

## **Adamantinoma of Bone**

The adamantinoma of bone is an extremely rare primary sarcoma of bone, accounting for about 0.3 per cent of all malignant tumors of bone. In 90 per cent of the cases, it will be seen in the diaphyseal portion of the tibia, especially in the anterior cortex. It occurs equally in males and females, typically in the second and third decades of life. The tissue of origin still remains a mystery but immunohistochemical stains suggest an epithelial origin that might account for why the tumor is more common just beneath the skin in the anterior cortex of the tibia. Radiographically, the adamantinoma takes on the appearance of a fibrous dysplastic lesion or perhaps osteofibrous dysplasia of the tibia. The lesion appears benign with a central lytic core surrounded by dense, reactive fibro-osseous bone that frequently dilates the anterior cortex and may be multiloculated in appearance. The tumor is quite slow growing and is usually painful, whereas with fibrous dysplasia and osteofibrous dysplasia, the patients remain asymptomatic. If a so-called fibrous dysplastic lesion continues to grow past maturity, a physician should suspect adamantinoma, especially if the lesion is painful, and look for the characteristic nests and cords of epithelioid-looking cells surrounded by fairly benign-appearing fibro-osseous tissue on histological specimens. It is very rare for this tumor to metastasize to different sites but occasionally it will metastasize to regional lymph nodes and the lung.

Treatment for this low-grade lesion is purely surgical, consisting of a wide local resection, frequently a segmental resection of the mid-portion of the tibia, and reconstruction with a large bone allograft over an intramedullary nail. There have been a few rare cases in the literature where adamantinoma has arisen out of a pre-existing osteofibrous dysplastic lesion.