Indian Journal of Surgical Oncology (March 2019) 10(1):180–183
https://doi.org/10.1007/s13193-018-0843-6

CASE REPORT

# Ameloblastic Fibrosarcoma in Pregnancy: an Unreported Entity!

Suvendu Maji[1,2] · Indranil Ghosh[1]

Received: 16 June 2018 / Accepted: 23 November 2018 / Published online: 28 November 2018


**Abstract**
AFS is a rare tumor of odontogenic origin. Less than 100 cases have been reported so far in the literature. Due to its extreme rarity, we do not have clear management guidelines at present. The entire clinical spectrum and natural history of this uncommon malignancy is unknown. Most of the cases present with facial swelling along with varying degrees of oral signs and symptoms. AFS has higher incidence in male and common in third decade of life. However, presentation in female can be unique and may have a different course and outcome then what is known to us. The association of pregnancy and AFS is interesting and has hardly been emphasized before. Pregnancy can dramatically boost the growth of this tumor leading to impending airway obstruction or fatal hemorrhage. We report a case of an 18-year-old pregnant female who presented with rapidly enlarging bleeding mandibular mass and respiratory distress and present the challenges in its surgical management along with it long-term outcome on follow-up. We also discuss and explore the probable role of AFS in context of pregnancy and its oncological outcome.

**Keywords** Ameloblastic fibrosarcoma · Pregnancy · Mandibulectomy · Tracheostomy · External carotid artery

## Introduction

Ameloblastic fibrosarcoma is a rare mesenchymal neoplasm of odontogenic origin. Its exact incidence is unknown. Most of them arise de novo while few of them have been reported to arise from ameloblastic fibromas. They have male predilection. The usual mode of presentation is a mandibular mass with oral paraesthesia or dysphagia. Mandible is the most frequent site of occurrence while maxilla and base of the skull are the rare sites reported in literature. Diagnosis is based on histology and IHC is corroborative. It is a locally aggressive tumor with a tendency to recur locally. However, the outcomes are better when resected with adequate margins. Association of female hormones with certain malignancy is "known." For instance, pregnancy has been known to accelerate solid tumors like breast cancer. However, their role in odontogenic neoplasm has been only speculative. While several case reports linking the role of reproductive hormones and ameloblastoma and ameloblastic odontoid exists in literature, the probable association of AFS and pregnancy has never been scrutinized before. Our case report provides strong credibility to the fact that female hormones can fuel explosive growth in an otherwise slow growing tumor leading to potentially life-threatening complications.

## Case Report

An 18-year-old lady from Sunder ban presented to us in outpatient department with a huge painless facial swelling (Fig. 1) along with bleeding from the tumor and mild respiratory distress. She could barely speak and had difficulty in eating and swallowing. She had no other systemic complaints. She initially had a small nodular swelling in the left gingivobuccal sulcus for 3 years opposite second premolar tooth which she had ignored until last week. Apparently, the swelling grew rapidly in the last couple of days after she was diagnosed to be positive for pregnancy. Her medical records showed no significant past surgical and medical history. She has been married for 1 year and was prima gravidae. She was

✉ Suvendu Maji
drsuvendumaji@rediffmail.com

Indranil Ghosh
ighoshcnci@gmail.com

[1] Department of Surgical Oncology, Chittaranjan National Cancer Institute (CNCI), 37 SP Mukherjee Road, Kolkata, West Bengal 700026, India

[2] Department of Surgical Oncology, Chittaranjan National Cancer Institute, Kolkata, West Bengal 700026, India

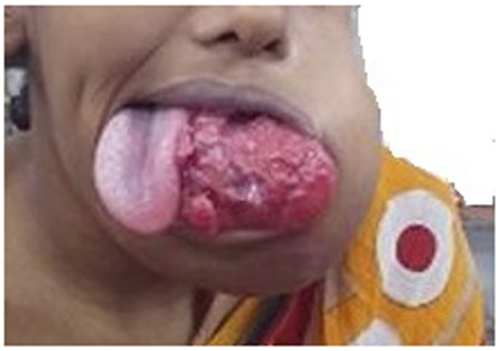

**Fig. 1** Huge mandibular mass occupying the entire mouth

14 days pregnant at the time of presentation. She was admitted on emergency basis for resuscitation. Physical examination was normal. On local examination, a huge swelling was present involving the left lower jaw. Superiorly, it extended from the level of ear lobule and zygoma up to the level of hyoid bone inferiorly. Medially, it extended from the mentum up to the angle of the jaw on the left side. On internal examination, a firm swelling extending from lower midline incisors anteriorly and posteriorly pushing the anterior tonsillar pillar was present. Tongue and floor of the mouth were displaced towards right. Loosening of teeth over the swelling was seen. The globular lump was warm, non-tender and bled to touch. The globular lump had restricted mobility and firm consistency. The skin over the swelling was stretched and shiny. No neck nodes were palpable. FNAC done outside from the lump was inconclusive. The patient condition improved transiently with resuscitation. An OPG done showed left-sided unilocular radiolucent tumor of left hemi mandible with destruction of bone suggestive of ameloblastoma, dentigerous cyst, etc. Color Doppler neck showed high vascularity in tumor along with compression of left external carotid artery and displacement of internal carotid artery posteriorly.

Computed enhanced tomography scan of face neck showed presence of an osteolytic lesion involving the left lower hemi mandible pushing the tongue, floor of the mouth to right in midline, and fauces posteriorly with no invasion of the same. Neck showed no significant lymphadenopathy. Her routine blood investigations were normal. An obstetrical and medical consultation was sought to evaluate her pregnancy. She was counseled about the possibility of spontaneous abortion as a result of surgery. Tracheostomy was done to secure an airway, and external carotid artery was ligated above the superior thyroid artery in view of vascularity of tumor. Tumor was then removed enbloc with overlying adherent skin which eventually resulted in left hemi mandibulectomy. The resulting defect was reconstructed with PMMC (Pectoralis Major Myocutaneous) flap. Neck was left untreated. Postoperatively, she was managed in intensive care unit. The tracheostomy was removed on the 7th postoperative day. Oral feeding was stated after 14 days. She had superficial surgical site infection which was managed with antibiotics and dressings. Macroscopically, the tumor was $16 \times 10.5 \times 6.5$ cm in size and on cut section, it showed presence of white lobulated tumor invading and destroying the mandible. Microscopically, the sections from the growth showed presence of ameloblastic tumor with atypical proliferation of fibroblastic component with frequent mitosis 2/10 HPF. This was consistent with a diagnosis of ameloblastic fibrosarcoma (low) grade. However, three margins of resection were positive. The bone was also involved by tumor. On immunohistochemistry (IHC), tumor was vimentin positive in mesenchymal cells while p53 was positive in glandular component. Her case was discussed in multidisciplinary tumor board and she was advised adjuvant radiotherapy by the tumor board. However, she defaulted and was lost to follow-up. She returned back 15 months later for a checkup. At follow-up, we were surprised to find her healthy and without any evidence of local recurrence. Most importantly, her course of pregnancy had been uneventful in spite of her disease and she gave birth to a normal healthy baby via normal vaginal delivery. She has been in close follow-up for last 5 months (Fig. 2). She is healthy without any evidence of recurrence at the end 3 year post surgery.

## Discussion

Ameloblastic fibrosarcoma is an unusual neoplasm of odontogenic origin. This rare neoplasm was first described by Heath in the year 1887 [1]. These neoplasms have earlier been called as ameloblastic dentinosarcoma while others have termed it as ameloblastic odontosarcoma. However, the World Health Organization (WHO) classification has made it clear that AFS is a different entity from the former two types. The median age at presentation is 30 years while the age range is from 3 to 89 years [2]. Males are affected more than females [3]. The commonest site of affection is posterior part of mandible in the region of molars and premolars. There are varying modes of presentation. Most patients present with mandibular swelling with different grades of paraesthesia or dysthesia [4]. According to the literature, majority of them arise from preexisting ameloblastic fibromas while cases arising de novo have also been reported [5]. Variety of radiological investigations can be used to evaluate the tumor extent and nature; however, none of them are diagnostic. Orthopantogram is a low-cost easily available investigation which often shows a radiolucent and lytic area in the mandible or maxilla. Computed enhanced tomography scan of face neck can characterize the mass and helps in surgical planning and deciding the resectability. Confirmation is by histology only. Different techniques have been used to ascertain the diagnosis. Case reports exist on the use of FNAC and incisional biopsy to

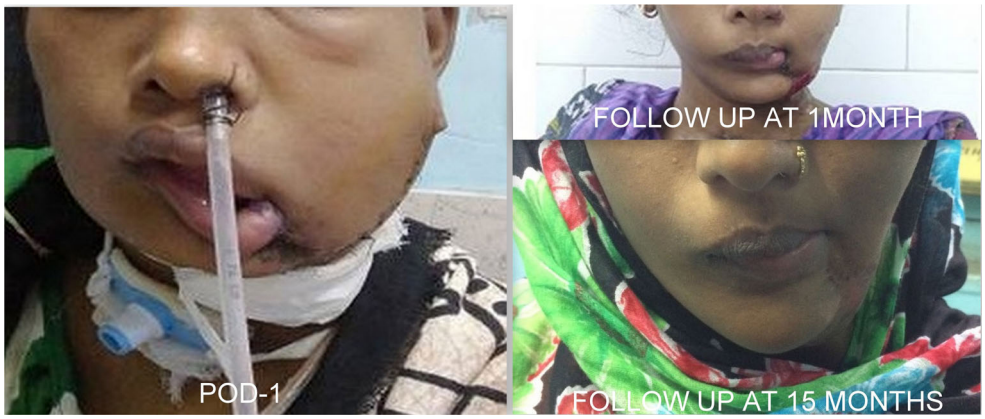


**Fig. 2** Postoperative serial images showing good cosmetic outcome at the end of 15 months

obtain cytological and histological diagnosis. Morphologically, the tumor may be endophytic or exophytic with predominantly solid components. Microscopically, the tumor consists of benign epithelial islands made up of columnar or cuboidal cells arranged in a palisading manner. The stroma consists of spindle-shaped cells which display varying degrees of pleomorphsm, hyperchromasia, and frequent mitoses. The cells are often arranged in storiform or herring bone pattern [6]. Treatment consists of wide surgical excision followed by appropriate reconstruction. There is no role of neck dissection as lymph node metastases are rare. AFS is known for local recurrence, the reported rate being 19% [7]. However, distant metastases occur rarely. The use of postoperative radiotherapy [8] and adjuvant chemotherapy [9] has been described in the English literature, but their use and role is uncertain at this point of time. The relationship of pregnancy with malignancy is well known. However, such an association is not common given its incidence being 0.07 to 0.1% as per western literature. Both benign and malignant neoplasm can affect pregnant females. Pregnancy-associated cancers are rare, the most common being those associated with the reproductive age group. Breast cancers and cervical cancer are the two most common cancers associated with pregnancy. Other cancers reported in pregnancy are lymphoma, leukemia, melanoma, and bone and soft tissue sarcomas [10]. However, occurrence of de novo AFS in pregnancy has never been discussed before. Extensive search of literature revealed a case report describing occurrence of an AFS in a pregnant Nigerian female in the year 2013 [11]. However, unlike our case, the AFS in the former arose from a preexisting ameloblastic fibroma. Ameloblastic fibrosarcoma is a rare malignancy. Only 67 cases have been reported so far in the literature. Association of AFS with pregnancy makes it extremely unique and rare. Our case highlights several practice points and sheds light into its rather unknown natural history. AFS can be quiescent for a long time but can take an aggressive course and lead to life-threatening situation as seen in our case. Although the disease is known for its recurrence, low-grade lesions like ours might be an exception to this thumb rule[12]. The absence of recurrence in spite of positive margins and without any postoperative adjuvant treatment (in form of either radiotherapy or chemotherapy) was peculiar and could not be explained by us. The role of immunity in eliminating the residual cancer cells is the only way this could be explained.

## Compliance with Ethical Standards

**Conflict of Interest** The authors declare that they have no conflict of interest.

## References

1. Reichart PA, Philipsen HP (2004) Odontogenic tumours and allied lesions, 1st edn. Quintessence Publishing, London
2. Williams MD, Hanna EY, El-Naggar AK (2007) Anaplastic ameloblastic fibrosarcoma arising from recurrent ameloblastic fibroma: restricted molecular abnormalities of certain genes to the malignant transformation. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 104(1):72–75
3. Lai J, Blanas N, Higgins K, Klieb H (2012) Ameloblastic fibrosarcoma: report of a case, study of immunophenotype, and comprehensive review of the literature. J Oral Maxillofac Surg 70(8):2007–2012
4. Kousar A, Hosein MM, Ahmed Z, Minhas K (2009) Rapid sarcomatous transformation of an ameloblastic fibroma of the mandible: case report and literature review. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 108(3):e80–e85
5. Muller S, Parker DC, Kapadia SB, Budnick SD, Barnes EL (1995) Ameloblastic fibrosarcoma of the jaws. A clinicopathologic and DNA analysis of five cases and review of the literature with discussion of its relationship to ameloblastic fibroma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 79:469–477
6. Reichar PA, Philipsen HP (2004) Odontogenic tumors and allied lesions. Quintessence Publishing, London
7. Marx RE, Stern D (2012) Oral and maxillofacial pathology: a rationale for diagnosis and treatment, vol 2, 2nd edn. Quintessence, Hanover Park
8. Kobayashi K, Murakami R, Fujii T, Hirano A (2005) Malignant transformation of ameloblastic fibroma to ameloblastic fibrosarcoma: case report and review of the literature. J Cranio-Maxillofac Surg 33(5):352–355

9. Demoor-Goldschmidt C, Minard-Colin V, Cassagneau E et al (2012) Ameloblastic fibrosarcoma of the mandible: report of 2 chemosensitive pediatric cases. J Pediatr Hematol Oncol 34(2): e72–e76
10. Pentheroudakis G, Pavlidis N (2006) Cancer and pregnancy: poena magna, not anymore. Eur J Cancer 42(2):126–140
11. Akinyamoju AO, Olusanya AA, Adeyemi BF, Kolude B (2013) Ameloblastic fibrosarcoma: report of a case. J Oral Maxillofac Pathol 17:424–426