Desmoplastic fibromas (DF) are extremely rare bone tumours that do not metastasize, but may be locally aggressive. They are considered to be a bony counterpart of soft tissue desmoid tumours and are histologically identical.

Clinical presentation

Incidence is ~0.3%. The most common areas of involvement include the mandible, pelvis and femur.

Mean age at presentation is 21, and there is no sex predilection.

Pathology

Desmoplastic fibromas histologically are identical to soft tissue desmoid tumors, with abundant collagenous stroma and little cellularity or pleomorphism. The main cell types that are seen include: fibroblasts, myofibroblasts, and undifferentiated mesenchymal cells

Radiographic features

Plain film

typically seen as a lytic bone lesions with a geographic pattern of bone destruction often has a narrow zone of transition and non-sclerotic margins internal pseudotrabeculation: > 90% 3 no matrix mineralisation widening of the host bone from gradual apposition of periosteal new bone formation: ~ 90%

MRI There is considerable overlap with other bony lesions on MR appearances. Signal characteristics include:

T1: typically low signal T2: has background intermediate to high signal with intrinsic low to intermediate intensity foci within T1 C+ (Gd): often shows heterogeneous enhancement Treatment and prognosis

Despite being benign it is a still locally aggressive tumour. En bloc resection is the current treatment of choice.