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Myofibroblastic Sarcoma of the Maxillary Sinus: Very Rare Entity

L. Laghsene[1]; Z. Lita[2]; M. Loudghiri[3]; W. Bijou[4]; Y. Oukessou[5]; S. Rouadi[6]; R. Abada[7]; M. Roubal[8]; M. Mahtar[9]
Department of Otolaryngology Head and Neck Surgery, University Hospital Ibn Rochd, Casablanca, Morocco.

Abstract:-

- ***Introduction:***

The Myofibroblastic Sarcoma is a recently discovered malignant tumor that mainly affects the face and neck region, but it occurs most often in the tongue and oral cavity. However, involvement of the maxillary sinus has only been reported a few times in the literature.

It is a slowly growing neoplasm with a high recurrence and metastasis risk.

- ***Case Presentation:***

18 years old female patient, admitted for a painless mass of the left hemi palate increasing progressively in volume evolving since 5 months, associated to a left nasal obstruction and purulent rhinorrhea. Once in our department, she underwent a facial CT scan, followed by a facial MRI, which showed a tissue process centered primarily on the left maxillary sinus, causing lysis of the inferior wall and bulging of the hard palate on the left side . The surgical decision involved combining an endoscopic and external approach, and was supplemented by radiation therapy. The follow up demonstrated a successful outcome. the patient did not present any recurrence or distant metastasis.

- ***Conclusion:***

The key lessons are the essential role of imaging in diagnosing and treating LGMS , as well as the crucial role of postoperative radiotherapy in preventing recurrence even though this subject is still controversial.

Keywords:- *Myofibroblastic Sarcoma , Low Grade , LGMS , Maxillary Sinus , Paranasale Sinuses , Head and Neck Cancer , Case Report . SMA. Radiation Therapy.*

I. INTRODUCTION

LGMS is an extremely rare tumor that has only recently come to light. The first such case was identified only up to 1998s. (1)

Low-grade myofibroblastic sarcoma (LGMS) represents an atypical tumor composed of myofibroblasts with a predilection for the head and neck, especially in the tongue and oral cavity. (2,3)

Myofibroblasts are mesenchymal spindle-shaped cells present in almost every soft tissue. LGMS usually occurs in adult patients with a slight male predominance. Children are rarely affected. (4)

Low-grade myofibroblastic sarcomas (LGMSs) have been reported in the literature under various terms, including, sarcomas of myofibroblasts, myofibrosarcomas, myofibroblast-rich fibrosarcomas, leiomyosarcoma myofibroblastic variant and spindle-cell sarcomas showing myofibroblastic differentiation (5).

Clinically, the most common reason for consultation is a painless swelling or expanding mass .It is a slowly growing tumor with a high tendency to local recurrence and metastasis, even after a long time . (6)

Radiological imaging reveals typically a tumor with regular borders, clear and visible destructive margins (7; 8).

II. CASE REPORT

A 18 years old female patient, with no particular pathological antecedents, was referred to our department for a painless mass of the left hemi palate increasing progressively in volume evolving since 5 months, associated to a left nasal obstruction and purulent rhinorrhea.

The oral cavity examination revealed an ulcerative budding mass of the left hemi palate that does not extend beyond the midline. The nasal examination revealed nasal smooth bulging of the left lateral nasal wall obscuring all landmarks. Furthermore, No lymphadenopathies were found.

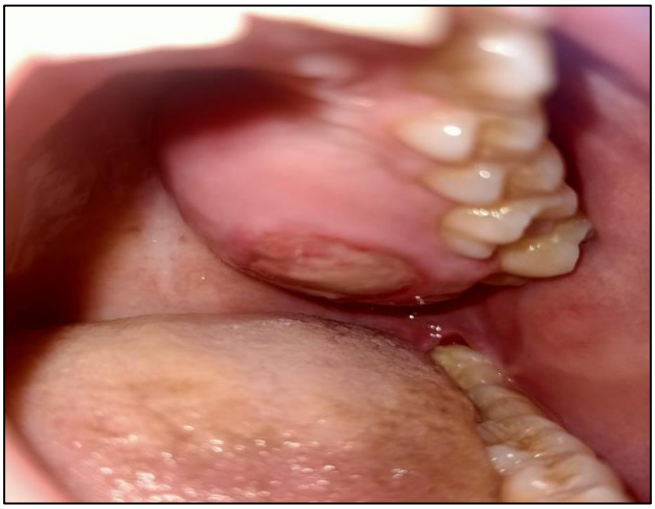

Fig 1 Intraoral Image showing the left Hemi Palate Budding Mass

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The Facial CT scan demonstrated a tissue density lesional process of the left maxillary sinus , enhanced after injection of the contrast product which lyses the alveolar process below , comes into contact with the cavum.

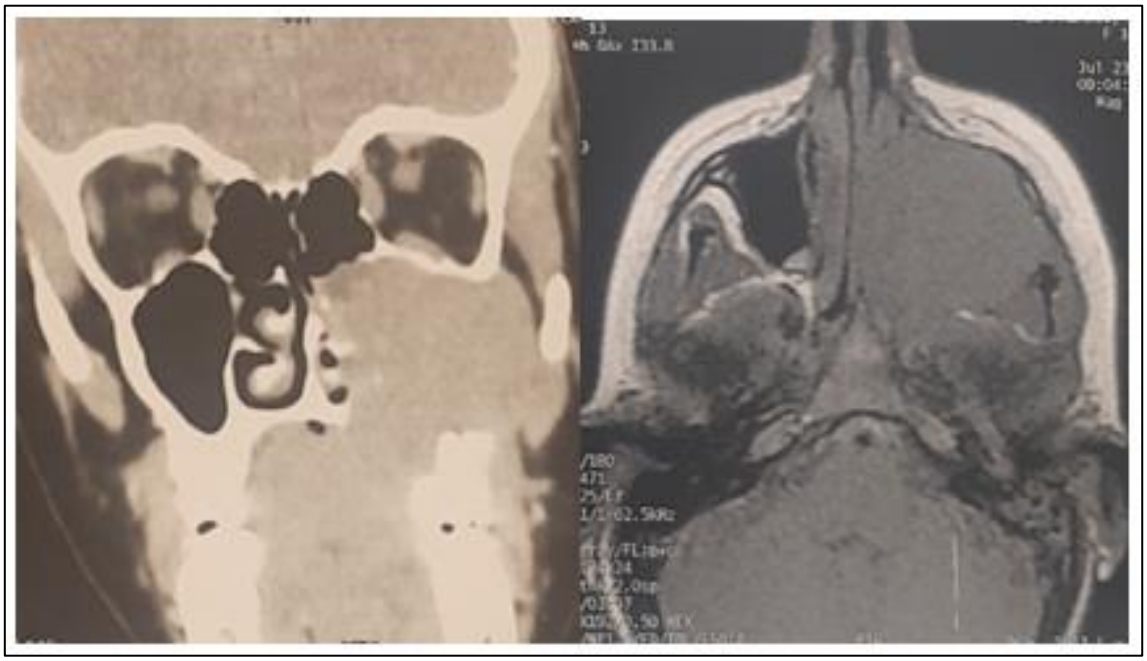

Fig 2 Axial / Coronal Facial CT scan images showing: a left maxillary sinus mass destroying the alveolar process

The Facial MRI showed a tissue process of the left maxillary sinus in T1 isosignal , T2 hypersignal , well limited and enhanced after gadolinium injection with invasion of the bony palate and lysis of the external , internal and anterior walls .

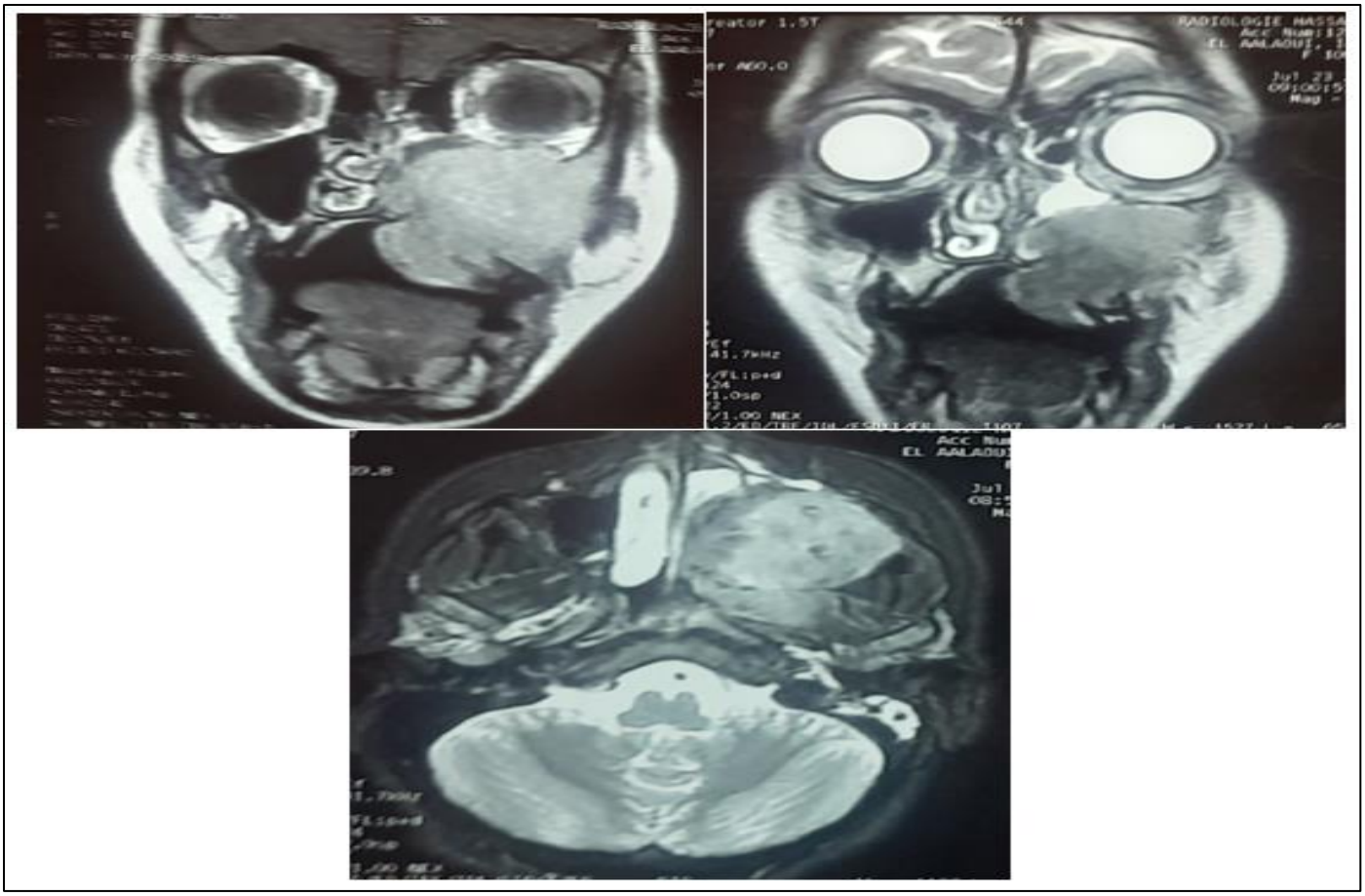

Fig 3 Axial and Coronal Facial MRI images showing the left maxillary sinus process in T1,T2 signal

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The patient underwent a biopsy of the left palatal lesion under local anesthesia and the histological study revealed a slightly atypical spindle cell proliferation.

The patient underwent under general anesthesia an inferior, anterior and internal maxillectomy with 8-tooth excision. The neck dissection was not performed . The loss of palatal substance was filled with a dental prosthesis. the Treatment was completed with adjuvant radiation therapy.

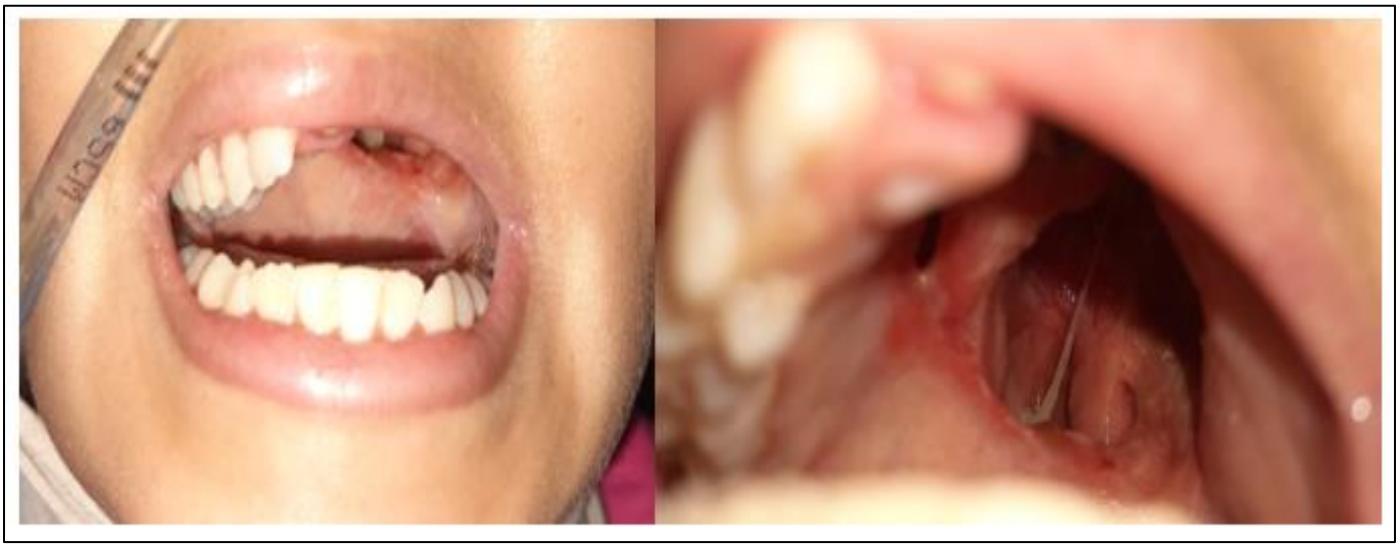

Fig 4 Post-Operative Images

The definitive Histopathological examination supported by immunohistochimiy revealed a Low Grade Myobroblastic Sarcoma.

The clinical evolution was very favorable , The patient did not show any signs of recurrence or distant metastasis at her 6-month follow-up and did not experience any significant swallowing dysfunction.

III. DISCUSSION

LGMS of the maxillary sinus is extremely rare; only seven cases have been diagnosed to date. It is a low-grade malignant tumor with a high tendency for recurrence and distant metastases, even after many years (9, 10)

LGMS has an affinity for the head and neck region, particularly the oral cavity and tongue. The most usual location of LGMS after the tongue is the mandible, palate , neck, maxilla, larynx, , and lips (10). The myofibroblastic sarcoma of the maxillary sinus and buccal mucosa have been very rarely documented in world literature until date.

The myofibroblastic sarcoma can also be confused with both benign and malignant affections such as fibromatoses , nodular fasciitis, , fibrosarcoma and leiomyosarcoma . (11)

Radiologically , Facial MRI should invariably be prescribed in the presence of any suspicion of soft tissue sarcoma of the head and neck (NCCN Guidelines version 2. 2022)(12).

Histologically, this tumor is made up of slender spindle cells with Eosinophilic cytoplasm and a spindle nucleus ,associated to a Focal expression of smooth muscle actin SMA , the Mitotic figures ranges from 2/10 to 20/10 (7) . in our case the cell proliferation was fusiform expressing SMA , the cytoplasm was eosinophilic with an elongated sometimes undulating nucleus and mitotic index was estimated at 2 /10 .

Immunohistochemistry is very essential for exact pathological diagnosis.

The recurrence rate of LGMS is about 25.45% and peaks at the highest degree, particularly when the tumor is found in the nasal and paranasales sinuses.

Although no consensus has been arrived about treatment of LGMS yet. The currently recognized gold standard treatment for managing sarcomas invading bones is radical surgical excision of the tumor with a tumor free margin (14.15).

But In cases of positive margins, performing radical excision is contraindicated or when the tumors invade the blood vessels or nerves, radiotherapy or chemo radiotherapy should be taken into account. (14 ; 16 ; 17) .

Radiotherapy relatively gives higher rate of recurrence, even according to Maruyama et al. radiotherapy should not be administered after tumor resection, as it raises the risk of postoperative recurrence. (13).

Although, qualifying patients with pre-existing poor prognostic factors may also be associated with a heightened risk of recurrence. Among these factors, the resection status, positive margins, regional lymph node involvement, and age over 60 years are cited (14).

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