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Giant Odontogenic Myxoma of the Maxillary: A Rare Entity

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Abstract **Review Article**

***Introduction*:** Maxillary odontogenic myxoma is a locally invasive benign mesenchymal tumor. The objective of this case observation was to discuss the difficulties related to the management of a case of giant maxillary odontogenic myxoma. ***Observation*:** This was a 32-year-old patient who consulted in the stomatology and maxillofacial surgery department of the CHU-CNOS Pr HT of Bamako for a swelling of the left hemiface evolving for 3 years with deterioration of the general condition. She has no particular medical or surgical history. On physical examination, there was a large swelling of the left hemiface measuring 35/21cm, hard and painless to palpation. There was also exophthalmos of the left eye, filling of the left nasal fossa, left suborbital hypoesthesia, and ipsilateral submandibular adenopathy. A maxillofacial CT scan and operability assessment were performed. After intense renutrition, surgical excision was performed under general anesthesia with a primary tracheotomy. Histological examination of the surgical specimen concluded that it was an odontogenic myxoma. The outcome was favorable, there was no recurrence after 12 months. ***Conclusion*:** Odontogenic myxoma is a benign tumor with slow progression and can be locally invasive, making surgical management difficult.

Keywords: Odontogenic myxoma, Giant, Maxilla, Bamako.

INTRODUCTION

Odontogenic myxoma of the maxilla was initially described by Thoma and Goldman in 1947 [1]. It is a benign and rare mesenchymal tumor [2]. It represents 3 to 7% of odontogenic tumors and 0.41% of bone tumors [7]. This tumor more frequently affects young adults between the second and fourth decade of life in women [2, 3].

Although histologically benign, it presents a very aggressive locoregional malignant behavior with invasion of adjacent tissues requiring appropriate management [2,1]. Its diagnosis is therefore not easy and it must be differentiated from malignant tumors such as sarcoma and other benign tumors such as ameloblastoma [2, 3]. Advanced forms called giant or "historical" are described in the literature [4, 3].

The objective of this study was to report a case of giant myxoma by describing its characteristics, and comparing it to cases reported in the literature.

This was a 32-year-old patient who had consulted for a painless swelling of the left hemiface that had been developing for 3 years. She had no contributory medical or surgical history.

On admission, she had a deterioration in her general condition (WHO score = 3) with malnutrition folds.

On exo-oral examination, a large swelling of the left hemiface exceeding the midline was noted (Figure 1). The swelling is covered with tight skin, with an “orange peel” appearance, measuring 35/21 cm in the major axis. It was fixed, painless, firm, and integral with the maxilla on the left. Grade II left exophthalmos, left nasal cavity filling and left suborbital hypoesthesia were also noted with a firm, mobile and painless 1cm

adenopathy in sector Ib of the homolateral type. On intraoral examination, filling of the upper vestibular was noted due to swelling with exposure of the oral mucosa. The swelling was covered by healthy mucosa with venous laci. The floor of the mouth and tongue were free. Dental signs such as tooth loss, tooth displacement and tooth mobility were present in quadrant II. Maxillofacial CT scan revealed a large necrotic and osteolytic mass of the left maxilla (Figure 2).

The treatment began with intense refeeding, then a biopsy was performed followed by histopathological examination which revealed a myxoma. The surgical procedure performed under general anesthesia after a first tracheotomy, consisted of a total left maxillectomy via the Weber Ferguson approach, corresponding to a Cordeiro III type left maxillary substance loss (Figure 3, 4, 5). The reconstruction of the vestibulopalatine mucosa substance loss was done by left jugal flap and Bichat ball. The postoperative course was simple. The evolution was favorable (Figure 6). There was no recurrence after 2 years.

Figure 1: Image at admission

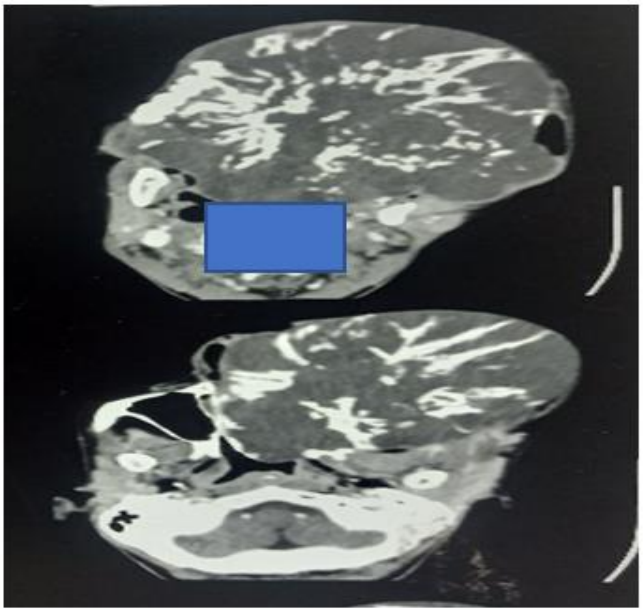

Figure 2: Scanographic appearance of the tumor

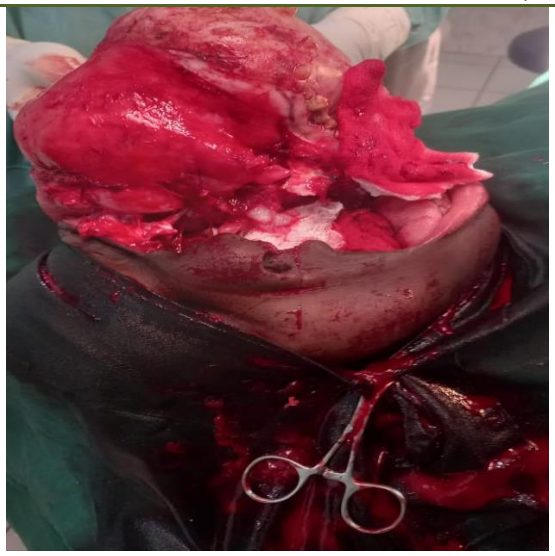

Figure 3: Per-operative appearance

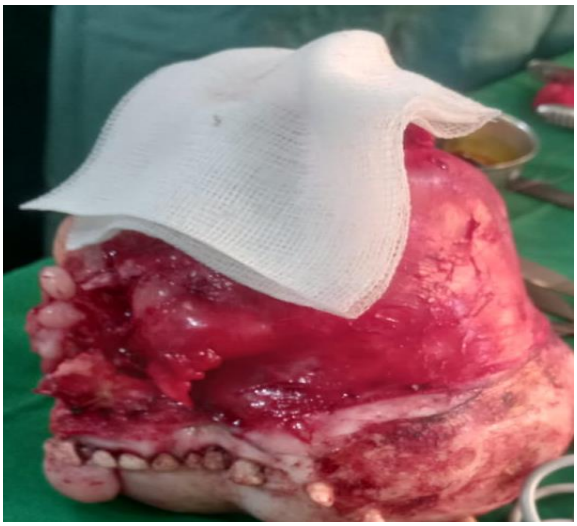

Figure 4: Operating room

Figure 5: Appearance after excision

Figure 6: Post-operative appearance

DISCUSSION

Although rare, myxoma is a benign odontogenic tumor that ranks second among benign odontogenic tumors [5-7]. At the cervicofacial level, two forms are described: Bone myxoma, it derives from the mandible, maxilla, nasal bone and rarely from the petrous bone. As for soft tissue myxoma, it derives from the parapharyngeal tissues, parotid and larynx. Bone myxoma affects the mandible more than the maxilla [5, 8, 9]. Studies of isolated cases with maxillary localization are frequent [2, 10, 11, 8]. Most studies report a female predominance [12, 13, 1]. However, many authors report isolated cases in men [3, 11, 7]. This tumor more frequently affects young adults between the second and fourth decade [2, 13, 12]. However, isolated cases in children have been described [3, 8, 14]. The symptomatology is dominated by swelling [13, 3, 14, 1]. The swelling can be significant with difficulties in feeding and speaking as in the present case [3, 15, 1]. The swelling is generally accompanied by pain, hypoesthesia, cervical adenopathy, dental mobility, dental displacement or even dental avulsion [3, 14]. In the reported case, he had an altered general condition listed as WHO3 which would be due to the large oral swelling preventing feeding. The average time between the first symptoms and treatment varies between 1 and 5 years, reflecting its slow growth [13, 1]. However, cases of short-term evolution have been described << less than a year >> [8, 15, 9]. The classic radiological appearance is that of a clear polygeodic radiograph [9]. However, the appearance of a monogeodic cystic lesion is possible. The myxoma can sometimes invade the cortices and soft tissues requiring a differential diagnosis with a malignant process [9,3]. A biopsy is thus performed before surgery to confirm the diagnosis [3, 1]. The tumor volume often requires tracheal intubation after tracheotomy as observed in the present case [3, 4]. The treatment of choice is surgery, the protocol of which depends on the site, tumor size, and appearance of the limits [3, 1, 13]. Surgery can be conservative with enucleation and curettage [12]. Surgical intervention can be radical with significant loss of substance requiring reconstruction as in our case [1, 3]. Long clinical and radiological follow-up is required in order to diagnose possible recurrences [12, 3].

CONCLUSION

Odontogenic myxoma is a rare, locally invasive benign tumor. Clinical and radiographic aspects are not characteristic and make the diagnosis difficult. A biopsy is often necessary to make the final diagnosis. The treatment is surgical, often radical, with the use of a flap. The prognosis is good.

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