Desmoplastic Fibroma of Bone

Desmoplastic fibroma of bone is an extremely aggressive, but benign, fibrous lesion seen in long bones as well as the mandible. It represents one per cent of all bone tumors and occurs typically in children and young adults with equal sex distribution. The long bones involved include the distal femur, proximal tibia, proximal humerus and the lower half of the pelvis. Radiographically, these lesions have an aggressive and motheaten appearance, eroding into cortical bone and breaking out into the adjacent soft tissue, much like the low-grade fibrosarcoma. Histologically, the tumor appears identical to aggressive fibromatosis seen in soft tissue with intense collagen formation by aggressive-appearing fibroblasts and an occasional mitotic figure. The recurrence rate is very high with curettement alone (around 40 per cent) and for this reason most authorities suggest a wide surgical resection, similar to what would be used for a soft tissue aggressive fibromatosis, for which a prosthetic implant might be required. This tumor never metastasizes to the lung.