CASE REPORT

Ameloblastic fibrosarcoma: Report of a case

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ABSTRACT
Ameloblastic fibrosarcoma (AFS) is a rare odontogenic malignancy with benign epithelial and malignant ectomesenchymal components. About 66 cases have been reported in the medical literature. We therefore report an additional case as well as a review of literature to add to the existing knowledge on this rare lesion.

Key words: Ameloblastic fibrosarcoma, odontogenic tumor, ectomesenchyme

INTRODUCTION

Ameloblastic fibrosarcoma (AFS) is a rare neoplasm composed of benign ameloblastomatous epithelium and malignant ectomesenchyme.[1] About 66 cases have been reported in the medical literature.

AFS has a predilection for the mandible; and it is most commonly seen in the third decade of life with a male preponderance.[1,2] Patients often present with a painful swelling,[1,2] and radiographically as a multilocular radiolucent lesion with indistinct margins.[2] Metastasis is rare, but recurrences have been reported.[2] Treatment is by radical surgery.[1]

We present a second case of AFS from our institution.

CASE REPORT

A 28-year-old asthmatic Nigerian female presented at the Oral Surgery Clinic of the University College Hospital with a 4-year history of a gradually enlarging right maxillary swelling which accelerated in growth following two pregnancies. Extraorally, the lesion extended anterioposteriorly from the right alar of the nose to the right tragus and superioinferiorly from below the infraorbital rim to the right commissure of the mouth, measuring approximately 14 × 10 cm in size [Figure 1].

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Intraorally, there was no limitation of mouth opening. However, there was an intraoral extension of the tumor from 11 to 18, also involving the right side of the palate. The overlying mucosa was adherent to the lesion and had irregular areas of ulceration.

Computerized tomography scan showed a well-defined, homogenous, isodense mass filling the right maxillary antrum with erosion of the lateral, posterior and anterior walls. The mass also extends to the floor of the right orbit, displacing it upwards without perforating it. The adjacent ethmoidal air cells and nasal cavity were invaded by the lesion [Figure 2].

The incisional biopsy done was inconclusive, but was suggestive of a malignant lesion. The patient had right hemimaxillectomy; right external ethmoidectomy and sphenoidectomy performed by a combined team of maxillofacial surgeons and otorhinolaryngologists through a Weber-Ferguson incision with an infraorbital extension. The surgical specimen was then subjected to histopathological examination which revealed a low grade malignant neoplasm displaying mesenchymal and epithelial differentiation [Figure 3a and b]. The mesenchymal component displayed storiform [Figure 3c] and herring bone pattern in a fibromyxoid stroma, while the epithelial component was composed of focal ameloblastic islands made up of columnar cells arranged in a palisaded pattern with a central area of stellate reticulum-like cells. Focal areas of necrosis and mitotic figures were seen. The features were those of AFS. Patient was rehabilitated with a feeding plate in the immediate postoperative period and later with an obturator. She has remained disease-free 29 months after surgical intervention [Figure 4].

DISCUSSION

AFS was first reported by Heath in 1887 in which he described a spindle cell sarcoma that had epithelial cells resembling those of the enamel organ.[1] The most concise reviews till

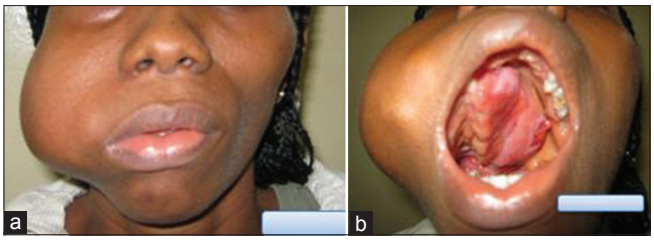

Figure 1 (a and b): Right maxillary swelling with a massive intraoral extension involving the palate

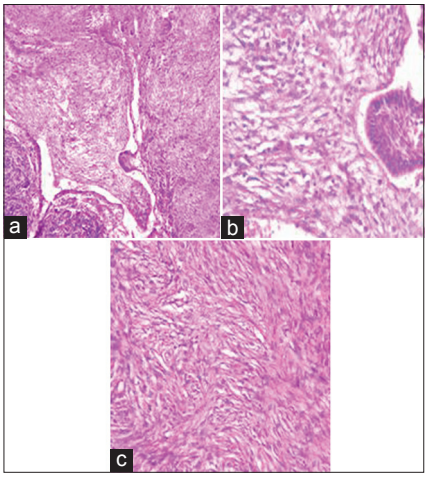

Figure 3: Photomicrograph showing low grade malignant neoplasm displaying mesenchymal and epithelial differentiation (H&E stain, a:×50, b:×400). (c) The mesenchymal component displayed storiform and herring bone pattern (H&E stain, ×100)

date are those of Muller *et al.*, where 51 cases of AFS were reviewed and Bregni *et al.*, with the review of 62 cases.[2,3] Bregni *et al.*, reported a male to female ratio of 1.6:1 and a predilection for the mandible over the maxilla.[3] Between 2001 and 2009, at least four more cases have been reported.[4,5]

The most commonly proposed pathogenesis of AFS is transformation of an existing ameloblastic fibroma (AF). Muller *et al.*, reported that 44% of AFS had a previous diagnosis of AF.[2] Incisional biopsies, however, may present a potential source of error because malignancy may have arisen from focal areas not biopsied.[6] Thus, serial sampling of the surgical specimen of AF is essential for accurate diagnosis. Also, a high index of suspicion should be entertained in patients with AF. Long-term follow-up for recurrences and close monitoring for transformation in addition to complete surgical excision should be done in patients diagnosed as AF.[2]

Our patient had a right maxillary lesion of four years duration and gave a history of accelerated growth of the tumor during pregnancy, which raises speculations on the role hormones may play in the progression of this tumor. Also, our patient was 28-years-old at presentation, similar to the mean age reported for AFS which is 27.5 years.[2] The tumor had previously not been operated on, thus implying that our case was a de novo malignancy.

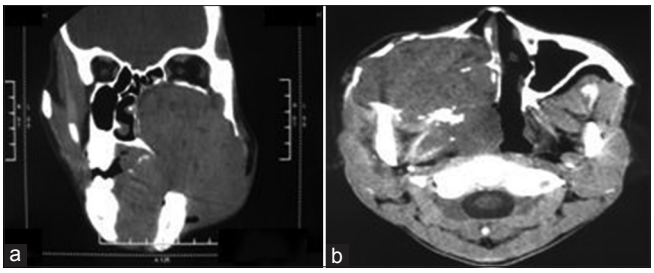

Figure 2 (a and b): A computerized tomography scan which shows an isodense area that has eroded the antral walls and destroyed the maxilla

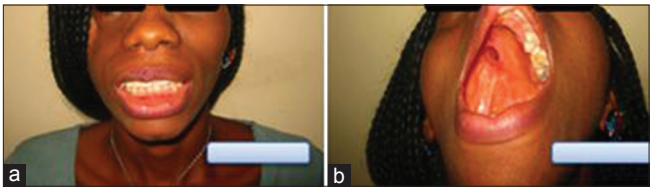

Figure 4 (a and b): Postoperative profiles

AFS has a predilection for the mandible, but a few cases have been reported in the maxilla. Also, our patient had an episode of epistaxis late in the presentation, raising a possibility that this tumor arose centrally. This is further corroborated by the absence of tumor cells within the antral lining. AFS has been reported to originate from the antrum in an infant[4] and has also been reported to involve skull base and surrounding regions in a 48-year-old male.[5] Uncommon finding with AFS is the involvement of the cervical or submandibular lymph nodes.[2]

The histological architecture of AFS is characterized by benign epithelial islands that are similar to those of follicular ameloblastoma.[1] These islands are composed of peripheral cells that are columnar or cuboidal in shape and are arranged in a palisading pattern. At the center of these islands are polyhedral cells reminiscent of stellate reticulum.[1] The epithelial components are widely separated by a malignant ectomesenchymal stroma which makes up the bulk of the tumor. This consists of hypercellular connective tissue with plump polygonal to fusiform stromal cells which show mild to moderate cytologic atypia and numerous mitotic figures in a myxoid matrix.[1] Our case had very few odontogenic nests compared to the extensive stroma, a feature reported to be commonly seen in recurrent lesions in which the sarcomatous component completely overgrows the epithelial component.[2]

Further investigations done by Yamamoto *et al.*, in their study, showed the presence of keratin in the columnar and polyhedral cells of the epithelial component and vimentin in the ectomesenchymal component verifying the biphasic nature of this tumor.[7] Also, Williams *et al.*, demonstrated alterations of the p53 and c-KIT genes in the sarcomatous component of an anaplastic AFS that transformed from a recurrent AF.[8]

Presently, surgical excision with a margin of normal tissue

is the treatment of choice. Other treatment modalities such as adjuvant chemotherapy and radiotherapy were found to be ineffective.[1] Furthermore, some authors have proposed that AFS should be considered as a low grade fibrosarcoma[6] in view of its more favorable prognosis in comparison to fibrosarcomas of the orofacial region. Our patient has remained tumor-free 29 months after wide surgical excision, which is the recommended treatment for AFS.

In conclusion, AFS is a rare tumor in which the first reported case from our institution was by Daramola *et al*., in 1979.[9] Also, Adekeye *et al*., in 1978[10] reported a case in northern Nigeria. To the best of our knowledge, this is the third reported case in Nigeria and the only one from Nigeria with more than 6 months follow-up after treatment. Long-term follow-up would provide more information on survival and recurrence rates of this tumor.

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