Ameloblastic fibro-odontosarcoma: a case report


Abstract. This paper reports one case, of an ameloblastic fibro-odontosarcoma (AFOS) affecting the mandible, in a 12-year-old girl. This neoplasm is a rare odontogenic neoplasm. To the authors’ knowledge this is the fifteenth case of AFOS reported in English. The patient’s chief complaint was a swelling in the face for 6 months. An incisional biopsy was performed diagnosing the case as an ameloblastic fibroma. After radiography ameloblastic fibro-odontoma was diagnosed. Computed tomography was performed and a stereolithography model made to plan the surgical procedures. A hemimandibulectomy followed by a vascularized fibular flap was then proposed. The surgery was uneventful. Microscopic features diagnosed an AFOS. After 23 months of close follow-up there is no sign of recurrence or metastasis. Dental implants were recently placed in the fibular flap.

Keywords: ameloblastic fibro-odontosarcoma; hemimandibulectomy; reconstruction; fibula; stereolithography.

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The sum of radiographic features and histopathology suggested an ameloblastic fibro-odontoma (AFO). Computed tomography revealed a radiolucent lesion in the left mandible with evidence of a retained molar associated with dense focal radiopacity. A stereolithography model was made to plan the surgical procedure and to guide the bending of the titanium plates. A hemimandibulectomy followed by reconstruction was planned because of the size of the tumor, the mandibular destruction and to minimize the patient’s morbidity after the surgery.

Under general anesthesia, the patient was operated on by 3 teams (Oral and Maxillofacial Surgery, Head and Neck Surgery, Plastic Surgery). Intermaxillary fixation was performed using Erich’s splints and wires to hold the occlusion in a centric position. Through a neck incision, a sub-platysmal dissection created an ‘apron flap’. The lesion was then exposed and excised, with inclusion of the oral mucosa, by a left hemimandibulectomy. The condyle was preserved. One titanium reconstructive plate, bent previously, was adapted to bridge the defect, achieving optimal contour (2.4 System, MDT, Brazil). A vascularized fibular flap was transplanted to the site and fixed with two titanium miniplates (2.0 System, MDT, Brazil) and wires. The fibular vessels were sutured to the facial artery and to the external jugular vein. All surgical wounds were closed in layers. The donor site was sutured and a cast was applied. The patient’s recovery was uneventful and she was discharged after 5 days. Ten days after surgery, the patient underwent 99mTc scintigraphy. It showed normal mandibular blood flow and uptake fully compatible with the surgery.

Histological examination showed a biphasic neoplasia with areas of epithelial and mesenchymal elements (Fig. 3). The epithelial component combined strands and buds with peripheral layer of cells exhibiting reverse nuclear polarity. In some fields, the connective element consisted of stellate cells resembling normal fibroblasts. Other fields presented a solid stroma with cellular atypia, nuclear pleomorphism and numerous mitotic figures (Fig. 4). Structures resembling dentinal tubules and immature enamel formation were noted. These patterns were recognized as an AFOS arising in a pre-existing AFO. All surgical resection margins were free of tumor.

Despite the malignant presentation, chemotherapy has not been used because its use is controversial. After 12 months’ follow-up the graft was in place and the occlusion was maintained (Fig. 5). After 23 months of close follow-up there is no sign of recurrence or metastasis. Recently, dental implants were placed in the fibular flap. The donor site recovered uneventfully and the patient walks without problems.

**Discussion**

AFS are rare, true, mixed neoplasms exhibiting a benign epithelial component and malign ectomesenchymal cells. Some authors consider that AFS are the malignant counterparts of the AFO. AFO are considered possible precursor lesions for AFDS and AFOS. CARLOS et al. state that, 64 cases of AFS and 14 cases of AFDS/AFOS have been reported in the English-language literature, up to 2005. To the authors’ knowledge, the case presented here is the fifteenth case of AFDS/AFOS published in English. It is the fifth case in a female patient. Table 1 presents the characteristics of AFS and related lesions.

The diagnosis of AFS is made histologically; some features of the mesenchymal pattern, such as cellularity, cellular atypia, mitosis and palisading patterns must be observed as well as the possible step-wise progression of benign to malignant presentation. The first biopsy taken from this case showed only the benign profile of the tumor. Histological evaluation of the whole specimen revealed dentin, enamel and atypical stroma with mitotically active cells. These findings led the authors to recognize an AFOS arising in a pre-existing AFO.

The case presented here was not microscopically striking so no immunohistochemical markers were used. Nevertheless, other cases might require...
adjuvant diagnostic tools. Although scarce, immunohistochemical staining reports could provide information about tumor growth and behavior. The study of Ki-67, p53, and PCNA showed immunolocalization in the connective tissue of the AFS, revealing a high growth profile in the mesenchymal component, not evident in AF. Poorly differentiated tumors tend to show greater stromal cellularity with a decrease in the epithelial component. Lee et al. described cytokeratin immunostaining as a helpful key in identifying epithelial nests, excluding pure sarcomas. They also reported that mesenchymal cells of AFS showed diffuse and moderate expression of CD34.

Although malignant, AFS rarely metastasizes. Fatal cases are associated with uncontrolled local tumor infiltration after numerous recurrences. Some of the recurrences are linked to curettage or enucleation of a previously diagnosed AF. According to Kobayashi et al., 20% of the patients treated for AFS died within 3 months to 19 years, due to locally aggressive tumor growth. In the 12 months after the present patient’s surgery, there was no sign of recurrence or metastasis. A surgical protocol with wide resection was used in accord with published reports and has the best prognosis. Microscopically there were no signs of tumor infiltration in the surgical resection margins. The use of titanium plates, bent before the surgery, helped to preserve the patient’s facial contour, masticatory function and teeth occlusion. Shaping the plates before surgery reduced the operating time. Healing was uneventful. Scintigraphy, performed 10 days after surgery, and post-surgical radiographs allowed close follow-up for 23 months. The patient is now undergoing dental implant treatment.

In conclusion, the authors present a case similar to others, with some peculiar aspects. The malignization of a benign lesion suggests an unexpected profile and a brief evolution of the case. At diagnosis, the mean age of AF is 14.8 years. For AFS, arising in a pre-existing AF, the mean age is 33 years. In the present case, an AFOS arising in an AFO, occurred in a 12-year-old girl. The authors agree with Bregni et al., Carlos et al., Hayashi et al. and Lee et al. that AFS have the same biological behavior as AFDS and AFOS. The difference between them is the formation of tooth-like structures. An AFO can change its profile to a malignant tumor, such as an AFS or AFDS/AFOS, therefore it is advisable to investigate all cases of AF/AFO because they can disguise a malignant tumor.
### Table 1. Characteristics of AF, AFS and related lesions.

<table>
<thead>
<tr>
<th></th>
<th>AF</th>
<th>AFD</th>
<th>AFO</th>
<th>AFS</th>
<th>AFDS/AFOS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender predilection</td>
<td>Males</td>
<td>Males</td>
<td>no predilection</td>
<td>Males</td>
<td>Males</td>
</tr>
<tr>
<td>Mean age</td>
<td>14.8 years</td>
<td>14.8 years</td>
<td>8-12 years</td>
<td>27.5 years</td>
<td>30 years</td>
</tr>
<tr>
<td>Rarity</td>
<td>Rare</td>
<td>Rare</td>
<td>Less common than AF/AFD</td>
<td>Very rare</td>
<td>Very rare</td>
</tr>
<tr>
<td>Age range</td>
<td>7 weeks – 62 years</td>
<td>7 weeks – 62 years</td>
<td>X</td>
<td>3 – 83 years</td>
<td>12-83 years</td>
</tr>
<tr>
<td>Rarity</td>
<td></td>
<td></td>
<td>Very rare</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anatomic predilection</td>
<td>Mainly in the posterior mandible</td>
<td>Posterior mandible</td>
<td>None</td>
<td>Posterior mandible</td>
<td>X</td>
</tr>
<tr>
<td>Clinical features Imaging</td>
<td>Asymptomatic</td>
<td>Asymptomatic</td>
<td>Asymptomatic</td>
<td>Swelling and pain Ill-defined radiolucency</td>
<td>Asymptomatic, swelling Ill-defined radiolucency, dental hard tissues; radiopacities</td>
</tr>
<tr>
<td>Odontogenic ectomesenchyme</td>
<td>Benign</td>
<td>Benign</td>
<td>Benign</td>
<td>Malign</td>
<td>Malign</td>
</tr>
<tr>
<td>Epithelial strands and buds</td>
<td>Benign</td>
<td>Benign</td>
<td>Benign</td>
<td>Benign</td>
<td>Benign</td>
</tr>
<tr>
<td>Dental structures</td>
<td>No</td>
<td>No</td>
<td>Dysplastic dentin</td>
<td>Dental hard structures</td>
<td>No</td>
</tr>
<tr>
<td>Treatment</td>
<td>Enucleation and curettage</td>
<td>Enucleation and curettage</td>
<td>X</td>
<td>Wide surgical resection</td>
<td>Wide surgical resection</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Rare malignant progression</td>
<td>Rare malignant progression</td>
<td>Excellent</td>
<td>Recurrence without distant metastasis</td>
<td>Recurrence without distant metastasis</td>
</tr>
</tbody>
</table>

AF, ameloblastic fibroma; AFD, ameloblastic fibrodentinoma; AFO, ameloblastic fibro-odontoma; AFS, ameloblastic fibrosarcoma; AFDS, ameloblastic fibroodontosarcoma; AFOS, ameloblastic fibro-odontosarcoma; X, no data from consulted authors.


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