

Osteofibrous Dysplasia

Osteofibrous dysplasia is a rare condition occurring typically in children less than 10 years of age. This benign condition has radiographic and microscopic features similar to fibrous dysplasia of the tibia, the main difference being the presence of more heavily ossified tissue at the periphery of the lesion giving it a somewhat soap-bubbly appearance on X-ray. Microscopically, it has a similar "alphabet soup" metaplastic bone appearance in a benign fibrous tissue stroma but with the additional feature of heavy osteoblastic rimming of the trabeculae not seen in fibrous dysplasia. It typically arises from the anterior cortex of the tibia at mid-shaft and causes a progressive anterior bowing of the tibia over time, creating a cosmetic deformity associated with pain of a dull, aching nature. The radiographic appearance is very similar to and impossible to separate from that of the malignant adamantinoma that also affects the tibia in children and, for this reason, a biopsy must be performed to rule out the malignant possibility. Occasionally, osteofibrous dysplasia of the tibia can progress gradually into a well-differentiated form of the adamantinoma, which has microscopic features of both osteofibrous dysplasia and adamantinoma in the same lesion. As far as treatment is concerned, early surgical debridement and bone grafting before age 15 years results in a high local recurrence rate and should be discouraged until the child reaches full bone maturity at which time a definitive debridement and bone grafting procedure can be performed without significant risk of recurrence. This lesion can be seen bilaterally in a very small percentage of cases and can also involve the fibula on the same side. This process in newborns appears to be more osteolytic and destructive in nature.