Juvenile Psammomatoid Ossifying Fibroma

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History

A four year male Asian-Pacific Islander presented with a six month history of a slowly expansile lesion of the right posterior mandible. Slight facial asymmetry was clinically evident.

Radiographic Features

Imaging studies revealed a well demarcated unilocular lesion of the right posterior mandible. There was circumferential expansion of the mandible with sparing of the condyle. The cortex was expanded but intact. The body of the lesion exhibited a relatively homogenous ground glass, but subtly variegated, radiodense appearance. The characteristic narrow radiolucent rim between the residual osseous cortex and the body of the lesion is readily visualized (Fig. 1).

Treatment

Incisional biopsy was followed by surgical resection of the mass via hemi-mandibulectomy.

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Diagnosis

Histologic examination revealed a cellular proliferation of uniform, cytomorphologically bland fibroblastic spindle cells. Despite the tumor's cellularity, mitotic figures were not evident. Minimal extracellular collagen deposition was present. Interspersed between the fibroblastic cells were numerous irregular and spherical ossicles exhibiting varying degrees of calcification. These psammoma body-like ossicles were relatively acellular, with a concentric pattern of lamination (Fig. 2). The fibroblastic tumor cells extended to a margin of reactive new bone representing expansion and replacement of the cortex. The constellation of histomorphologic, radiographic and clinical features of this lesion supported an interpretation of juvenile psammomatoid ossifying fibroma (JPOF).

Discussion

Within the spectrum of benign fibro-osseous lesions of the jaws, ossifying fibromas represent the neoplastic entities. The ossifying fibromas (OF) are subdivided into conventional and juvenile clinicopathologic subtypes. On the basis of morphologic features, juvenile ossifying fibromas are further separated into trabecular (JTOF) and psammomatoid (JPOF) variants [1]. The juvenile variants are characterized by distinctive trabecular or psammomatoid matrix production, occurrence in younger patients relative to conventional OF and a predilection for the bones of the paranasal sinuses, the periorbital region and the maxilla [2–4]. In contrast, conventional OFs tend to be more common in the mandible.

The ground glass nature of this JPOF is typical, however in addition to the radiographic findings described above;

Fig. 1 Axial CT scan (bone window) demonstrates an expansile mass lesion involving the right posterior mandible and ramus. A developing tooth is present at the anterior medial margin of the mass

Fig. 2 Characteristic JPOF histologic pattern of psammomatoid ossicles set in cellular fibroblastic tissue

JPOF imaging findings can include multilocularity, cystic degeneration with fluid levels. Variations in radiodensity from purely radiolucent to mixed density with prominent radiopacities have been described. Additionally, serial radiographs of JPOF may reveal progressive calcification of the matrix [2]. Tumors affecting the tooth bearing portions of the jaws can cause root resorption or tooth displacement. Entities considered in the radiographic differential diagnosis of this lesion include fibrous dysplasia

and mixed odontogenic tumors. Fibrous dysplasia also presents as a ground glass radiopacity, however it can be excluded on the basis of the well demarcated border with a radiolucent rim instead of a blended, indistinct margin. The overall configuration of the JPOF is more rounded than the typically fusiform outline of fibrous dysplasia. The mixed odontogenic tumors, odontoma and ameloblastic fibro-odontoma also occur in younger patients and present as mixed density mass lesions in the tooth bearing portions of the jaws. These tumors are well demarcated, often intimately associated with an unerupted tooth and contain coarse opacities that are similar in radiodensity to tooth structure [5].

Treatment of JPOF consists of complete surgical removal and incomplete excision has been associated with a high local recurrence rate [2, 4].

The precise nature and classification of the juvenile ossifying fibromas has been the source of considerable debate among pathologists, resulting in a confusing proliferation of competing nomenclatures [6]. This controversy is exacerbated by the fact that this group of tumors is not restricted to pediatric patients with morphologically identical lesions identified in adults. Fortunately, a degree of consensus has been established and most sources accept the existence of juvenile ossifying fibromas which are further subdivided into JPOF and JTOF subtypes.

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