

## Case Report

# Ameloblastic fibrosarcoma of the upper jaw: Report of a rare case with long-term follow-up

Maryam Khalili<sup>1</sup>, Pouyan Amini Shakib<sup>2</sup>

<sup>1</sup>Department of Oral and Maxillofacial Pathology, School of Dentistry, Tehran University of Medical Sciences, Tehran, <sup>2</sup>Department of Oral and Maxillofacial Pathology, School of Dentistry, Babol University of Medical Sciences, Babol, Iran

## ABSTRACT

Received: May 2012  
Accepted: November 2012

**Address for correspondence:**  
Dr. Pouyan Amini Shakib,  
Babol University of Medical  
Sciences, School of  
Dentistry, Ganjafroz Ave,  
Babol, Iran.  
E-mail: pouyanshakib@  
yahoo.com

Ameloblastic fibrosarcoma (AFS) is a rare malignant mixed odontogenic tumor which is usually considered as the malignant counterpart of ameloblastic fibroma. Only mesenchymal component represents sarcomatous alterations and ameloblast-like epithelial nest remains bland in AFS. Here, we report a case of AFS in a 26-year-old man in the maxilla, which was regarded as an uncommon location for this tumor. After 2 years follow up, no evidence of recurrence was noted. We also emphasize on comprehensive clinical, radiographic, and histopathologic evaluation of such patients rather than immunohistochemical staining to make an accurate diagnosis.

**Key Words:** Ameloblastic fibrosarcoma, maxilla, odontogenic tumor

## INTRODUCTION

Ameloblastic fibrosarcoma (AFS) is a rare mixed odontogenic tumor composed of benign epithelial and malignant ectomesenchymal components. Although AFS is regarded as the malignant counterpart of ameloblastic fibroma, it may arise de novo without any pre-existing lesion.<sup>[1]</sup> Also, some authors have described “ameloblastic dentinosarcoma” and “ameloblastic odontosarcoma,” based on the mineralized structures found on microscopic examination.<sup>[2]</sup> To our knowledge, only 71 cases of AFS have been reported until now in the English literature.<sup>[3,4]</sup> Histologically, the tumor is usually defined as a low-to intermediate-grade malignancy and clinically shows locally aggressive behavior and high recurrence rate but rare metastasis.<sup>[4,5]</sup> This entity is more common in young adults with approximately equal gender distribution. The majority

of reported cases have been found in the posterior region of mandible and rarely maxillary involvement is indicated.<sup>[6]</sup> The most common clinical findings are swelling and pain. Paresthesia is also observed. Radiographically, the tumor usually shows a destructive radiolucent pattern with ill-defined borders suggestive of malignancy.<sup>[7]</sup>

It is critical to differentiate AFS, especially low-grade variant, from ameloblastic fibroma, and recently, several immunohistochemical studies with Ki67, Bcl-2, PCNA, c-KIT, and P53 have been performed, suggesting that proliferating markers in association to histopathologic features could be useful for identifying the malignant tumor.<sup>[3,5,8]</sup>

In this article, we reported a case of maxillary AFS. The clinical, radiographic, and histopathological features were described in addition to the patient's management and present status.

## CASE REPORT

A 26-year-old man, educated in an engineering field, from a city in North of Iran was referred to School of Dentistry of Tehran University of Medical Sciences by an oral and maxillofacial surgeon (OMFS) for oral pathology consultation in winter of 2010. He

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complained of continuous dull pain in the upper left region of his face from 2 months ago and visual disturbances in recent weeks. The upper left second and third molars had been extracted by a dentist one month ago, assuming that the pain was of dental origin. As expected, the pain persisted and healing of the extraction site was not normal. Therefore, a more serious process was suspected and the patient was referred to an OMFS for further examination. He performed an incisional biopsy and sent it to a general pathology service for histopathologic examination. The microscopic diagnosis was “adenomatoid odontogenic tumor” which did not match the clinical and radiographic findings. The surgeon decided to ask for a second opinion and sent the documents to our center for consultation.

Clinical examination revealed a bony hard swelling of the left posterior region of maxilla. Intraorally, nontender buccal and palatal expansion of the alveolar ridge at the same area was observed. Also, the extraction site of the second and third molar was filled with an irregular exophytic ulcerated mass which bled easily on palpation [Figure 1]. No other intra- or extra-oral sign was observed; the remaining head and neck examination and general physical evaluation were unremarkable and no significant event was noted in the medical history.

Panoramic radiographs demonstrated an ill-defined unilocular radiolucent lesion in the left posterior region of maxilla and the left maxillary sinus was totally obliterated. Computed tomography scan showed extension of the lesion medially and vertically to the nasal septal bone and the inferior orbital rim, respectively [Figure 2].

Microscopic examination of the slides revealed a neoplastic tissue with scattered benign appearing cords and nests of epithelial cuboidal or columnar cells with peripheral palisading in a myxoid to highly cellular mesenchymal background mostly composed of plump stellate and ovoid to spindle cells. Nuclear hyperchromatism, pleomorphism, and scattered mitotic figures were identified in some areas as well as giant and bizarre cells and the tumor was diagnosed as “ameloblastic fibrosarcoma” accordingly [Figures 3a and b].

No evidence of regional lymphadenopathy or distant metastasis was found and a segmental resection of the maxilla along with wide excision of the surrounding soft tissues was performed.

Histopathologic examination of the whole specimen confirmed the diagnosis of AFS. All surgical margins were free of tumor infiltration. However, adjuvant radiotherapy was also performed in order to ensure a better outcome. The patient was under close follow up and after 6 months, rehabilitative prosthetic treatment was started. At present, after 2 years, the patient is free of any recurrence and in a good health status [Figures 4a and b].

## DISCUSSION

AFS is a very rare intraosseous odontogenic malignancy with approximately 71 cases published to date. Approximately, one third of reported cases appear to originate from recurrence of a previous ameloblastic fibroma. Therefore, some investigators recommend a more aggressive treatment for ameloblastic fibromas as compared to previous procedures.<sup>[9]</sup> Our case is best considered as *de novo*, since the patient did not report any previous lesion in the area.

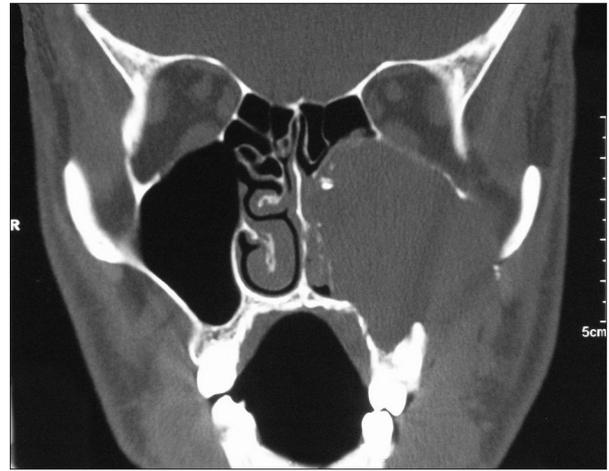
The mean age at the time of diagnosis of AFS for 62 cases reviewed by Bregni *et al.*<sup>[1]</sup> was 27.3 in a wide age range, from 3 to 83 years. According to these 62 published cases, the tumor is more common in males than females (59.7% vs 37.1%), although in two cases (3.2%) gender was not specified; and also AFS is more frequent in the mandible (79%) than the maxilla (21%) and the majority of them were found in the posterior region of the mandible. Considering additional 9 cases reported so far,<sup>[2-6,8-11]</sup> no significant alteration was observed in age and gender distribution [Figure 5] or location prevalence (the mandible to maxilla ratio: 3.4).

AFS is a highly recurrent lesion. To date, 25 (35%) of the 71 reported cases<sup>[1-6,8-11]</sup> have had at least one recurrence during follow-up period and 14 patients (19.7%) have died of their disease within 3 months to 19 years. Clinical findings vary among reported cases but usually include pain and swelling.<sup>[10]</sup> Also, despite regional lymph node involvement or distant metastasis reported in a few cases, some authors have considered AFS as a low-grade sarcoma or even a semi-malignant lesion.<sup>[6]</sup> In our patient, the lesion was located in the posterior maxilla with maxillary sinus involvement which is a rare location for this tumor. No evidence of regional or distant metastasis was noted.

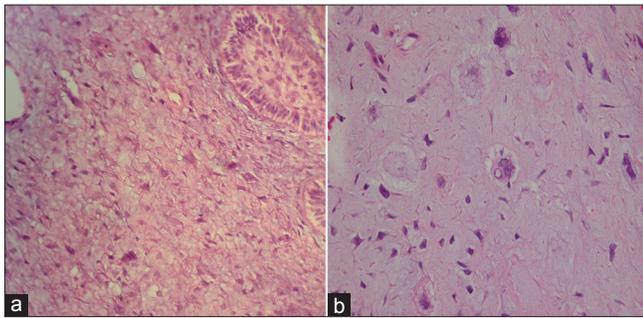
Radiographically, AFS usually appears as an expansile destructive radiolucency with ill-defined margins.<sup>[6,7]</sup> In



**Figure 1:** Intraoral view shows buccal and palatal expansion in the left maxillary region and the exophytic ulcerated mass



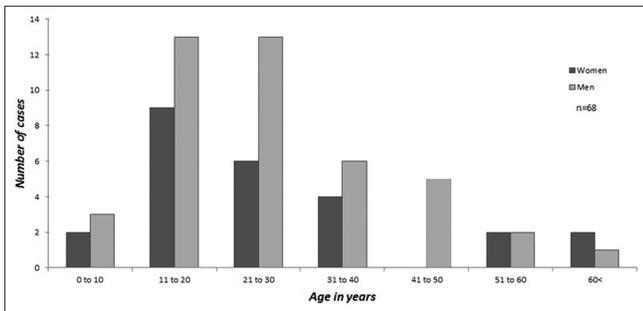
**Figure 2:** Computed tomography scan shows extensive destruction of the left maxillary sinus



**Figure 3:** Photomicrographs showing (a) benign-appearing epithelial islands and highly cellular mesenchymal component (H and E, original magnification  $\times 100$ ); (b) pleomorphism and scattered bizarre cells in mesenchymal component (H and E, original magnification  $\times 100$ )



**Figure 4:** Intraoral examination and (a) CT scan evaluation (b) revealed no evidence of recurrence after two years



**Figure 5:** Age and gender distribution of 68 cases of Ameloblastic fibrosarcoma (age or gender of three cases of 71 reported AFSs have not been specified)

our case, destructive behavior of the lesion was obvious in both conventional radiographs and computed tomography images. Failure to correctly evaluate the panoramic radiographs and establish a clinical differential diagnosis by the general practitioner should be given special attention. This led to unnecessary extraction of the teeth and delayed diagnosis of the

tumor. In case of oral and maxillofacial pain and symptoms, dentists are usually the first healthcare practitioner people consult. Therefore, their role in early diagnosis of diseases especially malignancies is crucial. Unfortunately, in most cases, jaw pain is interpreted as of dental origin and the patient is treated accordingly. Dentists should be familiar with signs and symptoms of malignant tumors of the jaws, especially rare cases with unusual presentation. This knowledge along with a high index of suspicion results in correct differential diagnosis and early detection of serious cases and could have a tremendous impact on the prognosis and total outcome of the disease.

Microscopically, the bland epithelial component of AFS is similar to that seen in ameloblastic fibroma, although it is frequently less prominent. The definitive diagnosis of AFS is established based on histopathologic evaluation of the mesenchymal component which usually demonstrates various features of malignancy including cellular atypia, pleomorphism, and mitotic figures. However, sometimes, it is seriously difficult to differentiate AFS, especially low-grade tumors from ameloblastic fibroma.<sup>[9]</sup> When odontogenic epithelium is less prominent or absent, other benign locally

invasive or malignant spindle cell tumors such as desmoplastic fibroma and neurofibrosarcoma should also be considered.<sup>[6]</sup> In our case, highly cellular areas of mesenchymal cells with pleomorphism and scattered bizarre giant cells were observed, although mitotic figures were not prominent. Therefore, also considering epithelial component, the diagnosis based on histopathological examination was not a serious problem.

Recently, some authors have suggested a panel of biomarkers associated with cell proliferation (Ki67, PCNA, and c-KIT) and apoptosis (Bcl2) to overcome difficulties in the diagnosis of low-grade tumors and evaluate the growth potential in the mesenchymal component without considering clinical and radiographic findings.<sup>[3,5]</sup> The results revealed that all the studied proteins were overexpressed in the malignant mesenchymal portion of AFS compared with ameloblastic fibroma, suggesting that nuclear proliferative factors such as Ki67 and proliferative cell nuclear antigen, in association to histopathologic features, may be useful for identifying the malignancy.

The treatment of choice for AFS is radical surgical excision without primary neck dissection. Some investigators recommend adjuvant chemo- and/or radiotherapy, but others are uncertain about their benefits. Because of rarity of cases, it is almost impossible to estimate long-term prognosis accurately and many patients have died from uncontrolled local invasive disease.<sup>[4,6]</sup> Our patient underwent radical surgical resection and radiotherapy with no evidence of recurrence or metastasis after 2 years, which could be considered as a favorable outcome.

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**How to cite this article:** Khalili M, Shakib PA. Ameloblastic fibrosarcoma of the upper jaw: Report of a rare case with long-term follow-up. *Dent Res J* 2013;10:112-5.

**Source of Support:** Nil. **Conflict of Interest:** None declared.