

Osteofibrous dysplasia

Definition

Rare tumour related to Adamantinoma found principally in the tibia.

Epidemiology

Found in the first decade.
Slightly more common in boys.

Site

This is typically found on the anterior aspect of the tibia, in the diaphysis

Aetiology

May be part of a spectrum between fibrous dysplasia and adamantinoma.

Clinical

Usually presents because of deformity from the anterior tibial bowing.

Radiological

Frequently associated with marked anterior bowing of the tibia.

Characterized by an expanding intracortical osteolysis.

Pathology

There is a similar appearance to fibrous dysplasia, with fibrous tissue amongst trabecular bone, but the lining osteoblasts are much plumper in appearance. There may be production of lamellar bone by the osteoblasts.

Treatment

Bracing is recommended in young patients, because of the high recurrence rate. Consider resection, and bone transport with the Ilizarov technique.

Prognosis

There is usually progression throughout childhood, but this tends to stop at skeletal maturity.

If surgery is undertaken prior to age 10 there is a high recurrence rate, so if possible surgery should be delayed until after this.

Adamantinoma

Definition

Extremely rare tumour related to ossifying fibroma, characteristically located in the tibia.

Epidemiology

Found in the second and third decades

Site

Anterior aspect of the tibia and occasionally the fibula.

Aetiology

May represent a regressing osteofibrous dysplasia, or may be the precursor to this lesion.

Clinical

May be asymptomatic, or mildly painful.

Radiology

Well demarcated lytic or mixed lytic/sclerotic lesion involving the cortex. May be soft tissue extension.

MRI: T1 low, T2 high signal

Pathology

Biphasic pattern, with islands of epithelial like tissue surrounded by fibrous tissue.

Treatment

Radical excision.

Prognosis

Recurrence occurs after limited resections.

Metastases occur in around 30%.