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CASE REPORT

Ameloblastic fibroodonto sarcoma – An extremely rare case report with clinical and radiological insights

Sivan Sathish*, Upender Malik, Haritma Nigam, Shilpi Srivastava, Tabiha Khan, Vaishali Malik

Department of Oral Medicine and Radiology, Teerthanker Mahaveer Dental College and Research Centre, Teerthanker Mahaveer University, Moradabad, Uttar Pradesh, India

Background: Ameloblastic fibro-odontosarcoma (AFOS) is a rare malignant odontogenic tumor with benign epithelial and malignant mesenchymal components. It typically affects younger individuals, with a predilection for the posterior mandible. With fewer than 25 cases reported globally, it remains a diagnostic challenge due to overlapping features with other odontogenic lesions and limited documented information.

Case presentation: This is a case of a 21-year-old male who presented with a progressively enlarging swelling on the right side of his face, painful growth in the posterior mandible, and numbness in the right lower lip. Clinical examination revealed a bony hard swelling with an ulcerative lesion in the mandibular alveolar mucosa. Cone-beam computed tomography showed a heterogeneously hypodense, expansile lesion with bicortical expansion, cortical erosion, and hyperdense masses resembling enamel and dentin. Histopathological analysis confirmed AFOS, showing benign odontogenic epithelial islands within a malignant mesenchymal stroma. The patient underwent surgical resection with wide margins, followed by radiotherapy and chemotherapy to address metastatic lymph node involvement.

Conclusion: This case highlights the diagnostic complexities and aggressive behavior of AFOS. A multidisciplinary approach integrating clinical, radiographic, and histopathological findings is critical for accurate diagnosis and effective management.

Keywords: ameloblastic fibroodontosarcoma, sarcoma, odontogenic tumors

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Introduction

Malignant odontogenic neoplasms are rare, accounting for only about 1-2% of all odontogenic tumors. These neoplasms are categorized into carcinomas and sarcomas, each presenting unique clinical and histopathological features and biological behaviors. Among them, ameloblastic fibro-odontosarcoma (AFOS) is an extremely rare, locally aggressive tumor classified under odontogenic sarcomas by the World Health Organisation. Ameloblastic fibro-odontosarcoma is a biphasic malignant neoplasm comprising benign epithelial elements and a malignant mesenchymal component that produces odontogenic hard tissues, specifically dentin and enamel matrix [1]. This odontogenic sarcoma subtype can either arise de novo or develop through malignant transformation of a pre-existing benign ameloblastic fibro-odontoma [2].

Ameloblastic fibro-odontosarcoma occurs in younger individuals, with a predilection for the posterior mandible and an age distribution skewed towards the second and third decades of life. Due to its rarity and atypical presentation, AFOS poses considerable diagnostic challenges for clinicians. The limited pathological information available and the lack of standardized diagnostic criteria further complicate its identification. To date, not more than 25 cases of AFOS have been documented worldwide, highlighting the extreme rarity of this condition and the need for additional case reports to enhance understanding of its clinical, radiographic, and histopathological features [3-5].

* Correspondence to: Sivan Sathish
E-mail: sivansathishmfds@yahoo.co.in

Case Presentation

A 21-year-old male patient presented to the Department of Oral Medicine and Radiology with a chief complaint of a progressively enlarging swelling on the right side of his face and a painful growth in the right lower back tooth gum region, which had been present for the past three months. Three months prior, the patient noticed a slight swelling on his right cheek, which slowly increased in size. The swelling was initially painless. Two months before the consultation, a mobile right lower back tooth (the seventh tooth from the midline) exfoliated on its own. Following the tooth's exfoliation, the patient noticed a small, painful growth in the gum region. The pain was moderate, continuous, and throbbing, radiating throughout the jaw. It was aggravated by eating and talking. For the past month, the patient also experienced a loss of sensation in his right lower lip.

On extraoral examination, the face appeared asymmetrical due to a dome-shaped swelling on the right side of the face. The swelling was noted in the right side angle of the mandibular region. The skin over the swelling appears to be normal in color and texture. The swelling extends superiorly from 1 cm below the alar-tragus line and inferiorly extending up to the lower border of the mandible, anteriorly extending from the anterior border of the masseter, and posteriorly extending up to the angle of the mandible. On palpation, the dome-shaped swelling was bony hard in consistency, which was non-tender and demonstrated eggshell crackling. The entire dome-shaped swelling was measured at about 3x5 cm in diameter (Figure 1A).

On lymph node examination, a single palpable lymph node was noted in the right jugulodigastric region. It was

oval in shape, firm in consistency, discrete, non-tender, and immobile, measuring approximately 1x2 cm. The overlying skin was pinchable.

On intraoral examination, a single, punched-out ulcerative lesion was noted in the alveolar mucosa of the 47 region. The ulcer had an undermining edge and everted margins with erythematous and edematous mucosa. The floor of the ulcer was covered with pseudomembranous slough. The ulcer was measuring about 1x2 cm in diameter. The ulcer was severely tender on palpation. Further, there was a dome-shaped bicortical expansion involving the buccal and lingual bone of the molar-ramus region, extending anteriorly distal to 45 and posteriorly up to the retromolar region. The gingiva overlying the dome-shaped swelling was erythematous and edematous. On palpation, all the inspection findings were confirmed. The dome-shaped swelling was bony hard in consistency and moderately tender on palpation (Figure 1B).

Following clinical examination, the patient was advised for radiographic investigations. Cone-beam computed tomography of the right mandible revealed an expansile heterogeneously hypodense lesion in the molar-ramus region. The axial (Figure 2) and coronal sections revealed bicortical expansion of the molar-ramus region with erosion in the buccal cortical plate. The sagittal section revealed impacted 48 that was displaced apically to the lower border of the mandible.

The displaced molar was completely developed, and its CEJ was the point of origin of this heterogeneously hypodense lesion. This heterogenous hypodense lesion extends from the lower border of the mandible to the alveolar margin in relation to 46, 47, and 48. The margin of the expansile heterogeneously hypodense lesion appears scalloped. The internal radiodensity of this hypodense lesion is heterogeneous due to localized multiple hyperdense masses measuring 1mm to 0.5 cm in diameter. These hyperdense masses show varied radiodensities resembling enamel and dentin. The entire heterogeneously hypodense lesion measured about 3x5 cm in diameter.

Gray-scale ultrasonographic investigations of the cervical lymph node revealed a single, discrete, homogenously hypoechoic jugulodigastric lymph node with loss of hilum and transverse diameter measuring about 2 cm in diameter. The nodal margin appears to be sharp (Figure 1c).

As per Sivan's classification of jaw lesions based on 3D visual volumetric analysis (2024), this lesion is described as a homogeneous bicortical hypervolumetric lesion, suggestive of a fibrous tumor with internal calcified components. (Figure 3) [16]. Additionally, 3D volumetric analysis also revealed multiple cortical plate erosions, thus radiographically confirming malignant counterpart of the above lesion.

Based on clinical features, CBCT, and ultrasonographic features, it was provisionally diagnosed as ameloblastic fibro odontosarcoma. Differential diagnosis considered were ameloblastic fibrosarcoma, ameloblastic fibro-odontoma and adenomatoid odontogenic tumor. An incisional biopsy was performed, and microscopic examination confirmed a biphasic tumor with two distinct components: benign odontogenic epithelial islands and a malignant mesenchymal stroma. The epithelial component consisted of islands and strands of ameloblast-like cells, while the mesenchymal component showed marked nuclear pleomorphism, hyperchromatism, and increased mitotic activity. The tumor also produced irregular dental matrix material, diagnostic of AFOS. Immunohistochemistry with cytokeratin 14 and 19 highlighted the odontogenic epithelium and vimentin

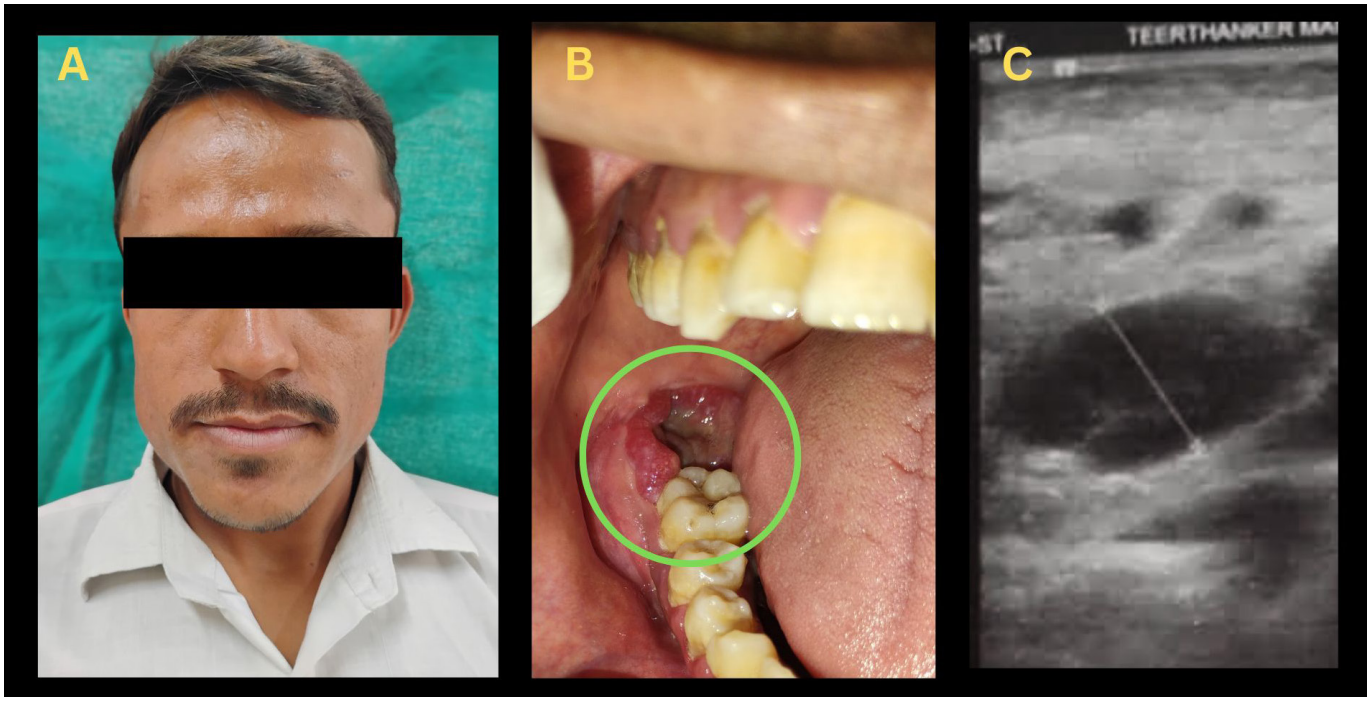

Fig 1. A. Extraoral swelling on left side of the face; B. Intraoral examination revealing a punched-out ulcerative lesion in the alveolar mucosa of the 47 region; C. Ultrasonographic examination revealing a homogenously hypoechoic jugulodigastric lymph node with loss of hilum.

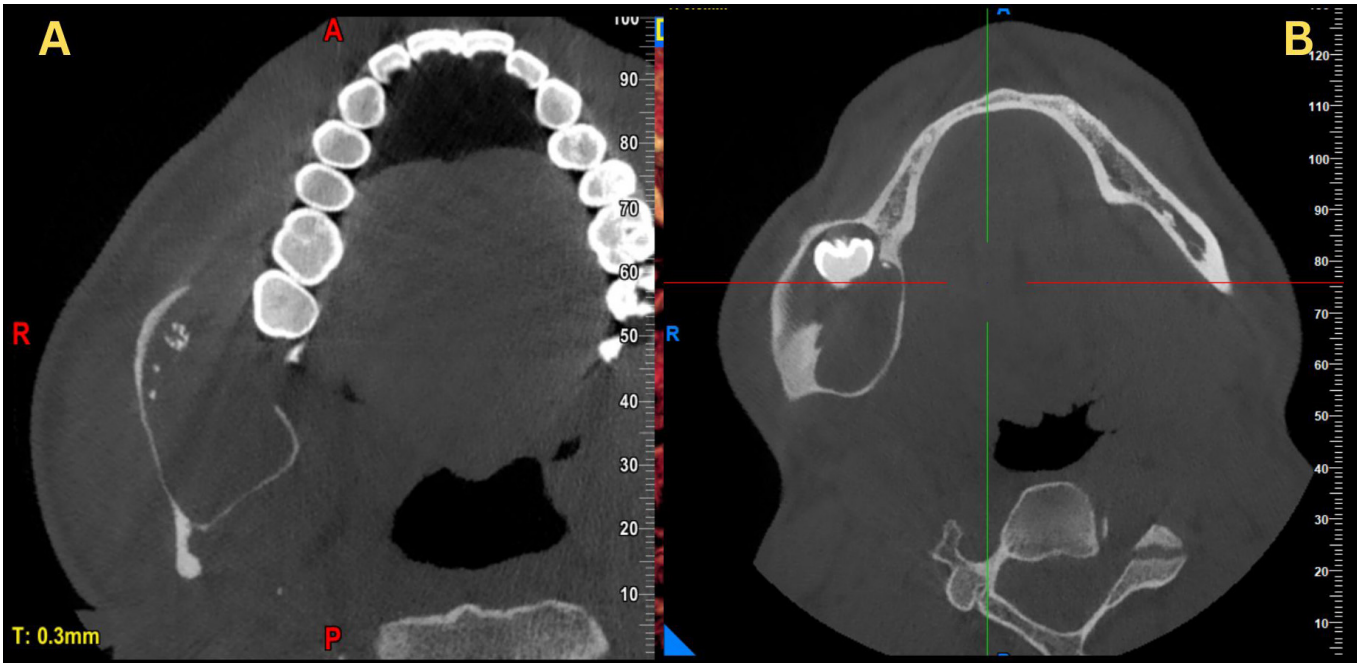

Fig. 2. Axial section revealing bicortically expansile heterogeneously hypodense lesion that is originating from cementoenamel junction of impacted third molar that was pushed apically to the lower of mandible. The radioattenuation of the internal structure of this hypodense lesion is heterogeneous due to multifocal aggregation of hyperdense spicules that shows radioattenuation similar to that of enamel and dentin. These hyperdense spicules sizes ranging from 0.3 to 0.5 mm in diameter and they are discrete and not getting conglomerated with each other. A similar hyperdense structure that is necessarily contacting the distal surface of 46 apical to cementoenamel junction that shows radioattenuation similar to that of enamel and dentin with shape resembling that of tooth structure and measuring about 1 cm in length.

with a high Ki-67 index confirmed the malignant mesenchymal nature.

Thus, based on the clinical features, results of the imaging studies, and histopathological examination, the final diagnosis was ameloblastic fibro-odontosarcoma of the right side posterior mandible. Management of ameloblastic fibro-odontosarcoma requires an intensive, multidisciplinary strategy. In this case, segmental mandibulectomy with wide surgical margins, along with ipsilateral modified radical neck dissection was recommended to address

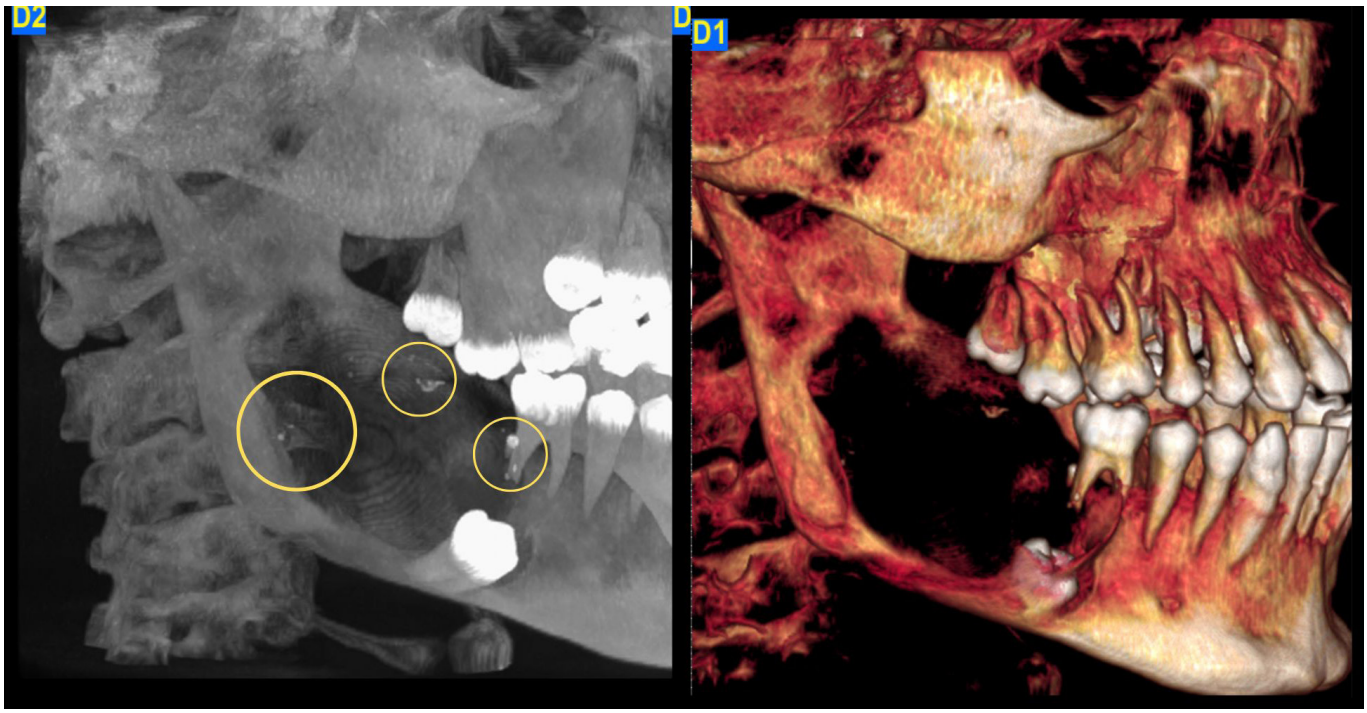

Fig. 3. 3D reconstructed section revealing bicortically expansile heterogeneously hypodense lesion that is originating from cementoenamel junction of impacted third molar and pushed apically to the lower of mandible. The radioattenuation of the internal structure of this hypodense lesion is heterogeneous due to multifocal aggregation of hyperdense spicules that shows radioattenuation similar to that of enamel and dentin.

metastatic jugulodigastric lymph node involvement. The mandibular defect was reconstructed using a reconstruction plate. Postoperatively, adjuvant external beam radiotherapy for 6 weeks combined with systemic chemotherapy was recommended. At present, the patient's postoperative examinations show satisfactory healing, with no evidence of recurrence, and he remains under close follow-up with periodic imaging and clinical evaluations.

Discussion

Ameloblastic fibro-odontosarcoma (AFOS) is an exceptionally rare odontogenic malignancy, with fewer than 25 cases documented in the English literature. The 2005 WHO Classification of Odontogenic Tumors categorizes odontogenic sarcomas into ameloblastic fibrosarcoma (AFS), which lacks dentin and enamel matrix, and ameloblastic fibro-odontosarcoma/fibrodentinosarcoma (AFOS/AFDS), which includes them. The rarity of AFOS underscores the diagnostic and therapeutic complexity of this lesion [1–4].

AFOS commonly presents as a painful swelling, with symptoms ranging from two months to over ten years. The average age at diagnosis is 23.5 years, though it has been reported in patients aged 4 to 83 years. There is a slight male predominance (M:F = 1.2:1). It primarily affects the mandible—especially the retromolar and ramus regions—with only one known maxillary case. The tumor may arise de novo or through malignant transformation of a pre-existing ameloblastic fibro-odontoma (AFO) [5–6].

Radiographically, AFOS often appears as a unilocular or multilocular expansile radiolucent lesion with poorly defined margins and focal radiopaque elements. Cortical perforation or ill-defined borders raises suspicion for malignancy. Histologically, AFOS is characterized by benign epithelial components resembling the enamel organ (including reverse polarization and stellate reticulum-like areas) interspersed in a malignant mesenchymal stroma with marked nuclear pleomorphism, hyperchromatism, and increased mitoses.

In the present case, clinical and radiographic evaluations played a pivotal role in establishing the diagnosis and ruling out differential diagnoses. Although unicystic ameloblastoma was initially considered due to the lesion's location and association with impacted teeth, specific clinical and imaging features helped differentiate it. Clinically, our case showed an ulcerative lesion with everted borders in the alveolar mucosa—an uncommon presentation in unicystic ameloblastoma, which typically presents without mucosal ulceration. Radiographically, CBCT revealed a heterogeneously hypodense, multicystic lesion with scalloped borders and multiple hyperdense, tooth-like masses—unlike the well-defined, unilocular, uniformly hypodense lesions typically seen in unicystic ameloblastoma [7].

Adenomatoid odontogenic tumor (AOT), another benign odontogenic tumor, usually occurs in the anterior maxilla and rarely affects the mandibular molar-ramus area, which was involved in our case. Furthermore, soft tissue ulceration is atypical for AOT. Radiographically, AOTs present as hypodense expansile lesions that engulf the impacted tooth entirely and may contain small, scattered radiopacities (<0.5 mm) that do not resemble teeth. In contrast, AFOS exhibits larger, tooth-like hyperdense structures, and the lesion originates around the CEJ without engulfing the entire tooth, further ruling out AOT [8–9].

Ameloblastic fibrosarcoma (AFS), a malignant counterpart of ameloblastic fibroma, shares similarities with our case in terms of swelling and soft tissue involvement. However, the absence of tooth-like radiopaque structures in AFS distinguishes it radiographically from AFOS [10].

Ameloblastic fibro-odontoma (AFO), though benign, presents with radiographic features resembling AFOS—expansile, heterogeneously hypodense, multicystic lesions with scalloped borders and tooth-like radiopacities. However, AFO typically lacks clinical features such as mucosal ulceration and everted borders, which were seen in our case, thus supporting a malignant diagnosis [11].

The presence of aggressive features, including mucosal ulceration, soft tissue invasion, and a palpable fixed jugulodigastric lymph node, further supported a diagnosis of malignancy. These features were inconsistent with benign odontogenic tumors such as unicystic ameloblastoma, AOT, and AFO.

Among malignant odontogenic tumors, AFOS and AFS were primary considerations. However, the presence of radiopaque, tooth-like structures strongly favored AFOS over AFS [12].

AFOS is classified as a low- to intermediate-grade malignancy, generally exhibiting local aggressiveness rather than widespread metastasis. Recurrence is reported in approximately 10% of cases. Rare instances of lung metastasis and a single reported case of local metastasis emphasize the tumor's unpredictable nature. Given its rarity, diagnosis and treatment require a multidisciplinary approach. While radiographic and clinical evaluations are vital, histopathological confirmation is essential. Radical surgical resection with clear margins remains the treatment of choice. Long-term follow-up is crucial to monitor for recurrence or potential metastasis [13–15].

Conclusion

This case report contributes valuable insights to the growing body of knowledge on AFOS, enhancing our understanding and supporting the development of evidence-based diagnostic and therapeutic strategies for this rare tumor.

Authors' Contribution

SS (Formal analysis; Investigation; Methodology; Patient Care; Validation; Visualization; Writing – original draft; Writing – review & editing)

UM (Conceptualization; Data curation; Validation; Visu-

alization; Writing – original draft; Writing – review & editing)
HN (Conceptualization; Data curation; Validation; Visualization; Writing – original draft; Writing – review & editing)
ShS (Conceptualization; Data curation; Validation; Visualization; Writing – original draft; Writing – review & editing)
TK (Data curation; Formal analysis; Investigation; Methodology; Patient Care; Validation; Visualization; Writing – review & editing)
VM (Data curation; Formal analysis; Investigation; Methodology; Patient Care; Validation; Visualization; Writing – review & editing)

Conflict of Interest
None to declare.

Ethical Statement
The patient has given permission to publish this case report.

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