Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology 32 (2020) 57–60

Contents lists available at ScienceDirect

Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: www.elsevier.com/locate/jomsmp

Case Report

Ameloblastic fibrosarcoma—A case report

J.N. Khanna, Radhika Ramaswami*, Kiran Thorat

Consultant Oral and Maxillofacial Surgeon, Saifee Hospital, Mumbai, Maharashtra, India

ARTICLE INFO

Keywords:
Ameloblastic fibrosarcoma
Ameloblastoma
Rare tumours of jaw

ABSTRACT

Ameloblastic fibrosarcoma is an extremely rare mixed odontogenic tumour. It may arise de novo but usually it arises from a pre existing ameloblastic fibroma. The treatment widely accepted for the same is wide excision. We report a case of a 16 year old boy who reported to us with complain of swelling over then left side of mandible extending to right. He gave a history that he was operated elsewhere two years back for a smaller swelling on left side mandible which was reported as ameloblastic fibroma earlier. We confirmed the diagnosis of ameloblastic fibrosarcoma and treated him with wide excision. Patient has been on regular follow up, has no signs of recurrence since almost 7 years and has been rehabilitated.

1. Introduction

The ameloblastic fibrosarcoma (AFS) is a very rare malignant odontogenic tumour composed of benign epithelial element scattered throughout malignant mesenchymal tissue [1]. AFS is a rare mixed odontogenic malignant tumour that was first described by Heath [1–3]. Ameloblastic dentinosarcoma and Ameloblastic odontosarcoma are the variants which show enamel and dentin. According to English literature, 92 documented cases of AFS were found with just 3 of 92 metastatic lesion presentations [1].

AFS can arise from ameloblastic fibroma or can arise as a malignant tumour. In roughly 2/3rd of the cases reported the AFS arises in ameloblastic fibroma and in 1/3rd cases, they arise de novo [2].

AFS is equally distributed in both sexes. AFS is seen in second and third decade of life though literature has given a wide range between 3–89 years [2,3].According to Wood et al, the average age at occurrence of this sarcoma is 26.1 years [4].

AFS is commonly seen in the mandibular body region. Maxillary lesions are rarely seen. AFS is a locally aggressive destructive lesion with a high rate of recurrence in conservatively treated cases. Metastasis is rare but reported in few cases. AFS invades the surrounding tissues and has a low mortality rate.

We report a case where the patient was operated for ameloblastic fibroma at the age of 14 years. He was operated elsewhere and records were not available. After 2 years, he came back with a large extensive lesion which on biopsy confirmed diagnosis of ameloblastic fibrosarcoma.

2. Case report

A male patient, 16 years of age reported to us with swelling involving left parasymphysis and symphysis region crossing midline and extending to right side (Fig. 1). Patient complained of pain and hypoesthesia.

He gave a history that he had a swelling three years back in the left mandibular body region which was painless, slow growing. There was no paresthesia then as mentioned by him. He was operated elsewhere, the lesion was treated with peripheral osteotomy and the diagnosis of ameloblastic fibroma was made. He had no relevant medical history.

On clinical examination, there was a firm, non fluctuant swelling extending from mandibular left premolar to mandibular right canine region. Over lying skin was free from the tumour. Swelling extended into the surrounding soft tissues, the sublingual space; obliteration of the buccal and lingual sulcus. The tongue movements and sensations were normal. Patient reported hypoesthesia over the lower lip and chin. The buccal and lingual cortical bone were perforated at multiple places.

Computed tomography showed a large destructive lesion from right canine to left first molar with buccal and lingual expansion (Figs. 2–4). Bone was perforated at multiple places with destruction of lower border. Biopsy gave the report of ameloblastic fibrosarcoma.

Under general anesthesia, extended submandibular incision was taken, subplatysmal dissection was done. Tumour was exposed, a precontoured reconstruction plate was fixed with screws (Fig. 5). The plate was removed and tumour was resected (Fig. 6). The reconstruction plate was repositioned and fixed with screws. The specimen was sent for frozen section which revealed clear margins.

Histopathological report showed islands of ameloblastic epithelium

* Corresponding author at: Saifee Hospital, Mumbai, India.
E-mail address: radhika.ramaswami@gmail.com (R. Ramaswami).

https://doi.org/10.1016/j.ajoms.2019.06.004
Received 2 November 2018; Received in revised form 28 March 2019; Accepted 11 June 2019
Available online 09 December 2019

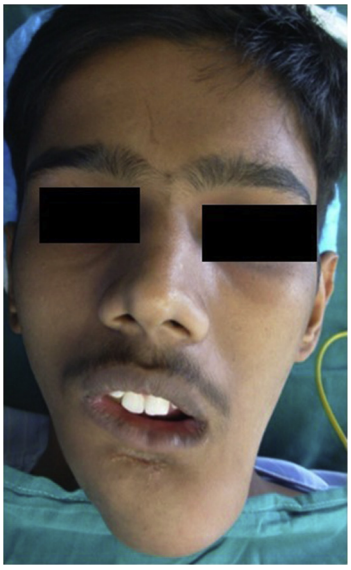

Fig. 1. Extraoral view of the patient.

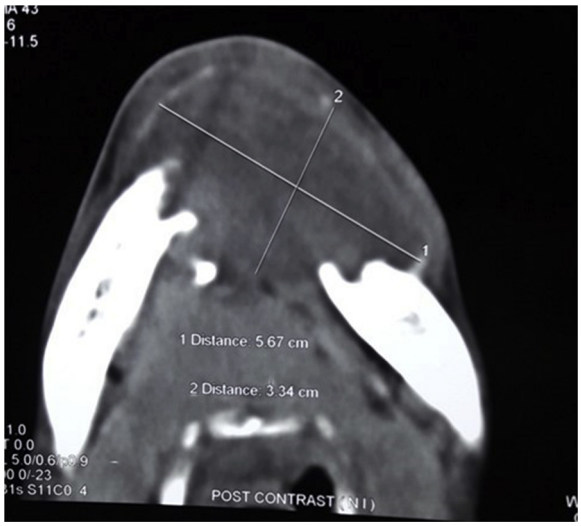

Fig. 2. Axial computed tomography (CT) scan of the patient post contrast revealing the extent of the lesion.

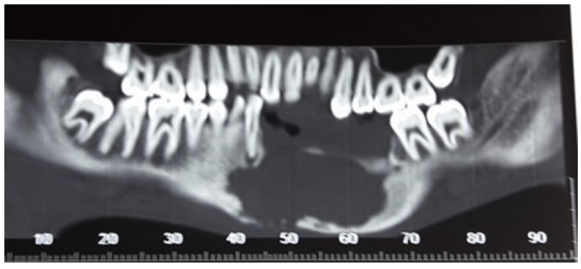

Fig. 3. Orthopantomograph showing the radioluscent lesion from left molar to right canine.

surrounded by more cellular neoplastic spindle cell stroma. Strands of benign amoeloblastic epithelium with elongated nuclei within columnar basal cells showing vacuolization, and reverse polarization surrounded by cellular neoplastic spindle cell stroma. No dysplasia is identified in the epithelium. (Figs. 7–9). Malignant mesenchymal component displayed predominantly hypercellular areas composed of nondescript sheets of stellate to oval spindle cells admixed with few low

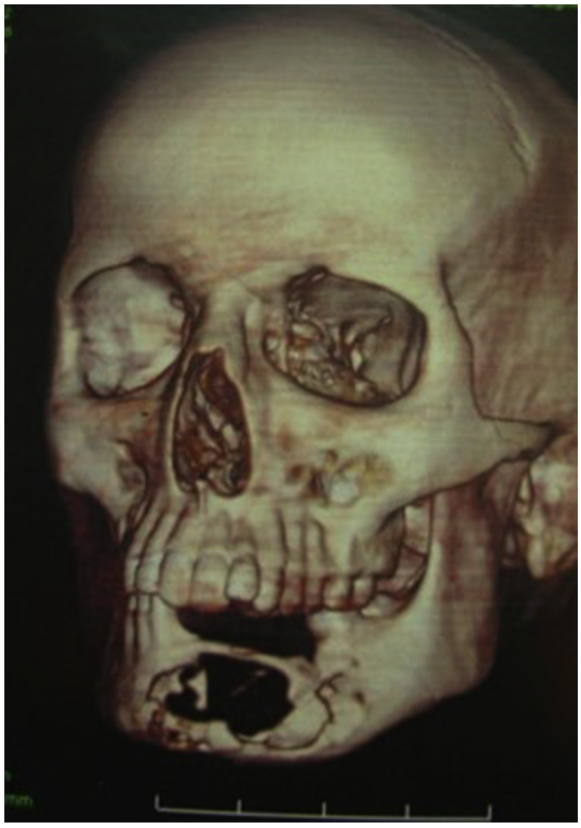

Fig. 4. CT scan with three dimensional reconstruction showing the tumour with multiple areas of perforation.

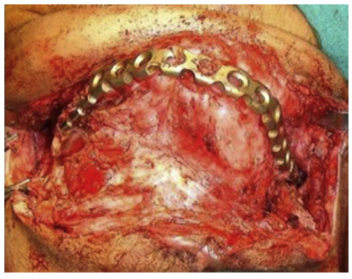

Fig. 5. Reconstruction plate fixed over the mandible.

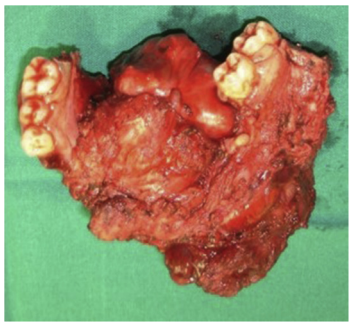

Fig. 6. Excised specimen.

cellular areas with edema. Middle level magnification images revealed no dysplasia in the epithelium (Fig. 10). Sarcomatous foci displaying cellular pleomorphism and mitotic activity confirmed the diagnosis of Ameloblastic fibrosarcoma (Fig. 11). The margins were reported free of tumour.

Patient was kept on regular follow up for two years and there were

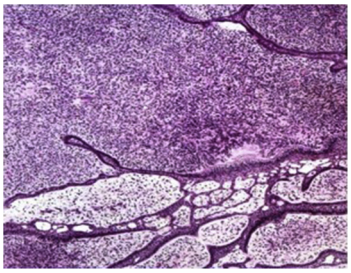

Fig. 7. low level magnification images (Hematoxylin and Eosin stain): Strands of benign amoeloblastic epithelium with elongated nuclei within columnar basal cells showing vacuolization, and reverse polarization surrounded by cellular neoplastic spindle cell stroma.

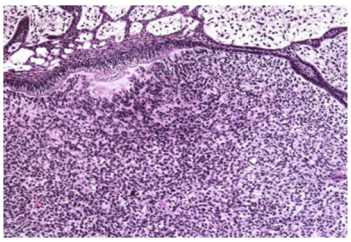

Fig. 8. low level magnification images (Hematoxylin and Eosin stain): Strands of benign amoeloblastic epithelium with elongated nuclei within columnar basal cells showing vacuolization, and reverse polarization surrounded by cellular neoplastic spindle cell stroma.

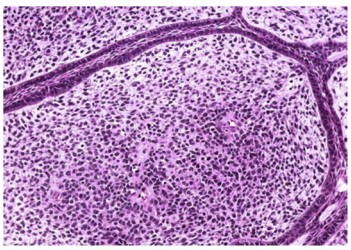

Fig. 9. Strands of benign ameloblastic epithelium surrounded by cellular neoplastic spindle cell stroma.

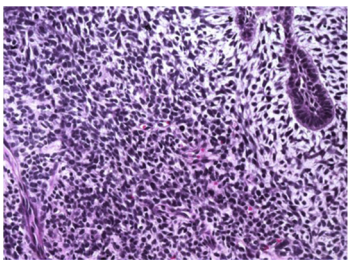

Fig. 10. middle level magnification images: Sarcomatous foci displaying marked cellularity, and cellular pleomorphism.

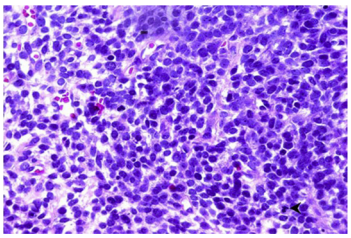

Fig. 11. Sarcomatous foci displaying cellular pleomorphism and mitotic figure in the lower right corner (black arrow).

no signs of recurrence. As patient is a football player, he refused free fibula. Iliac crest bone graft was harvested and fixed with reconstruction plate.

The patient has been on regular follow up and has no signs of the recurrence since over 5 years. Recently mandibular implants were placed in the patient as the graft had taken up well and adequate length with good quality of bone was available. Implants were loaded with bridge completing the rehabilitation.

3. Discussion

Ameloblastic Fibrosarcoma was first described by Heath in 1887 describing it as a spindle cell sarcoma that also had epithelial cells resembling the cells of the enamel organ [5]. Ameloblastic fibrosarcoma (AFS) is a malignant counterpart of ameloblastic fibroma in which the ectomesenchymal tissue shows sarcomatous features admixed with benign ameloblastic epithelium. This was included under the category of odontogenic sarcomas in the WHO classification of Odontogenic tumours in 2017 [6].

AFS can arise de novo or from a previous ameloblastic fibroma (AF). Kobayashi et al. suggest that up to two thirds of AFSs arise from transformation of an AF [7]. Howell and Burkes supported this thesis that most of the lesions arise from preexisting benign neoplasm as Ameloblastic fibroma is usually seen in younger group of patients while AFS is seen in older patients [8].

Leider et al [9] in 1972 presented a case and showed histologic changes in recurrent tumours where benign AF progressed to AFS and then fibrosarcoma with complete loss of odontogenic epithelium. They considered gradual disappearance of epithelial component as degenerative phenomenon. Park et al [10] suggested that anaplasia of mesenchymal tissue is correlated with degeneration of benign odontogenic epithelium. The reduction in benign odontogenic epithelium may result in overgrowth of malignant mesenchymal tissue.

Prein et al [11] 1979 concluded that AFS is a semimalignant tumour because of no record of metastasis and suggested the term proliferative ameloblastic fibroma. Chomete et al [12] reported an instance of pleuropulmonary mediastinal lymph node and hepatic metastasis.

Clinically, AFS arises as a painless or sometimes painful lesion which progresses rapidly compared to its benign counterpart. Duration of AFS may vary from few months to one year producing facial deformity. Involvement of inferior alveolar nerve leads to paresthesia or dysesthesia. Radiologically, AFS presents as a radiolucent mass with ill-defined borders, destruction of the buccal and lingual cortical plates with invasion of surrounding muscles and mucosa.

Grossly the tumor may be cystic or solid with a fleshy whitish to yellow appearance [2]

The histological architecture of AFS is characterized by benign epithelial islands that are composed of columnar or cuboidal peripheral cells arranged in a palisading pattern. At the center of these islands is polyhedral cell reminiscent of stellate reticulum. AF is the main differential diagnosis of AFS. Both neoplasms have a biphasic nature; however, AF has no malignant component, unlike AFS in which the mesenchymal component presents marked cellularity, nuclear pleomorphism, hyperchromatism and a moderate to high number of mitotic figures. Immunohistochemical markers can be helpful to distinguish AFS and AF, and the mesenchymal component of AFS is positive for p53 and PCNA unlike the negativity for these stains in AF [5].

Ameloblastic fibrosarcoma shows different behavior from fibrosarcoma. Metastases usually do not occur in the cases of ameloblastic fibrosarcoma in spite of anaplastic histologic features of mesenchymal component. It is thought that the epithelial component of the tumor exerts an organizational effect over the mesenchymal component, in both benign mixed odontogenic tumors and malignant ones [13].

AFS is considered locally aggressive neoplasm with a low potential for distant metastasis (4.5%) and an overall mortality rate of 25.4% [3].

AFS has a reported recurrence rate of 37% and a mortality rate of 12% [14].

Due to lack of clinical reports, there is no consensus on the treatment yet. In general, the treatment of choice is surgical excision with clear margins and long-term follow-up. Conservative approach shows high incidence of recurrence. Thus wide resection of the lesion with involved soft tissue is recommended.

Routine neck dissection is not recommended as spread is through hematogenous route [1]

Chemotherapy and Radiotherapy have been used however without any conclusive results [1,15]. There is no clear cut consistent pattern of adjuvant chemo or radiotherapy. It has been used in extensive recurrent lesions, and that gives more of regression than cure [1].

The case presented was operated elsewhere at age 14 years for ameloblastic fibroma where he was treated with conservative surgery. He reported after 2 years with ameloblastic fibrosarcoma. Wide resection of the tumour with clear margins was done. Patient was kept under regular follow up. After being disease free for over 2 years, reconstruction was done with iliac crest graft. A year later, when the graft was well settled, dental implants were placed and loaded. Patient has been on follow up for over 5 years with good rehabilitation and no recurrence. No radiation or chemotherapy was given to the patient. As recommended in many patients, AFS is best treated with wide resection with clear margins, regular follow up for at least 2 years before reconstruction and rehabilitation is carried out.

Ethical approval

Not required.

Declaration of Competing Interest

None.

References

[1] Pourdanesh Fereydoun, Mohamadi Mansoureh, Moshref Mohammad, Soltaninia Omid. Ameloblastic Fibrosarcoma of Mandible with distant metastasis. J Oral Maxillofac Surg 2015;73:2067. e1-2067.e7.
[2] Gilani SM, Raza A, Al-Khafaji BM. Ameloblastic Fibrosarcoma: a rare malignant odontogenic tumour. Eur Ann Otorhinolaryngol Head Neck Dis 2014;131:53–6.
[3] Lai Jonathan, Blanas Nick, Higgins Kevin, Kleib Hagene. Ameloblastic fibrosarcoma: report of a case, study of Immunophenotype, and comprehensive review of the literature. J Oral Maxillofac Surg 2012;70:2007–12.
[4] Wood RM, Markle TL, Barker BF, Hiatt WR. Ameloblastic fibrosarcoma. Oral Surg Oral Med Oral Pathol 1988;66:74–7.
[5] Balla Hyandavi, Uppala Divya, Majumdar Sumit, Utharavalli Siva Kalyan. Ameloblastic fibrosarcoma of mandible–a case report. Oral Surg Oral Med Oral Pathol 2017;29:383–6.
[6] El-Naggar AK, Chan JKC, Grandis JR, Takata T, Slootweg PJ. WHO classification of head and neck tumours. 4th ed. Lyon, France: IARC; 2017.
[7] Kobayashi K, Murakami R, Fujii T, Hirano A. Malignant transformation of ameloblastic fibroma to ameloblastic fibrosarcoma: case report and review of the literature. J Craniomaxillofac Surg 2005;33:352–5.
[8] Howell RM, Burkes EJ. Malignant transformation of ameloblastic fibro-odontoma to ameloblastic fibrosarcoma. Oral Surg Oral Med Oral Pathol 1977;43:391–401.
[9] Leider AS, Nelson JF, Trodahl JN. Ameloblastic fibrosarcoma of the jaws. Oral Surg Oral Med Oral Pathol 1972;33:559–69.
[10] Park Hae Ryoun, Shin Kum Bong, Sol Mee Young, Suh Kang Suek, Lee Sun Kyoung. A highly malignant ameloblastic fibrosarcoma report of a case. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1995;79:478–81.
[11] Prein J, Remagen W, Spiessl B, Schafroth U. Ameloblastic fibroma and its sarcomatous transformation. Pathol Res Pract 1979;166:123–30.
[12] Chomette G, Auriol M, Guilbert F, Delcourt A. Ameloblastic fibrosarcoma of the jaws: report of three cases. Pathol Res Prac 1983;178:40–7.
[13] Takeda Y. Ameloblastic fibroma and related lesions: current pathologic concept. Oral Oncol 2009;35:535–40.
[14] Bregni RC, Taylor AM, Garcia AM. Ameloblastic fibrosarcoma of the mandible: report of two cases and review of the literature. J Oral Pathol Med 2001;30:316–20.
[15] Nagori SA, Jose Anson, Bhutia Ongkila, Roychoudhury Ajoy, Kakkarb Aanchal. Ameloblastic fibrosarcoma developing 8 years after resection of ameloblastic fibrodentinoma: a unique presentation. Oral Surg Oral Med Oral Pathol 2015;27:143–6.