Fibrous Dysplasia

Fibrous dysplasia, a benign, non-neoplastic disease, is seen in all parts of the body. It appears in the otolaryngic area chiefly in the maxilla, but also in the mandible, ethmoids, frontal and temporal bones. Polyostotic forms occur but in the facial area the monostotic type is the rule and represents 90 percent of all cases of fibrous dysplasia. Most patients are under 30 with the disorder beginning in the first or second decade of life.

The gross specimen demonstrates abnormal bone gradually blending into the normal and it is easy to cut.

Microscopically, trabeculae of immature nonlamellar (woven) bone are scattered throughout nondescript fibrous tissue having variable cellularity. The margins of the bony fragments are indistinct and have no clear limiting edge of osteoblasts - a distinguishing feature from ossifying fibroma. The irregular trabeculae have C - and S-shapes like Chinese script writing. Multinucleated giant cells and cementum-like calcifications may be present.

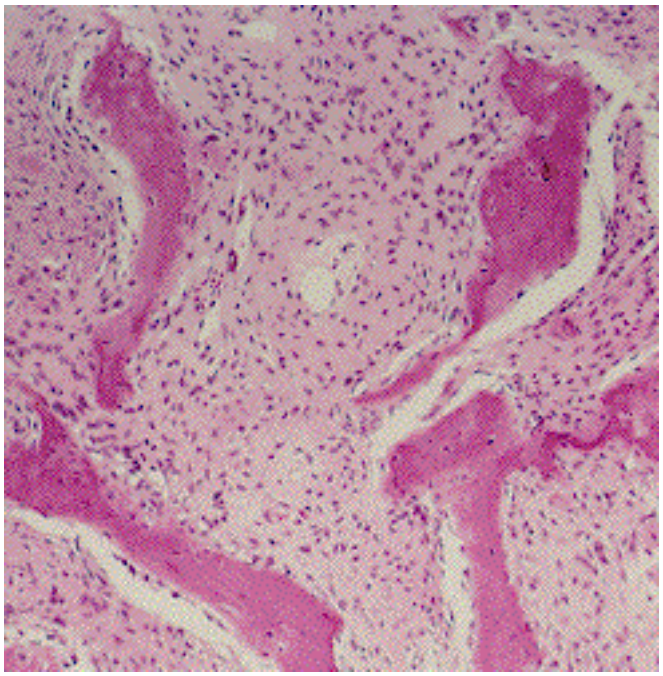
Ossifying Fibroma

This is a slow growing tumor often discovered incidentally on a radiograph and, as noted above, it differs from fibrous dysplasia microscopically in that ossifying fibroma shows the connected trabeculae of woven bone to be outlined by osteoblasts. In this example a cementum-like material is also present. It is not always possible to differentiate histologically between ossifying fibroma, fibrous dysplasia and cementoma but clinical correlations are helpful with special reference to radiographic findings.

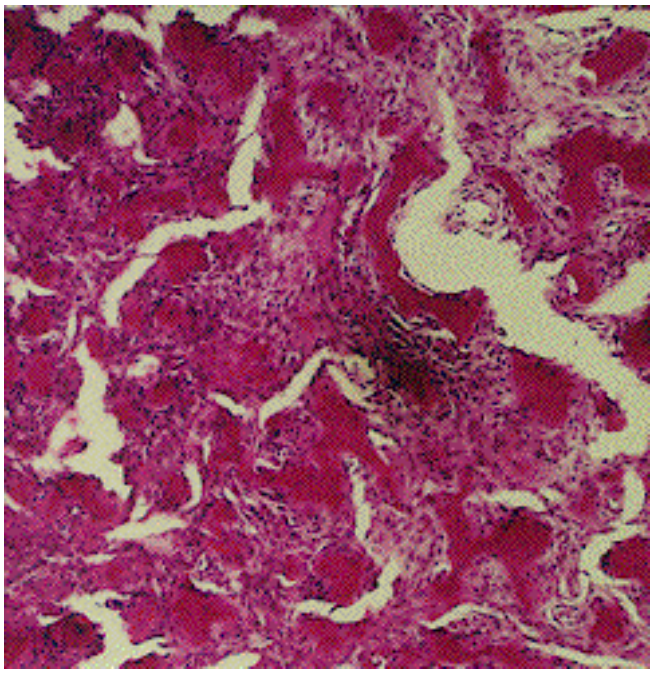
Cementoma (Cementifying fibroma)

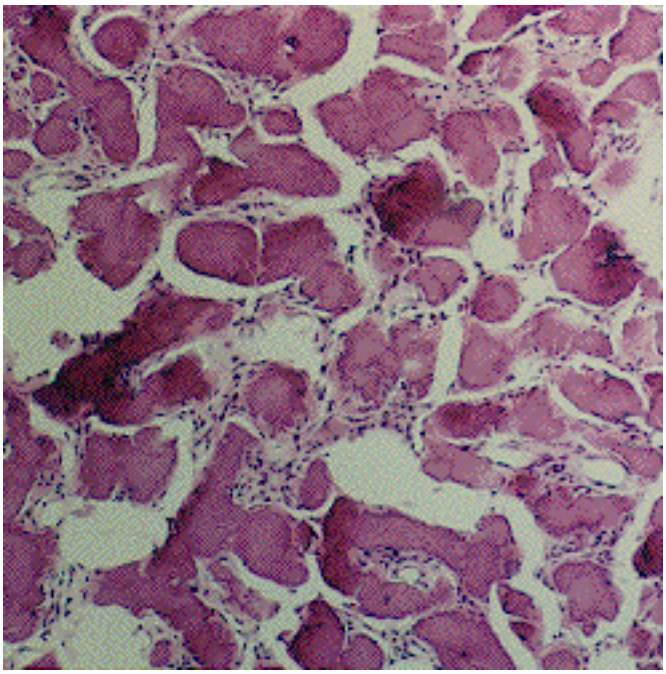
Cementoma is one of a group of conditions sometimes listed under fibrosseous lesions that include such entities as fibrous dysplasia as well as other conditions arising from the multipotential blastic cells of the periodontal membrane which are able to produce bone, cementum, and fibrous tissue. Cementum is a mineralized dental tissue covering the root of a live tooth and has the characteristics of compact bone, but is avascular. The tumor arises in the mandible in most cases, but is occasionally seen in the maxillary or ethmoid area and may have intranasal extension.

Fibrous dysplasia, maxilla. Woven bone with nonlamellar trabeculae resembling Chinese script characters C- and S- blend into the surrounding fibrous tissue. Note lack of osteoblasts at the periphery of the trabeculae.



Ossifying fibroma-Trabeculae of cancellous bone are rimmed by a prominent layer of osteoblasts.





Cemento-ossifying fibroma. The bony trabeculae have rounded configurations resembling cementum. This is a histologic variant of ossifying fibroma.

Clinical Aspects:

Because the affected bone in fibrous dysplasia becomes softer than the surrounding bone, radiologically it takes on a ground glass appearance with indefinite borders — a mark distinguishing the condition from ossifying fibroma. A lateral film of the skull may show involvement of the skull base in addition to maxillary involvement. Minimal cases need not be treated at all; for the advanced case, conservative surgery is preferred, although because of extent of the disease, this may amount to a major resection. The disease sometimes stabilizes at puberty. Malignant change is rare.