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Ameloblastic Sarcoma of the Mandible: Case Report

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Abstract **Case Report**

Ameloblastic fibrosarcoma (AFS) is considered as a rare malignant neoplasm composed of benign, odontogenic ameloblastomatous epithelium and malignant ectomesenchyme. It predominantly occurs in the posterior mandible with age ranging from 3 to 83 years. Pain and swelling are the most commonly associated clinical symptoms. The purpose of this report is to present a case of 23-year-old male patient who presented with an extra oral swelling in mandible region which was later proven histopathologically as AFS.

Keywords: Ameloblastic sarcoma, Malignant odontogenic tumor Sarcoma.

INTRODUCTION

Ameloblastic fibrosarcoma (AFS) is a malignant odontogenic tumor composed of a benign epithelium and a malignant mesenchymal component [1]. Itis defined as "a neoplasm with a similar structure to the Ameloblastic Fibroma, but in which the mesodermal component shows features of sarcoma [2]. AFS may arise denovo or may develop by malignant transformation of innocuous Ameloblastic fibroma [3-5]. Clinically, patients present with the chief complaint of pain and swelling. It presents in a wide age range of 3–83 years [3]. In this article, we are presenting the case of AFS.

CASE REPORT

The patient was 23-years-old without comorbidities. His medical history dates back to September 2023 with the appearance of a right mandibular swelling.

A mandibular MRI was performed, indicating a locally advanced malignant mandibular tumor process with retro-pharyngeal lymphadenopathy (Fig 4).

Furthermore, a Chest-Abdomen-Pelvis CT scan did not show any secondary lesions. A biopsy was performed, indicating an ameloblastic sarcoma (Fig 1, 2).

The appearance was suggestive of a spindle cell sarcoma, while the IHC profile was suggestive of an ameloblastic sarcoma:

Antibody anti CD34: negative
Antibody anti PS100: negative
Antibody anti cytokeratine AE1 AE3: positive
Antibody anti desmine: positive
Antibody antimuscle lisse: positive
Antibody antivimentine: positive
Ki67: 20%

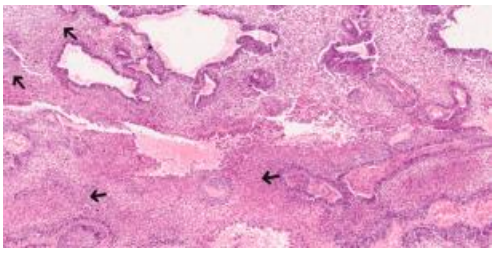

Figure 1

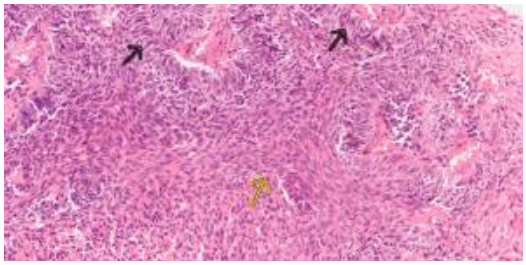

Figure 2

The patient received neoadjuvant chemotherapy (CMT) before surgery, with 4 cycles of Adriamycine+cyclophosphamide, in contrast the progression was marked by the clinical progression of the swelling.

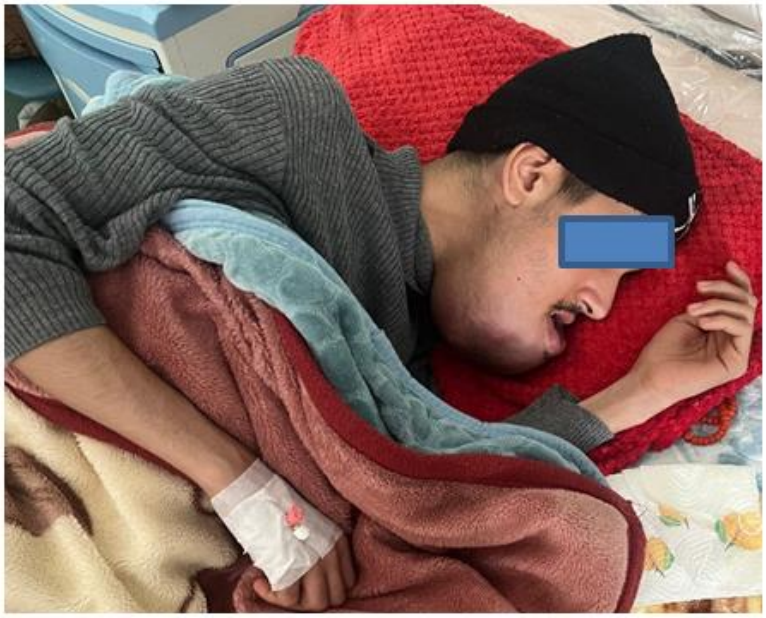

Figure 3: Right mandibular swelling

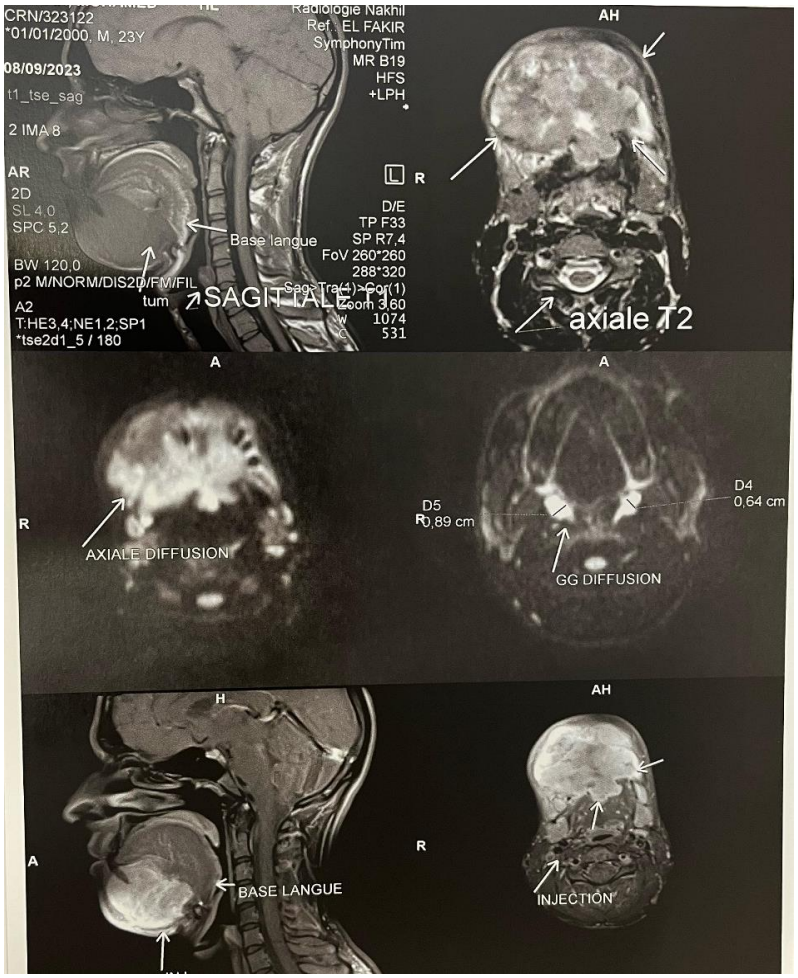

Figure 4: Locally advanced malignant tumor process of the mandible with retropharyngeal lymphadenopathy

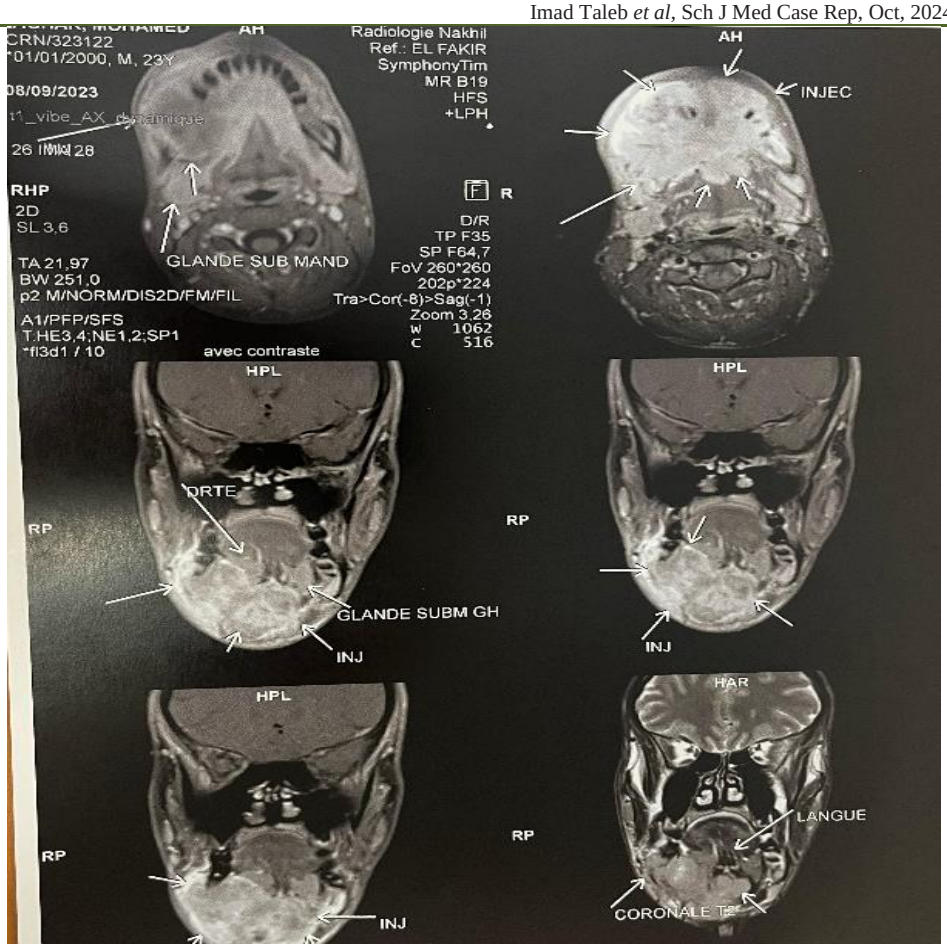

Figure 5: Locally advanced malignant tumor process of the mandible with retropharyngeal lymphadenopathy

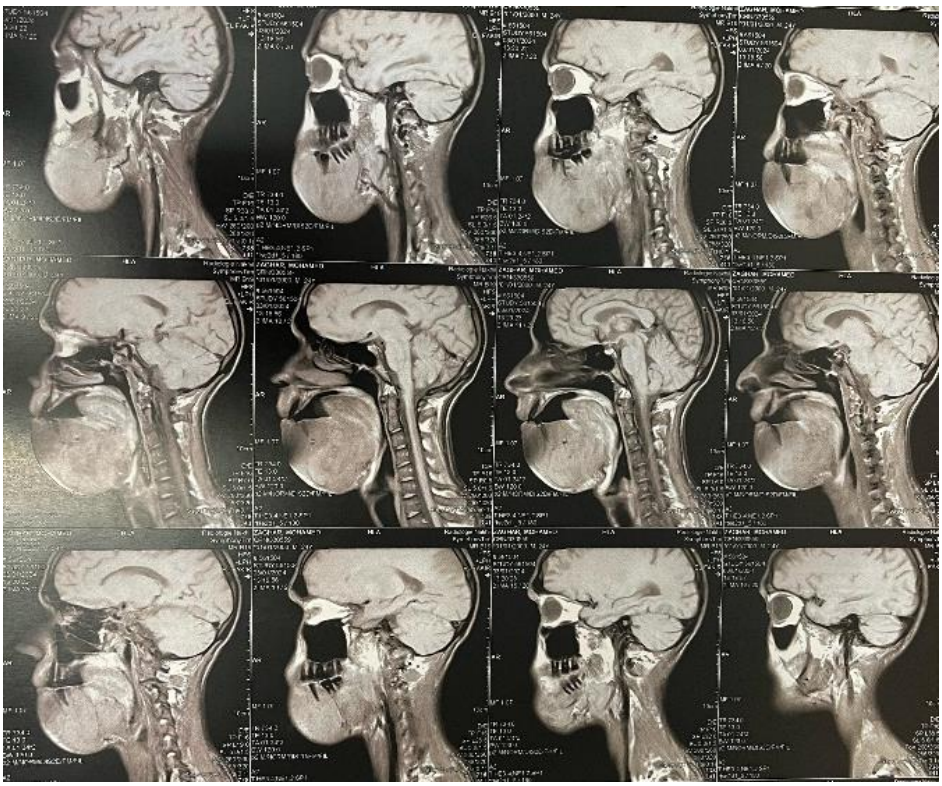

Figure 6: Increase in the volume of the tumor process of the mandibular arch by approximately 25% :3/1/2024 versus 8/9/2023

DISCUSSION

AFS is a rare malignant odontogenic tumor [4]. Heath published the first report of an AFS in 1887 where he described it as a 'spindle-celled sarcoma' of the mandible. The mean age of presentation ranges from 15 to 22 years and mandible is most commonly affected site [3, 6, 8, 9] similar to our case. Slight male predilection was noted [3]. Pain and swelling are the most common findings. Ulceration and paresthesia of lip have also been reported. Most of the cases arise in previously diagnosed Ameloblastic Fibroma cases [3]. Kegal [2], Leider *et al*., (1972) explained the mechanism of malignanttransformation by stating the role of surgical trauma to benign recurring tumors. Chomette *et al.*, 1983 stated that the functional involution of epithelial component might play a role in sarcomatous transformation of mesenchymal component [1].

The AFS has a very low potential for distant metastasis [7]. En bloc resection with wide margins and follow-up for atleast 10 years is the recommended treatment. Chemotherapy may be indicated as an adjuvant with radiotherapy to surgical resection, if a wide margin of resection is difficult to achieve, chemotherapy may be indicated as an adjuvant with radiotherapy to surgical resection.

Due to lack of clinical reports, there is no consensus on the treatment yet. In general, the treatment of choice is surgical excision with clear margins and long-term follow-up. Conservative approach shows high incidence of recurrence [10].

There is no clear cut consistent pattern of adjuvant chemo or radiotherapy. It has been used in extensive recurrent lesions, and that gives more of regression than cure [11] in our case, the patient was refractory to first-line chemotherapy.

CONCLUSION

Ameloblastic fibrosarcoma (AFS) is a rare malignant neoplasm there is no consensus on the treatment yet.

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