



## A CASE OF AMELOBLASTIC FIBROSARCOMA RAKESH CHANDRU K

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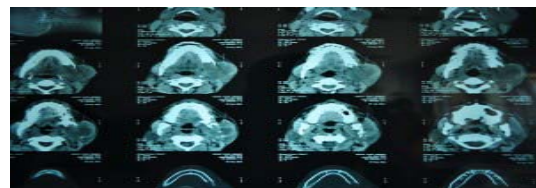
**Abstract** : Ameloblastic fibrosarcoma (AFS) is a rare odontogenic tumor comprising a benign epithelial component and a malignant ectomesenchymal component. AFS is a neoplasm with a similar structure to the ameloblastic fibroma, but in which the mesodermal component shows the features of a sarcoma. Here we present a case of ameloblastic fibrosarcoma of mandible in a 32 year old male patient for whom the pre operative incisional biopsy was diagnosed as ameloblastic fibroma. In histopathology of resected specimen some mesenchymal areas showed distinct cellular pleomorphism of an anaplastic sarcoma. So this may be due to recent transformation of ameloblastic fibroma to fibrosarcoma.

**Keyword** :Ameloblastic fibrosarcoma, ameloblastoma, odontogenic tumors.

A 32 year old male patient came with complaints of swelling in the left side of his face in the mandibular region for about 5 months duration. Initially it was very slowly progressive but for last 1 month there was a rapid increase in size. There was history of pain and difficulty in opening the mouth. On examination there was a hard diffuse swelling of size 8 \*6 cm at the mandibular region left side. Mobility has been restricted. Other clinical examinations of head and neck were normal.



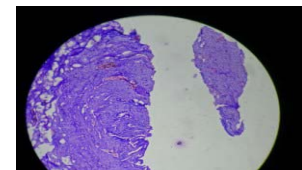
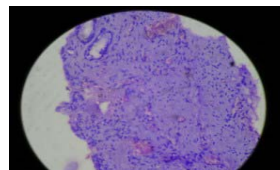
The CT scan of mandible showed ill-defined lytic lesion with soft tissue component in the body and ramus of left mandible extending to third molar alveolar margin and subcondylar region.



Pre operative incisional biopsy showed features of ameloblastic fibroma. Then total excision of lesion with left hemimandibulectomy with mandibular reconstruction was performed.



Histopathological study showed cellular neoplasm composed of round to polyhedral and spindle cells with round to ovoid nuclei, some with vesicular nuclei, prominent macronuclei, nuclear pleomorphism, atypia, and significant mitosis arranged in hypo and hyper cellular pattern. Focal area disclosing peritheliomatous arrangement of pleomorphic cells, areas of necrosis and hemorrhage seen.



Immune histochemistry shows cytokeratin and vimentin positive, S-100 negative. With these

impressions of ameloblastic fibrosarcoma was given. Post operatively radiotherapy given and patient is on regular follow up.

#### REVIEW OF LITERATURE

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. Most of them have occurred in the mandible and in the third decade of life. They may arise de novo or develop by malignant transformation of their benign counterparts like ameloblastic fibroma or ameloblastic fibroodontoma. So sufficient sampling from pathological specimen is necessary not to misdiagnose a malignant lesion since malignancy might have started in focal areas. This also explains the possibility of error in incisional biopsy findings. 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. Assessment of proliferative activity by immunohistochemistry or DNA ploidy has been helpful in differential diagnosis between AFS and ameloblastic fibroma. Clinical appearance and symptoms vary among the reported cases, but swelling and pain are the most constant findings. Ulceration, bleeding and paresthesia may be additional local findings. Pain can also precede the swelling. A rare case of AFS involving the anterior and middle skull base with intradural extension has been reported. Metastases are not unusual. Ameloblastic fibrosarcoma is a locally aggressive tumor and radical resection is indicated. The information available concerning the treatment, course, and prognosis of AFS is limited because of the paucity of cases reported. In view of the local aggressiveness and its high tendency to recur, 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. Adjuvant radiotherapy has a role in the treatment of this tumor for prevention of recurrence, especially in cases of incomplete surgical removal.

#### CONCLUSION

Ameloblastic fibrosarcoma (AFS) or ameloblastic sarcoma is an extremely rare odontogenic neoplasm. This tumor can be confused with ameloblastic fibroma and/or ameloblastoma. It is important to recognize variable features of ectomesenchymal components of AFS to provide a proper diagnosis and treatment. Ameloblastic fibrosarcoma is a locally aggressive tumor and radical resection is indicated. Adjuvant radiation therapy has a role in the management.

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