AMELOBLASTIC FIBROSARCOMA OF THE MANDIBLE - A POSSIBLE SARCOMATOUS TRANSFORMATION OF AN AMELOBLASTIC FIBROMA

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Ameloblastic fibrosarcoma is a rare odontogenic neoplasm regarded as the malignant counterpart of ameloblastic fibroma. It is composed of a benign epithelial component and a malignant mesenchymal component. Sometimes, the epithelial cells may be overgrown by the malignant mesenchymal cells and thus make diagnosis difficult. We report a case of a 26 year old female patient diagnosed on incisional biopsy as an ameloblastic fibroma but excision of the specimen revealed an ameloblastic fibrosarcoma with paucity of the epithelial component. The histopathology of the tumour revealed the possibility that the sarcomatous transformation of the mesenchymal component may have led to depletion of the epithelial component.

Key words: Ameloblastic fibrosarcoma, Ameloblastic fibroma, sarcomatous transformation.

Ameloblastic fibrosarcoma (AFS) is an extremely rare malignant odontogenic tumour which exhibits a biphasic pattern histologically. It is composed of both a benign odontogenic epithelial component resembling an ameloblastoma and a malignant mesenchymal component with features resembling a fibrosarcoma.[1] These tumours are known to either occur de novo or from a malignant transformation of an ameloblastic fibroma (AF)[2]. Sometimes the epithelial component may be overgrown by the malignant mesenchymal cells making diagnosis difficult. Here, we report a rare case of an AFS in a young female patient where we found both areas of benign and malignant mesenchymal components with scanty epithelial islands.

CASE REPORT

A 26 year old female patient presented with a complaint of a slow growing swelling of the left lower jaw since two years. Extraoral examination revealed profound asymmetry of the face due to an ill defined jaw mass involving the left posterior half of the mandible. (Fig. 1) Overlying skin was found to be normal. Intraorally, there was a large, firm, expansile bony mass causing obliteration of the buccal vestibule and displacement of the teeth towards the floor of the mouth. The swelling was erythematous and slightly tender on palpation. The patient's history revealed an incisional biopsy done 2 years back which was reported as an ameloblastoma . General physical examination and other laboratory tests were found to be normal. Panoramic radiograph showed an ill defined radiolucency present in the body of the mandible and extending into the ramus. The lesion had destroyed most of the mandible of the left side. (Fig. 2)

An incisional biopsy of the mass revealed odontogenic islands consisting of peripheral columnar cells exhibiting hyperchromatic nucleus and reversal of polarity along with a vacuolated cytoplasm. Centrally stellate reticulum like cells were seen. The intervening tissue was ectomesenchymal in nature with plump, stellate and ovoid cells in a loose matrix, with no evidence of atypia. (Fig. 3) A moderate chronic inflammatory infiltrate was present. A diagnosis of Ameloblastic Fibroma was arrived upon. But as the clinical and radiographic evidence suggested a highly destructive lesion, a hemimandibulectomy was carried out along with supraomohyoid neck dissection as few enlarged lymph nodes were found on surgery.

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Microscopic examination of the specimen revealed a highly cellular lesional tissue composed of ovoid, round, spindle shaped fibroblast like cells arranged in fascicles and whorls. These cells showed features of malignancy like cellular and nuclear pleomorphism, increased mitosis (both normal and abnormal) and varied nuclear hyperchromasia. (Fig. 4) Areas of increased vascularity and necrosis were seen. A transition of benign ectomesenchymal tissue to a full fledged sarcoma was seen at places. (Fig.5) However repeated serial sections did not reveal any odontogenic islands. Keeping in mind that the incisional biopsy showed ameloblastic islands, a final diagnosis of AMELOBLASTIC FIBROSARCOMA was given.

The lymph nodes were found to be reactive with no evidence of metastasis. The patient was given a dental prosthesis and recurrence has not been reported even after a period of 18 months.

DISCUSSION

Ameloblastic fibrosarcoma is a rare malignant odontogenic tumour of mixed origin wherein the epithelial component is bland and the ectomesenchymal tissue shows malignant changes. Clinically ameloblastic fibrosarcoma presents as a painful intraosseous mass most commonly in the posterior region of the mandible.[2] Occurrence is more common in males(ratio of 2:1) and the average age of presentation is 27.3 years.[2] The present case was that of a 26 year old female patient.

Two third of the reported cases have arisen de novo and the remaining cases have formed due to malignant transformation of an ameloblastic fibroma.[2] The difference in the mean age of diagnosis of ameloblastic fibroma (15-22 years) and ameloblastic fibrosarcoma (27.3 years) supports a possible stepwise progression from a benign to a malignant neoplasm.[1]

In the present case, the incisional biopsy revealed a diagnosis of ameloblastic fibroma because the mesenchymal tissue was devoid of any cellular atypia. However, the excisional biopsy exhibited highly malignant features like pleomorphism, hyperchromatism and mitotic figures in the mesenchymal tissue almost like a fibrosarcoma. In fact a transition from benign mesenchymal areas to malignant mesenchymal areas was seen. This is a very interesting finding and provides visual proof of the possibility of the patient previously having ameloblastic fibroma which underwent sarcomatous transformation.

Another exciting finding was the paucity of epithelial islands in the excised specimen. If it were not for the incisional biopsy which revealed ameloblastic islands, diagnosis would have been difficult. It has been noticed by some authors that greater the anaplasia of the mesenchymal component of AFS, lesser the occurrence of benign odontogenic epithelium.[3,4] Park suggested that the degeneration of the benign odontogenic component was because of the overgrowth of the malignant mesenchymal tissue.[4] Using MIB-1, Sano et al demonstrated a high proliferative potential in the mesenchymal cells of ameloblastic fibrosarcoma.[5] Williams also demonstrated differences in p53 distribution in areas resembling ameloblastic fibroma and the more malignant areas.[6] Since the reported case was highly anaplastic, these factors could explain the paucity of odontogenic epithelium seen. This has led some authors to speculate that some cases which were previously reported as fibrosarcomas of the jaws could actually be odontogenic in origin.[7] Hayashi has put forth a pathologic concept that the gradual loss of odontogenic epithelium can be correlated with the clinical malignant behaviour of AFS and suggested that AFS with total or partial loss of the epithelial component should be considered highly malignant and be treated as a sarcoma.[3]

Inspite of the anaplastic histological features of this neoplasm metastases has been reported in only one case in contrast to that of a conventional fibrosarcoma.8 It has been suggested that the epithelial component exhibits an organisational effect over the mesenchymal component and thus reduces the ability of cells to metastasise. In the present case

too, lymph nodes were found to be free of any malignant cells.[9]

The information related to the treatment and prognosis of this tumour is limited owing to its rarity although wide surgical excision and close follow up appear to be the treatment of choice.[10] In the present case the patient has been treated with hemimandibulectomy and has been kept on regular follow up.

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Fig. 1: Swelling involving the posterior left mandible

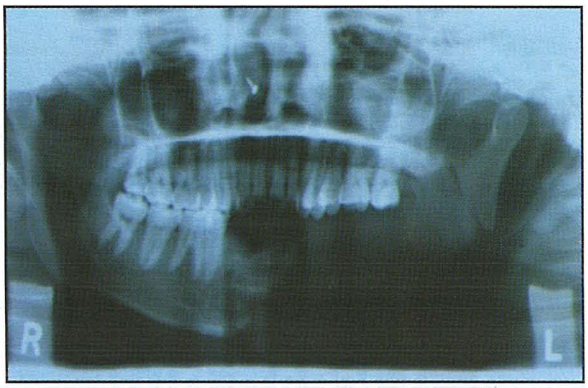


Fig. 2: Orthopentamogram showing destruction of the left side of the mandible.

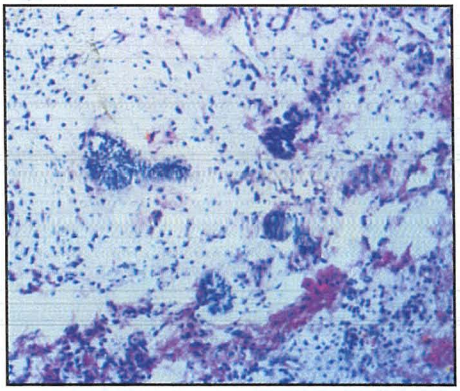


Fig. 3: Ameloblastic islands in a benign mesenchymal stroma (H-E staining x 10)

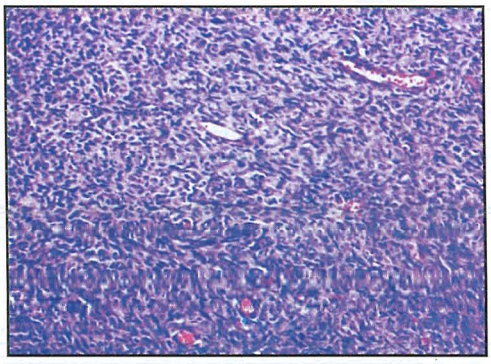


Fig. 4: Highly cellular areas of spindle cells arranged in fascicles, exhibiting pleomorphism, varied hyperchromatism and mitotic figures. (H-E staining x 20)

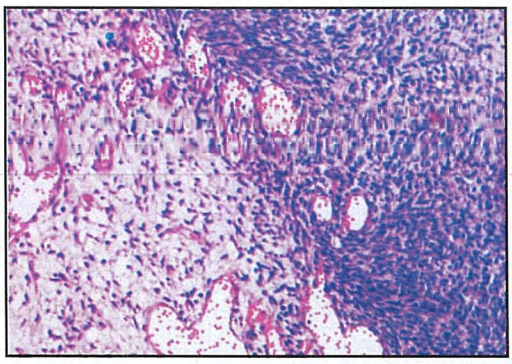


Fig. 5: Areas of transition from benign mesenchymal tissue to sarcomatous tissue . (H-E staining x 20)

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