Fibrous dysplasia of bone

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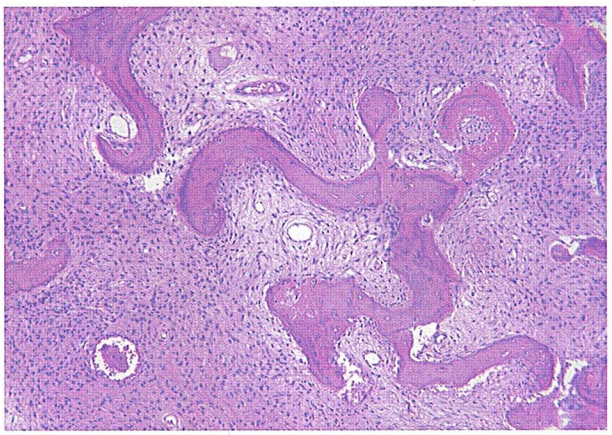

Figure 1. The spicules of woven bone are curvilinear and have a "Chinese character" shape, and they are set in a stroma of variably cellular fibrous connective tissue.

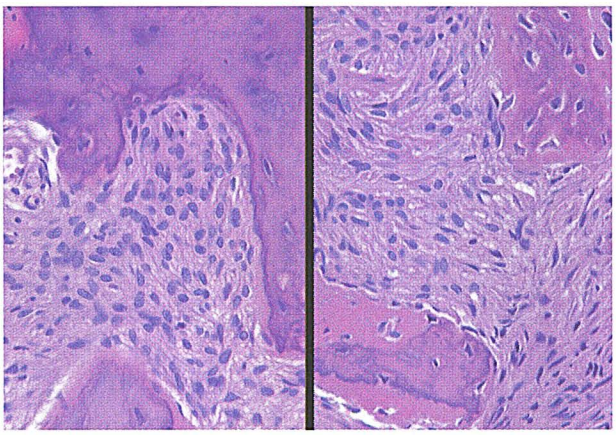

Figure 2. Two high-power images demonstrate the interface of the bone trabeculae with the fibroblastic stroma. The stromal cells have round-to-oval nuclei and an even chromatin distribution. The specimen on the right contains a few strands of collagen. No osteoblasts are seen.

Fibrous dysplasia (fibro-osseous metaplasia) is one of a diverse group of diseases that are characterized by alterations in bone growth. It is a developmental, tumor-like process of unknown etiology. Its initial clinical sign is usually a painless enlargement of the affected bone. It occurs in equal proportions in males and females, most often during the first two decades of life.

Fibrous dysplasia is subclassified into two main clinical subtypes: *monostotic* and *polyostotic*. The monostotic type, which accounts for 80% of cases, affects only one bone, usually the maxilla; the polyostotic type affects multiple bones. Polyostatic fibrous dysplasia is seen in both Jaffe-Lichtenstein and McCune-Albright syndromes, along with skin hyperpigmentation (café au lait tan macules) and sexual precocity (the most common endocrine disturbance). Radiologic studies typically demonstrate a poorly defined, ground-glass-type lesion; in the polyostotic form, multilocular radiolucencies may be seen.

Histologically, fibrous dysplasia features irregularly shaped trabeculae of immature, woven bone in a background of variably cellular, loosely arranged fibrous stroma (figure 1). The spicules of bone are often curvilinear or branching, and they have a "Chinese character" or "alphabet soup" appearance. The delicate trabeculae do not have osteoid rims, but they do have minimal osteoblastic rimming (figure 2). The fibroblasts usually have plump, ovoid nuclei, although elongated, narrow nuclei are sometimes also seen.

Treatment of fibrous dysplasia is challenging. Bone growth can stop spontaneously, but by then it might have already resulted in a cosmetic deformity that requires surgical excision. Regrowth occurs in as many as 50% of patients over time. Irradiation is not indicated.

Suggested reading

El-Mofty SK, Kyriakos M. Soft tissue and bone lesions. In: Gnepp DR, ed. Diagnostic Surgical Pathology of the Head and Neck. Philadelphia: W.B. Saunders, 2001:505-604.

Waldron CA. Bone pathology. In: Neville BW, Damm DD, Allen CM, Bouquot JE, eds. Oral and Maxillofacial Pathology. Philadelphia: W.B. Saunders, 1995:443-92.

Waldron CA. Fibro-osseous lesions of the jaws. J Oral Maxillofac Surg 1993;51:828-35.

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