Osteofibrous dysplasia
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03/11/2013

History
18 month old male with bowed leg.

Diagnosis
Osteofibrous Dysplasia

Discussion
Osteofibrous dysplasia (OFD) is a benign lesion occurring almost exclusively within the cortex of the tibial diaphysis. While typically confined to the tibia, OFD may occur less commonly in the ipsilateral fibula, or in the fibula alone. OFD is rare, accounting for roughly 0.2% of all primary bone tumors, and typically occurring in patients less than 20 years old. Patients with OFD typically present with soft tissue swelling in the leg with or without pain, or anterior bowing of the leg. Less commonly, pathological fracture or a radiological study for an unrelated concern may lead to the finding of OFD. Radiographically, OFD is typically observed in the anterior cortex of the tibia, potentially causing anterior bowing. Well margined lytic lesions occur within the cortex, and are often surrounded by a zone of sclerosis. The involved cortex may appear thickened, and while periosteal reaction is rare, when it does occur it is chronic in appearance. Histologically, OFD is characterized by spicules of woven bony trabeculae that are lined by osteoblasts, set within a fibrous backdrop. The osteoblast lining is key to differentiating OFD from fibrous dysplasia, which typically lacks this osteoblast lining. OFD must be differentiated diagnostically from adamantinoma (AD), a theoretically related and malignant condition. Along with plain radiographs, CT and MRI images of the lesion may provide additional information important for the diagnosis of OFD and differentiation from AD. A biopsy should be performed to confirm the diagnosis. Although OFD is considered benign, management and treatment remains controversial. Observation is typically recommended for treatment, with some recent recommendations for extraperiosteal resection. The prognosis for OFD is good, since progression usually ceases upon maturation of the skeletal system. Several large studies conducted recently all showed no progression of OFD with long term follow-up.

Findings
CR-Lucent medial cortical defect in the proximal tibia with gene varus

Reference

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