Chondromyxoid fibroma

Definition

A rare benign tumour derived from cartilage forming connective tissue. Jaffe and Lichtenstein originally described the tumour in 1948

Aetiology

May arise from the epiphyseal cartilaginous plate. Has a characteristic rearrangement on at 6q13.

Incidence

1.8% of all benign tumours in the Mayo data Chondroblastoma is 2.5 times more common than CMF

There is a slight male preponderance Most common in the second and third decades

Localisation

Typically in the metaphyseal region of a long bone. The lower limb is affected in 70% Rarely, involves both metaphysis and epiphysis Proximal tibial metaphysis is the commonest site. The small bones of the foot are commonly involved.

Symptoms

Pain Local swelling is found, rarely. Occasionally asymptomatic finding on XR

Physical findings

May be swelling and tenderness

Radiology

Characteristically an eccentric, sharply circumscribed zone of rarefaction that occasionally expands the bone. The defect can be round or oval. It frequently has a scalloped appearance, caused by the lobulated nature of the tumour. In many cases trabeculae appear to traverse the defect. These are actually corrugations on the surface of the cavity that contains the tumour. Chondromyxoid fibroma involving the surface of a bone tends to have extensive mineralization.

Bone scan is hot.

MRI typically shows low intensity on T1 and high intensity on T2.

Pathology

Macroscopic

Usually small; in the Mayo data the largest tumour was 5cm. Fragments appear firm, fibrous and semi translucent. If it is removed intact it appears lobulated and sharply demarcated

Microscopic

Mixture of myxomatous zones, fibrous zones and zones with chondroid appearance. There is a characteristic lobular pattern of growth, with a hypocellular appearance centrally and a hypercellular periphery.

At the edge of the periphery around 50% of tumours have scattered giant cells.

May be foci of cellular atypia.

The most important differential diagnosis is of a myxoid chondrosarcoma. These typically show liquefactive changes in the matrix, clear permeation of the surrounding bone, malignant XR features and most importantly hypercellularity throughout

Treatment

En bloc resection is the best treatment Curettage has around a 25% risk of recurrence, and bone grafting is often necessary. Bone grafting may reduce the risk of recurrence. Radiotherapy is not indicated except in the very rare surgically inaccessible region.

Prognosis

Recurrence rate of around 25% Sarcomatous change has not been convincingly demonstrated. It may occur after radiotherapy.