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Maxillary Bone Myxoma: Case Report and Literature Review

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Abstract: *Maxillary myxoma is an odontogenic tumor derived from embryonic mesenchymal tissue. It's a rare benign tumor, slow-growing but locally aggressive. Clinical and radiological manifestations can guide the diagnosis, which remains essentially histological. Surgery is the treatment of choice.*

We report a case of 42 year-old patient complaining of a right cheek swelling. An biopsy was performed . Histology confirmed the diagnosis of maxillary myxoma. En-Bloc resection was performed. At the time of the last follow up 2 years postoperatively there was no sign of recurrence and the cosmetic appearance was satisfactory.

Keywords: *Myxoma, Benign Tumor, Rare Tumor.*

1. INTRODUCTION

Maxillary myxoma is an uncommon benign tumor. According to World Health Organization (WHO) is a locally invasive neoplasm of ectomesenchymal origin with or without odontogenic epithelium and it appears to originate from the dental papilla, follicle or periodontal ligament. It's a rare tumor comprising around 0,41-7,19% of all maxillary tumors [1-2]

Only the confrontation of clinical and radiographic elements with the anatomo-pathological examination makes it possible to establish the diagnosis and to propose an adapted surgical treatment which must be as conservative as possible.

2. REPORT CASE

A 42 year old male patient, consulted for a right cheek swelling evolving for 2 years There were no other symptoms or past medical history of note.

The clinical examination revealed facial asymmetry with a painless bony hard swelling in the right cheek of approximately 3 cm, the overlying skin was free and of normal color.

Oral examination revealed a nontender, hard swelling extending from the maxillary right first molar to the right maxillary tuberosity, thereby obliterating the right buccal vestibule. However, the adjacent gingiva and oral mucosa appeared normal. (Figure 1). The motricity and the sensitivity of the face were preserved. No masses or lymphadenopathy were revealed on neck examination.

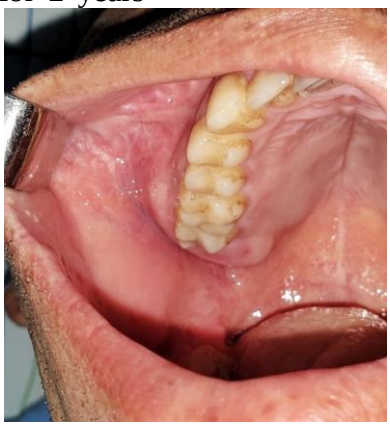

Figure1. *Pre-surgical intra-oral view*

Computed tomography CT scan with injection revealed a lesional process occupying the entire right maxillary sinus. This mass remains well limited despite being lytic, with no extension to the nasal fossa or to the inferior orbital wall and absence of anomalies of the pharyngo-laryngeal sector.**(Figures 2-3)**

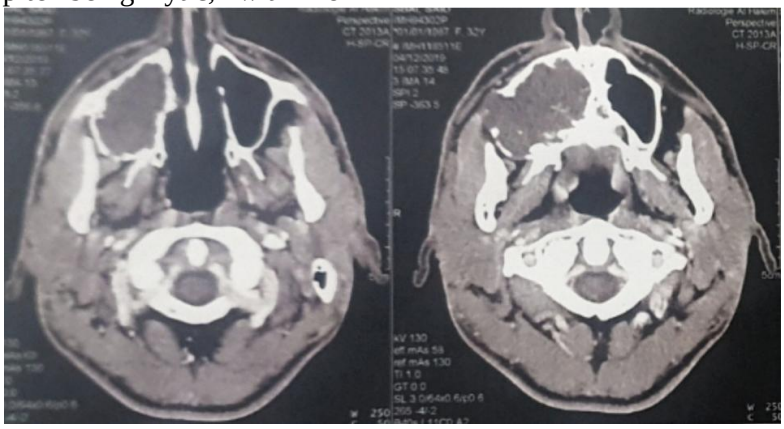

Figure2. *Axial computed tomography images showing the tumoral process occupying the entire right maxillary sinus.*

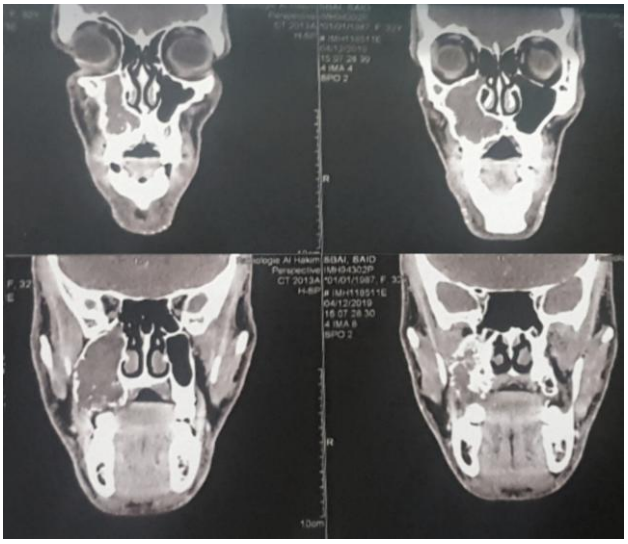

Figure3. *Coronal computed tomography images showing the extension of the maxillary tumor.*

An endobuccal biopsy of the tumor was performed, the histopathological examination showed a benign-looking tumor proliferation with no histological sign of malignancy, first evoking a myxoma. The patient underwent a removal of the tumor with a*enbloc* excision of the maxillary using vestibular approach. (**Figure 4).** The final histopathological examination confirmed the diagnosis of Maxillary odontogenic myxoma. Postoperative course was straightforward, with no recurrence at 24months follow-up. (**Figure 5)**

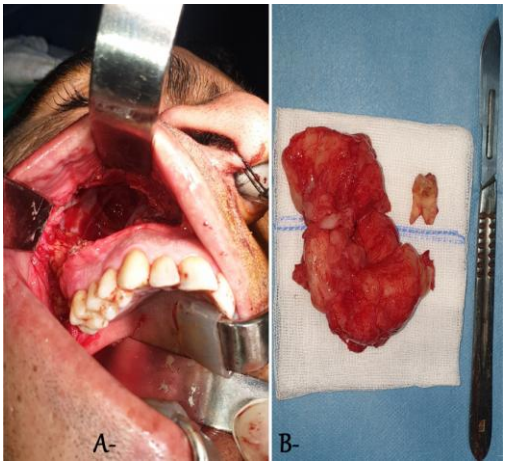

Figure4. *A-Per-operative image after tumor removal. B-The excised specimen*

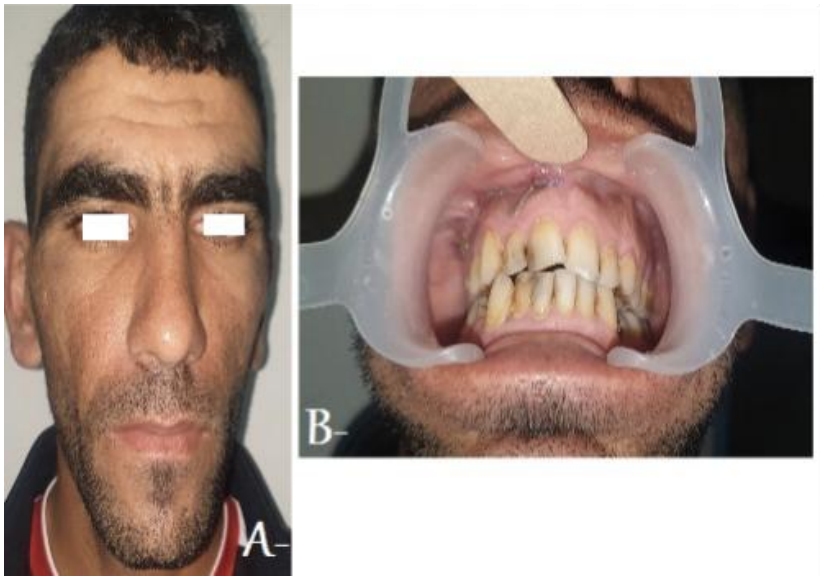

Figure5. 3 *months postoperative images*

3. DISCUSSION

Maxillary myxoma is an odontogenic benign tumor derived from embryonic mesenchymal tissue, first described by Virchow in 1863 [3].

It's a rare tumor comprising around 0,41-7,19% of all maxillary tumors [1,2], 3 to 6 % of odontogenic tumors and 0,41 % of all bone tumors [4]. The mean age occurrence is around 30 year-old, It is rarely seen in patients younger than 10 years of age or older than 50 [5,6]. No sex predominance reported [2-7].

The mandible appears to be more frequently affected than the maxilla [7-9]. Risk factors have been mentioned, like the Garney complex. This rare autosomal dominant syndrome, characterized by endocrine abnormalities, a variety of pigmented lesions of the skin and and multiple myxomas[10].

Clinically, the consultation is most often motivated by a painless slowly increasing swelling. Palpation reveals a mass of hard consistency, elastic when there is cortical effraction. The dental signs that can be associated are mobility, displacement or dental expulsion[2,11,12].In the maxilla, the intra sinusal development allows a long asymptomatic extension, which leads to an often late discovery, although pain, facial deformity respiratory distress or neurological disturbance can be found at a large size of the tumor [4].

Imaging determines the site and the extension of the tumor and helps guiding the diagnosis, Computed tomographic images of odontogenic myxomas may show multi- cystic bone lesion with cortical thickening, containing septa within a mass of tissue-like density[13,14].

On MRI, myxoma shows in hyposignal on T1 and hypersignal on T2-weighted sequences. This aspect is highly suggestive, but not systematic. Enhancement on contrast-medium injection is variable, the MRI allows more accurate study especially when the lesion is non-encapsulated and appears to be infiltrating [15].

Positive diagnosis relies on histopathologic examination, which macroscopically describes the myxoma with a whitish appearance, soft elastic and gelatinous consistency.

Microscopically, the myxoma is made up of loosely arranged spindle-shaped and stellate cells, many of which have long fibrillar processes that tend to intermesh. The loose tissue is not highly cellular, the intercellular substance is mucoid. Mitotic activity is weak, compatible with the slow growth of the tumor 16,17].

The treatment is exclusively surgical and the risk of recurrence dominates by far the therapeutic problem. Two different surgical approaches are found in the literature.

According to some authors simple enucleation or curettage may be sufficient.

Brian et al. reported no recurrence at a mean 8.5 years follow-up in maxillary myxoma resected without margins [4]. However, the recurrence rate varies between 10 and 33% depending on the series [13,19,20].Therefore a prolonged clinical and radiological surveillance is mandatory.

While other authors consider radical treatment as a treatment of choice because of local aggressive nature and the high risk of recurrence of this tumor. Either Vestibular or lateral rhinotomy ; the proper surgical approach is decided by the site of the tumor and the extent of its spread. with 1.5 cm on resection margins leading to a significant loss of substance requiring an immediate or deferred reconstruction or a implant restorations [21].

Maxillary myxoma it's unresponsive to chemotherapy and is not radiosensitive. Radio-clinical follow up is quarterly in the first year, semi-annually in the following two years, and then annually for ten years [11].

In the present case, the tumor was completely removed by *enbloc* resection using a vestibular approach and no recurrence was reported after 24 months of the surgery.

4. CONCLUSION

Maxillary myxoma is a rare odontogenic tumor, benign but locally aggressive. The clinical and radiological aspects are not specific and the positive diagnosis is only based on histologic specimen analysis. Treatment is surgical and strict prolonged surveillance is required due to the risk of recurrence.

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