Ameloblastic Fibrosarcoma arising denovo in mandible: A Case Report
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Abstract
Ameloblastic fibrosarcoma (AFS) or ameloblastic sarcoma is an extremely rare odontogenic neoplasm. Until now, 64 cases have been reported in the English literature. The authors report AFS in the mandible on the left side of a 26 year old female. The tumor was composed of odontogenic epithelium, resembling that of ameloblastoma, and a mesenchymal part exhibiting features of fibrosarcoma. The patient was treated with surgery and chemotherapy. The recovery was uneventful and the prognosis is under observation.

Keywords
Ameloblastic fibrosarcoma;
Neoplasm;
Ameloblastoma.

Introduction
Ameloblastic fibrosarcoma (AFS) is a rare odontogenic tumor comprising a benign epithelial component and a malignant ectomesenchymal component. AFS is regarded as the malignant counterpart of the ameloblastic fibroma, and along with ameloblastic fibrodentinosarcomas and ameloblastic fibro-odontosarcomas, belongs to the group of odontogenic sarcomas according to the 2005 WHO classification. AFS was first described by Heath in 1887. Development of this lesion in the jaws either de novo or from preexisting ameloblastic fibroma has been well documented. Ameloblastic fibrosarcoma occurs in a wide age range, from 3 to 83 years (mean, 27.3 years), but no case of this tumor has been reported in infancy. The male to- female ratio is 1.6:1. The most commonly affected site
within the jaw is the posterior part of the mandible. We discuss such a case of ameloblastic fibrosarcoma originating de-novo in the posterior region of mandible.

**Case Report**

A 26-year-old woman reported with a swelling of the left side of the mandible since 1 year. The swelling was well defined, with a smooth surface, non-tender and bony hard on palpation along the lower border of mandible and soft along its superior aspect. There was no palpable bruit or pulsations noted (Figure 1A).

Intraoral examination revealed a fungating, large, ulcerocarcinomatous lesion extending from the left angle of the mouth posteriorly till the retromolar area and extends lingually pushing the associated teeth and the tongue to the right side. The surface of the ulcer is covered with white-grey slough and anteriorly it is erythematous. The surface shows indentations of the maxillary teeth. The swelling was non-tender, with no bleeding or pus discharge on digital pressure (Figure 1B).

Panoramic view revealed an ill defined radiolucency in the left body region of the mandible showing destruction of the mandible with displacement of teeth 34, 35 and 37 and 38. Two radio-opaque masses are seen in the superior aspect of the radiolucency in the region of 36 and 38 with a radio-opaque scalloping at the lower border of the mandible (Figure 2A). Computerized tomographic scan showed solid mass destroying bone in an irregular fashion on left mandible with lateral displacement of hyoid bone (Figure 2B).

Hemi-mandibulectomy was carried out and the specimen was sent for histopathological examination. Light microscopy revealed irregularly arranged strands of islands of odontogenic epithelium surrounded by abundant mesenchymal tissue with the feature of fibrosarcoma (Figure 3). The fibrosarcoma cells were strongly positive on immunostain for vimentin (Figure 4) and ameloblastic cells were weakly positive for cytokeratin (Figure 5). S-100 and CEA were negative in both epithelial and sarcoma cells.

Based on the history, clinical examination and investigations a diagnosis of ameloblastic fibrosarcoma was made. Our patient received radiation therapy, 50 Gy in divided doses (150 cGy daily), after surgical removal of the tumor. During 4.5 years' follow-up, no recurrence occurred.
Ameloblastic fibrosarcoma (AFS) is a very rare tumor, and not a great deal of its morphologic diversity is known. A characteristic feature of AFS is a biphasic pattern comprising various amounts of the epithelial and mesenchymal components. Until now, 64 cases have been reported in the English literature. Most of them have occurred in the mandible and in the third decade of life. The present case was seen in the posterior mandible in a second decade female patient. A case has been reported wherein AFS mimicked adenoid cystic carcinoma. A state of confusion prevailed in differentiating AFS from odontogenic carcinosarcoma. In a study it was affirmed that odontogenic carcinosarcoma may be treated as an individual entity.

Clinical appearance and symptoms vary among the reported cases, but swelling and pain are the most constant findings. A rare case of AFS involving the anterior and middle skull base with intradural extension has been reported.

Ameloblastic fibrosarcoma is a locally aggressive tumor and radical resection is indicated. Metastases are not unusual. This tumor can be confused with ameloblastic fibroma and/or ameloblastoma. However, the distinction is easily done by paying attention to the cytological characteristics of the malignant mesenchymal spindle cells of ameloblastic fibrosarcoma that show frequent mitotic figures. Some of them also show aberrant, variation in nuclear chromatism and anisonucleosis.

The World Health Organization distinguishes odontogenic sarcoma devoid of dental hard tissue (AFS) from those displaying focal evidence of dentinoid (ameloblastic fibrodentinosarcoma) or dentinoid plus enameloid (ameloblastic fibroodontosarcoma),
but the World Health Organization panel acknowledges that presence or absence of dental hard tissue in an odontogenic sarcoma is of no prognostic significance.\textsuperscript{11}

A study was done to observe the interstitial cells of ameloblastic fibroma (AF) and ameloblastic fibrosarcoma (AFS) and elucidate their biological characters. The study concluded that decreased or lack of epithelia may be associated with excessive hyperplasia in AFS.\textsuperscript{12}

The information available concerning the treatment, course, and prognosis of AFS is limited because of the paucity of cases reported. According to previous reports, 20% of patients have died within 3 months to 19 years, resulting from locally aggressive tumor growth.\textsuperscript{5}

A rare case of anaplastic AFS has also been reported, where an AFS developed from ameloblastic fibroma 10 years after the primary excision.\textsuperscript{13}

In view of the local aggressiveness and its high tendency to recur (37% of the reported cases have had at least 1 recurrence), the treatment of choice is wide surgical excision with long-term follow-up. Because documented metastasis has been reported only once in the literature, routine neck dissection for AFS seems to be unnecessary.\textsuperscript{7}

AFS has been treated with chemotherapy as well. Two pediatric cases have been reported of chemosensitive AFS.\textsuperscript{14}

Our patient received radiation therapy, 50 Gy in divided doses (150 cGy daily), after surgical removal of the tumor. During 4.5 years’ follow-up, no recurrence occurred. Adjuvant postoperative radiotherapy, as in our patient, was used successfully in a case with a very extensive maxillary lesion\textsuperscript{3}. We recommend adding radiation to treatment of this tumor for prevention of recurrence, especially in cases of incomplete surgical removal.

References

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