Ameloblastic Fibrosarcoma of the Mandible

A 32-year-old Filipino woman presented with a 3-year history of slowly enlarging left hemimandibular mass with no associated symptoms. Previous biopsy showed ameloblastoma. Imaging revealed a translucent multiloculated mass with ill-defined borders. (Figure 1) On examination, the mass was irregularly shaped, measures 40 x 39 cm, slightly hyperpigmented and erythematous, warm with visible vessels. The patient underwent left segmental mandibulectomy with reconstruction and the specimen was sent for histopathologic evaluation.
Received was a mandibulectomy specimen weighing 1850 grams and measuring 17 x 14.5 x 14 cm. The body, angle, ramus and condyles of the left hemimandible are no longer identifiable grossly. There are ten teeth attached. Cut sections show multiple cystic spaces which measure from 0.8 to 15.0 cm in widest diameter, filled with abundant red-brown necrotic debris and yellow-brown purulent material. The mass has a grey-tan soft to fibrous cut surface with focal gritty areas. (Figure 2)

Microscopic examination shows a biphasic neoplasm composed of benign epithelial and malignant mesenchymal components. (Figure 3) The benign epithelial component is arranged in islands and strands with peripheral palisading, composed of bland cells without atypia. (Figure 4) The abundant mesenchymal component is composed of spindle cells in fascicles. The cells are moderately pleomorphic with enlarged hyperchromatic nuclei, prominent nucleoli, coarse chromatin and scant cytoplasm. The cells are suspended within loose stroma with variable degree of cellularity. Some mitoses are noted. (Figure 5)

Ameloblastic fibrosarcoma (AFS) belongs to a group of odontogenic

Figure 3. Ameloblastic Fibrosarcoma (AFS) biphasic morphology composed of benign epithelial and malignant mesenchymal components. (Hematoxylin-Eosin, 100X magnification).

Figure 4. The benign epithelial component is arranged in islands and strands with peripheral palisading, composed of bland cells without atypia. (Hematoxylin-Eosin, 400X magnification).
sarcomas in which the epithelial component is cytologically benign while the ectomesenchymal component shows malignant features. The AFS is the most common type among the odontogenic sarcomas. It is regarded as the malignant counterpart of ameloblastic fibroma (AF). Although most cases arise de novo, the documentation of a prior or pre-existing precursor lesion from ameloblastic fibroma suggests otherwise.\(^1,2\) A study by Kobayashi et al. suggested that up to one-third of AFS arise from AF while a review of literature by Lai et al. demonstrated that 51% of AFS had previously documented AF at the same site, providing supporting evidence of a malignant transformation.\(^3\)

Ameloblastic fibrosarcoma has a reported age range of 3 - 89 years, with overall mean patient age of 27.3 years. In cases of prior diagnosis of AF, AFS can occur at a mean patient age of 33 years whereas a mean patient age of 23 years where no prior diagnosis of AF was documented.\(^1,4\) It mainly affects males in the third or fourth decade of life and the posterior mandible is the most commonly affected site with ratio of mandibular to maxillary incidence of 4:1.\(^1,3,4\) The clinical presentation is usually that of a painful, enlarging mass and most lesions are radiographically translucent and multiloculated with ill-defined borders.\(^2,4\)

The histologic features of AFS reveal a mixture of benign odontogenic epithelium ranging from budding and branching cords to large islands composed of columnar or cuboidal peripheral cells arranged in palisading pattern, and an abundant malignant mesenchymal component showing marked cellularity, nuclear pleomorphism and moderate mitotic figures.\(^1,2,3\) Ameloblastic fibroma differs from AFS by having no malignant component and immunohistochemical stains have been suggested to provide distinction, particularly when identifying a low-grade fibrosarcoma. The malignant component of AFS will show positivity in p53 and PCNA and will have a higher Ki-67 expression than AF.\(^3,4\)

Although AFS are low to intermediate-grade sarcomas, a high incidence of recurrence is reported - about one third of patients experience recurrence and overall mortality rate is 25%. However, only less than 5% of cases with metastases have been reported.\(^1,3\) Long term follow up is thus indicated.