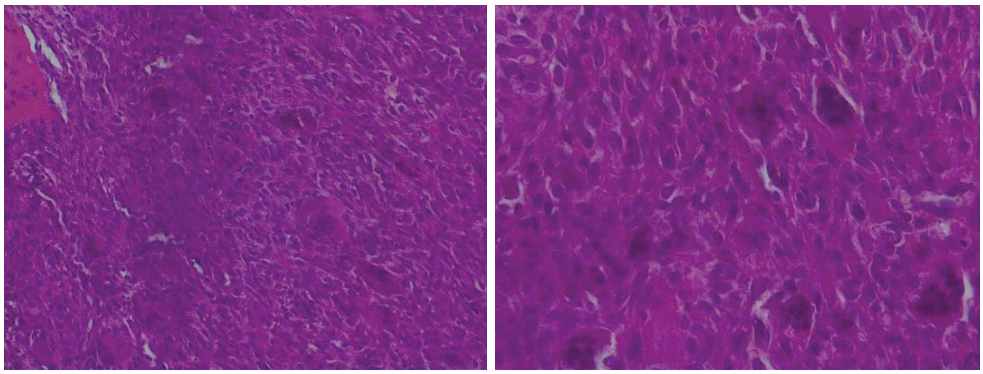

Figure 4. Multinucleated giant cells within a background of plump proliferating mesenchymal cells with extensive red blood cell extravasation

RESULTS

Post operative the patient had a good healing. Pain and discomfort after the procedure were managed with medication; the patient was given antibiotics, anti-inflammatory, and analgesic drugs. Mentally he was optimistic and pleased about the result. He tolerated well the soft diet for seven days and maintained a good oral hygiene. The sutures were removed after ten days.

The postoperative evolution was good, the patient being periodically called for control (Figure 5).

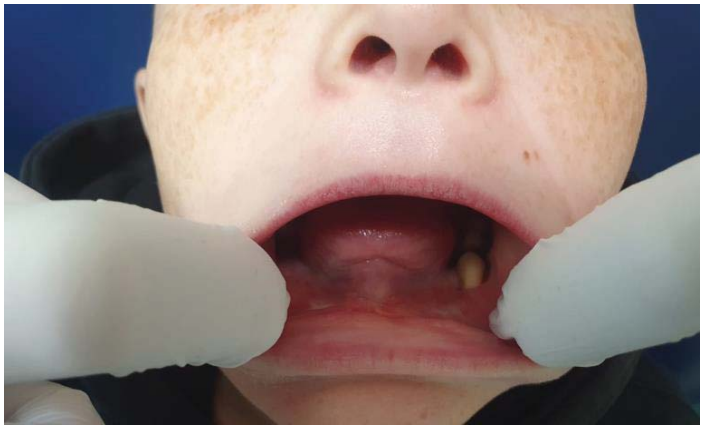

Figure 5. The clinical situation one year postoperatively

DISCUSSIONS

The frequent intake of carbohydrates, to support blood glucose levels, provides a substrate for oral cariogenic bacteria by implementing the risk of developing caries. Furthermore, the generalized growth retardation of these subjects could also explain the eruption delay of the dental elements [11,12]. GSD I b is a variant of GSD I a; it is due to the defect of the glucose-6-phosphate transporter and presents further manifestations, such as neutropenia, altered neutrophil migration and bactericidal activity [13]. Patients are more susceptible to oral ulcers, periodontitis and oral cavity infections [13,14].

Current management of giant cell granulomas is based on surgical resection combined with supported curettage or peripheral osteotomy, to reduce the risk of recurrence [15]. For aggressive lesions, an "en bloc "resection may be considered [16]. Several pharmacological treatments are described as effective alternatives to the surgical management of giant cell granulomas. These are essentially subcutaneous injection of calcitonin or alpha interferon and intra-lesional injection of corticosteroids, although they have significant drawbacks (long duration of treatment, need for additional surgery in case of ineffectiveness and side effects [17,18].

CONCLUSIONS

The diagnosis of giant cell granuloma of the floor of the mouth is essential for a good outcome. Communication between dentist, oral surgeon, geneticist and nephrologist is essential for the benefit of the patient. Oral hygiene is very important and needs serious attention from the patient because patients with glycogen disease type Ib are more susceptible to oral ulcers, periodontitis and oral cavity infections.

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