AMELOBLASTIC FIBRODENTINOSARCOMA- A CASE REPORT OF AN UNCOMMON ODONTOGENIC SARCOMA.


ABSTRACT

Odontogenic sarcomas are rare malignancies of the jaws, which includes ameloblastic fibrosarcoma (AFS), ameloblastic Fibrodentinosarcoma (AFDS), ameloblastic Fibrodontosarcoma (AFOS), and odontogenic carcinosarcoma (OCS). These sarcomas composed of a benign epithelium and malignant ectomesenchymal component. We report a rare case of AFDS in a 17 year girl which rapidly expanded to perforate cortical plates of right side of the mandible.

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INTRODUCTION

The ameloblastic fibrosarcomas (AFS) are a group of malignant odontogenic neoplasm composed of a completely benign epithelial element scattered throughout malignant mesenchymal tissue. Ameloblastic Fibrodentinosarcoma (AFDS) is a neoplasm similar to AFS, but in which limited amount of dysplastic dentine (dentinoid) is formed.1, 2, 4-9 In these tumours parts of the odontogenic epithelium have exerted sufficient inductive influence to result in dentin formation followed by the deposition of enamel matrix, even though the mesodermal component is sarcomatous.1-3 AFS has sometimes been divided into AFDS and ameloblastic fibrodontosarcoma (AFOS); however, many authors consider these tumors to be histologic variants of the same basic neoplasm.

Altini et al. suggested that these malignant tumors should be referred to as
ameloblastic fibrosarcoma to simplify a confusing nomenclature, because the histologic subtypes of ameloblastic fibrosarcoma do not vary in their biologic behavior.\(^2\) However, in the most recent World Health Organization (WHO) 2005 classification of odontogenic tumors, the AFDS and AFS are listed separately from the AFS. The review of literature shows that incidence of AFDS is considerably less than ameloblastic fibrosarcoma.\(^3\)

**CASE REPORT**

We report a 17-year-old female presented with a painless swelling on the right lower third of the face. The swelling had been noticed four months previously which enlarged to the present size. (Fig. 1)

![Fig 1: Presence of swelling in the right lower third of the face.](image1)

Intra oral examination revealed a firm swelling in the right gingivo-buccal region extending from the distal aspect of second premolar to retromolar region, the permanent molars were absent (Figure 2).

![Fig 2: Intra oral swelling seen in the right vestibular area](image2)

Radiographs showed large radiolucency in the body and ramus of mandible with scattered radio opacities. Mandibular 1st permanent molar was impacted and pushed to the lower border of the mandible, no evidence of formation of second and third molar was noted (Figure 3).

![Fig 3: Orthopantomogram](image3)

A 3D CT scan showed expansile mass which perforated the both buccal and lingual cortical plates (Figure 4).
Clinically it was diagnosed as benign odontogenic tumor. An incisional biopsy was performed and the histopathology picture showed odontogenic epithelium arranged in the form of follicles and strands scattered in the highly cellular ectomesenchyme (Figure 5).

The odontogenic epithelium didn’t show any carcinomatous features. The mesenchyme composed of closely arranged cells with cellular pleomorphism, nuclear hyperchromatism, and numerous mitotic figures, with dentinoid material formation(Figure6&7).

Histopathologically a diagnosis of AFDS was rendered. Partial mandiblectomy followed by surgical reconstruction was planned for this patient.

**DISCUSSION**

AFDS is a rare sarcoma, till date 16 cases of AFDS/AFOS have been reported in literature. 14 cases are documented in WHO’s 2005 Pathology and genetics of head and neck tumours. One case of AFOS has been reported by P.Mainenti et al in 2009 and the recent most case of AFOS by S Wang et al in year 2011. Nine cases are reported in men and six in women. The age range is 12-83 years with a peak in the third decade. We report a
case of 17 year old girl. Clinically, the
degree of malignancy of ameloblastic
fibrosarcomas has been varied. Leider et al
stated that AFS histologically behaves
similar to low-grade fibrosarcoma.\textsuperscript{2,5,6} The
presence of a benign epithelium as well as
a malignant mesodermal component
suggests the possibility that this tumor
may have originally been a benign
ameloblastic fibroma or ameloblastic fibro
dentinoma which underwent sarcomatous
change or it may also arise de novo.\textsuperscript{2,5} The
present case arose de novo; the patient had
no previous history of any other lesion.
Prein et al studied the metastasis and
conclude that ameloblastic fibrosarcomas
are semi malignant because of no record
and suggested the term proliferative
ameloblastic fibroma. Chrome et al
reported an instance with
pleuropulmonary, mediastinal lymph node
and hepatic metastasis.\textsuperscript{2} The present case
was clinically aggressive, although no
metastasis was observed. Radio
graphically, AFDS/AFOS can show a
unilocular or multilocular radiolucency
with poorly defined outlines being
associated with tooth and one or more
dense opacities. However, a case with an
irregular border and expansion and
perforation of the cortical plate should be
interpreted with caution and the possibility
of malignant odontogenic tumour should
be suspected.\textsuperscript{8,10} The present case showed a
multilocular appearance associated with
the right mandible with dense opacities
and an impacted 1st molar. These findings
and the location of the mass highly
suggestive of odontogenic tumour. The
histological pattern of AFDS resembles
AFS in which the epithelial tissue is
benign but the connective tissue
component is malignant. The epithelium
proliferates in the form of strands, cords
and islands. Larger islands have a border
of columnar cells with hyperchromatic
nuclei. A hypercellular connective tissue
stroma displaying mitotically active cells
surrounds the epithelial component.\textsuperscript{7,10} A
similar histologic picture was seen in the
present case also. Multiple samples were
taken from the resected mass which
showed only dentinoid mass, no enamel
was found. Poorly differentiated tumors
tend to show greater stromal cellularity
with a decrease in the epithelial
component. Park et al reported a case of
highly malignant AFS which showed
sheets of more anaplastic and poorly
differentiated mesenchymal cells were not
associated with the benign odontogenic
epithelium, whereas less anaplastic
mesenchymal tissue was closely associated
with it. They suggested that the anaplasia
of mesenchymal tissue is correlated with
the degeneration of benign odontogenic
epithelium, and that the loss of benign
odontogenic epithelium results from
overgrowth of the malignant mesenchymal
portion of the lesion.\textsuperscript{2,7,8}

The study of Ki-67, p53 and PCNA showed
immunelocalization in the connective
tissue of the AFS, revealing a high growth
profile in the mesenchymal component,
which was not evident in AF.\textsuperscript{9}

The histopathological findings were
characteristic and no
immunohistochemical staining was done
for the present case. Partial mandiblectomy
with preservation of the condyle was
performed followed by facial
reconstruction to preserve the patient’s
facial contour, masticatory function. After
six months of close follow up the patient is
doing well with no signs of recurrence and
metastasis.

CONCLUSION

Information available concerning the
behaviour, treatment, course and prognosis
of AFDS/AFOS is limited because of
inadequate cases reported. An extensive
sampling of resected mass is advised to
check the formation of dental hard tissue
to render a specific diagnosis of
AFDS/AFOS. It is likely that AFDS
behave in a similar manner to an
ameloblastic fibrosarcoma. A regular
follow up is required to check recurrence
and metastasis.

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