Fibrous Dysplasia

Fibrous dysplasia of bone is a relatively common hamartomatous developmental process in the skeletal system that can be either monostotic or polyostotic. The polyostotic form can be associated with endrocrine abnormalities such as precocious puberty in females in the McCune-Albright Syndrome. This condition occurs primarily in females and can include café-au-lait spots on the skin that have a rough, serrated edge compared to the smooth edges in neurofibromatosis. The monostotic form has an appearance similar to non-ossifying fibroma except that the lytic component has a smoky appearance caused by metaplastic bone formation in the fibrous tissue. In the more severe polyostotic form, representing only about 10% of fibrous dysplasia, the disease tends to amplify on one side of the body more than the other causing leg length discrepancy. An example of this is a characteristic chronic deformity of the proximal femur , referred to in the literature at the shepherd's crook deformity, that frequently requires surgical correction.

The most common locations for fibrous dysplasia are the proximal femur, followed by the tibia, pelvis, humerus, radius, ribs and, occasionally, the cranio-facial anatomy. Other endocrinopathies associated with polyostotic disease include hyperthyroidism, acromegaly, Cushing's syndrome and, occasionally, hypophosphatemic osteomalacia. When soft tissue myxomas are present with fibrous dysplasia, it is referred to as the Mazabraud's syndrome. Radiographically, there will be diffuse dysplastic changes throughout the involved long bones with gradual dilatation and thinning of the cortices, and a soap-bubbly appearance but with a smokey look compared to non-ossifying fibroma. Histologically, the picture is that of benign fibrous tissue matrix studded with metaplastic islands of bone with a so-called alphabet soup appearance but without osteoblastic rimming of the trabeculae. Occasionally one will see macrophages, osteoclasts, foam cells and even cholesterol deposits similar to what is found in non-ossifying fibroma. There is a one per cent chance that fibrous dysplasia will convert to a secondary sarcoma in adult life in which case it would present as an osteosarcoma, malignant fibrous histiocytoma or even a chonodrosarcoma.

Treatment usually consists of corrective osteotomies for secondary developmental deformities, along with bone grafting, including fibular strut grafts and occasionally intramedullary devices to prevent repeated stress fractures. Recently there has been enthusiasm for the use of bisphosphonates to inhibit the osteoclastic activity in growing children that produces progressive weakening of the bone. It is best not to perform bone-grafting procedures in children because of the high recurrence rate in children under 16 years of age.