CHAPTER 12

Fibrogenic Tumours

Tumours of fibrogenic origin do not have a mineralizing matrix but generally produce collagen; high grade tumours may not produce any matrix.

Desmoplastic fibroma is one of the most uncommon of bone tumours. It is identical to the much more common soft tissue desmoid and locally aggressive.

Fibrosarcomas range from the well differentiated tumours, which are difficult to separate from desmoplastic fibroma, to highly malignant tumours which are composed of small cells and simulate Ewing sarcoma. Distinction from fibroblastic osteosarcoma may be arbitrary and may depend on sampling.

Desmoplastic fibroma of bone

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Definition
Desmoplastic fibroma is a rare, benign bone tumour composed of spindle cells with minimal cytological atypia and abundant collagen production.

ICD-O code 8823/0

Synonyms
Desmoid tumour of bone, intra-osseous counterpart of soft tissue fibromatosis.

Epidemiology
The incidence is approximately 0.1% of all primary bone tumours. It tends to occur in adolescent and young adults with near equal gender distribution.

Sites of involvement
Desmoplastic fibroma may involve any bone but is most frequent in the mandible.

Clinical features / Imaging
Patients present with a variety of symptoms. Some have pain, others present because of deformity or loss of function. Radiographically, desmoplastic fibroma is usually a well defined, radiolucent lesion that may expand the host bone. Intralesional trabeculation is frequent. Larger lesions may breach the periostium and extend into soft tissue. Such erosive, destructive pattern may mimic other, more aggressive lesions. Desmoplastic fibroma has low signal intensity in both T1 and T2 weighted MRI images. The extent of disease and margins are best assessed with CT and MRI.

Macroscopy
The tumour is firm and the cut surface is creamy-white with a variegated whorled pattern. The advancing surfaces of the lesion tend to be scalloped and apparently well defined. The tumour may extend into soft tissue.

Histopathology
The lesion is composed of spindle cells (fibroblasts/myofibroblasts) on a richly collagenous, variably hyalinized background. The degree of cellularity is variable but cellular atypia and pleomorphism are minimal or absent. Mitoses are rare.

Genetics
FISH analyses of desmoplastic fibroma suggest that trisomies 8 and 20 represent nonrandom aberrations in a subset of these lesions, analogous to similar findings in soft tissue desmoid tumours {267}.

Prognostic factors
The tumour behaves in a locally progressive/aggressive manner. Recurrence following curettage and resection are 72% and 17%, respectively {832}. Local relapse has been reported as late as eight years following primary surgery. There is a single reported case involving the spine that showed little detectable change over a follow up period of nine years without therapy {1482}.

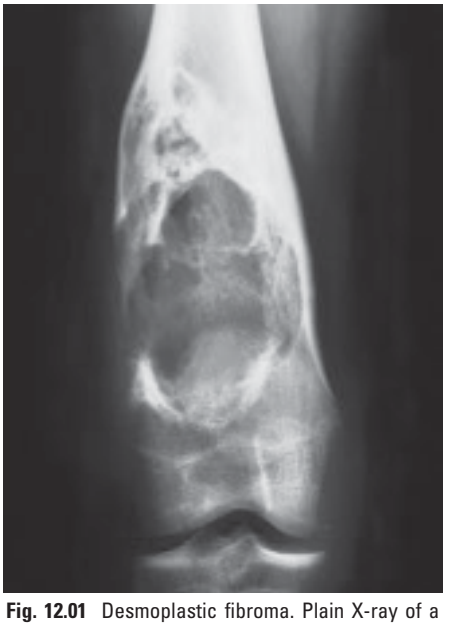

Fig. 12.01 Desmoplastic fibroma. Plain X-ray of a tumour involving the distal femur. The lesion is large, lobulated, and has a sclerotic rim.

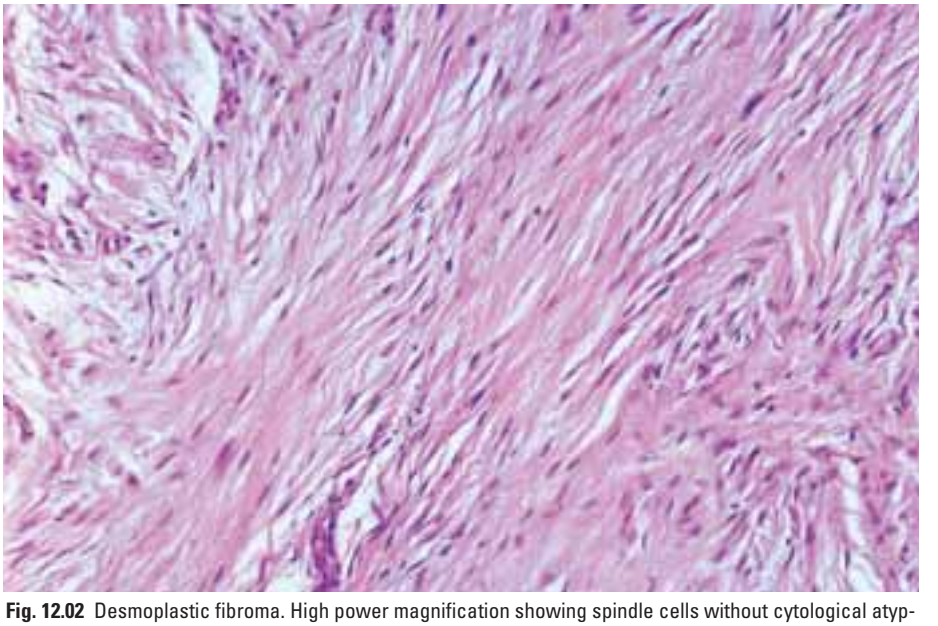

Fig. 12.02 Desmoplastic fibroma. High power magnification showing spindle cells without cytological atypia and large amounts of collagen.

Fibrosarcoma of bone

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Definition
A primary malignant spindle cell neoplasm of bone in which the tumour cells are typically organized in a fascicular or "herringbone" pattern.

ICD-O code 8810/3

Epidemiology
Precise epidemiological data pertaining to fibrosarcoma of bone is difficult to obtain due to inconsistent terminology usage for fibrosarcoma versus malignant fibrous histiocytoma.
Fibrosarcomas constitute up to 5% of all primary malignant bone tumours, with relatively uniform incidence over the second to sixth decades and equal gender distribution {991}. There have been occasional reports of cases occurring during infancy {167,425}.

Sites of involvement
Historical series indicate that fibrosarcomas most frequently involve the metaphyses of long bones. In one large series, the distal femur was involved in 48 of 102 of cases (47%) {2075}. Other frequent sites of involvement were the proximal femur (16%), distal humerus (14%) and proximal tibia (11%). A series of 130 cases also identified the distal femur as the most common site (21%) of involvement {991}.

Clinical features / Imaging
Pain and swelling are the usual symptoms. Up to one-third of patients have pathological fracture {1221}.
Radiographically, fibrosarcoma usually appears as a destructive geographic lesion, but may have an ill defined permeative, "moth eaten" appearance with cortical destruction and frequent soft tissue extension. A periosteal reaction is not infrequently present {2075}. The soft tissue extension may be better visualised by CT and MRI.

Aetiology
In most cases, the aetiology of fibrosarcoma of bone is not known. However, fibrosarcoma has been reported in association with a number of conditions including prior radiation therapy, Paget disease, giant cell tumour, osteochondroma, bone infarcts, chronic osteomyelitis, fibrous dysplasia, ameloblastic fibroma and hereditary bone dysplasia {85,644,886}.

Macroscopy
Well differentiated tumours produce large amounts of collagen, resulting in a firm consistency with a trabeculated, white cut surface and circumscribed margins. Poorly differentiated tumours have a softer, fleshy consistency with foci of necrosis; they vary in colour and are poorly marginated.

Histopathology
Histologically, fibrosarcoma of bone is composed of a uniformly cellular population of spindle shaped cells arranged in a fascicular or "herringbone" pattern with a variable amount of collagen production. Parts or all of the lesion may be more myxoid and such lesions have been labelled myxofibrosarcomas. Higher grade lesions tend to be more cellular with less collagen production, exhibit greater nuclear atypia and a higher mitotic count including abnormal mitoses than their better differentiated counterparts. Areas of necrosis may be seen.

Differential diagnosis
In cases with more severe cytological atypia, including tumour giant cells, fibrosarcoma may be difficult to distinguish from malignant fibrous histiocytoma. The presence of a storiform pattern and epithelioid type cells with "ground glass" cytoplasm would favour a diagnosis of malignant fibrous histiocytoma. In view of the identical clinical,

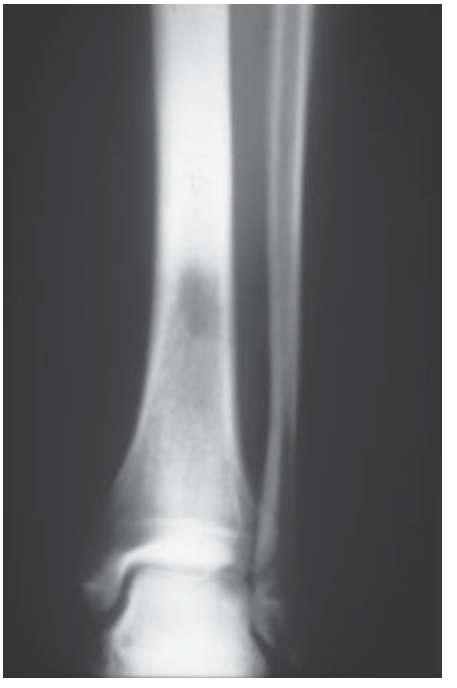

Fig. 12.03 Fibrosarcoma of tibia. Plain radiograph demonstrating ill defined purely osteolytic lesion involving distal third of tibia. The soft tissue extension of the tumour is not evident in this study.

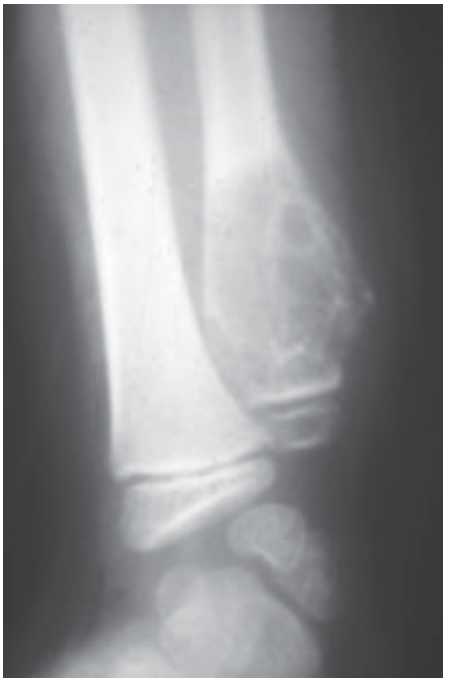

Fig. 12.04 Fibrosarcoma of ulna. Plain radiograph showing ill defined expansile osteolytic lesion of the metaphysis with cortical destruction on the medial aspect.

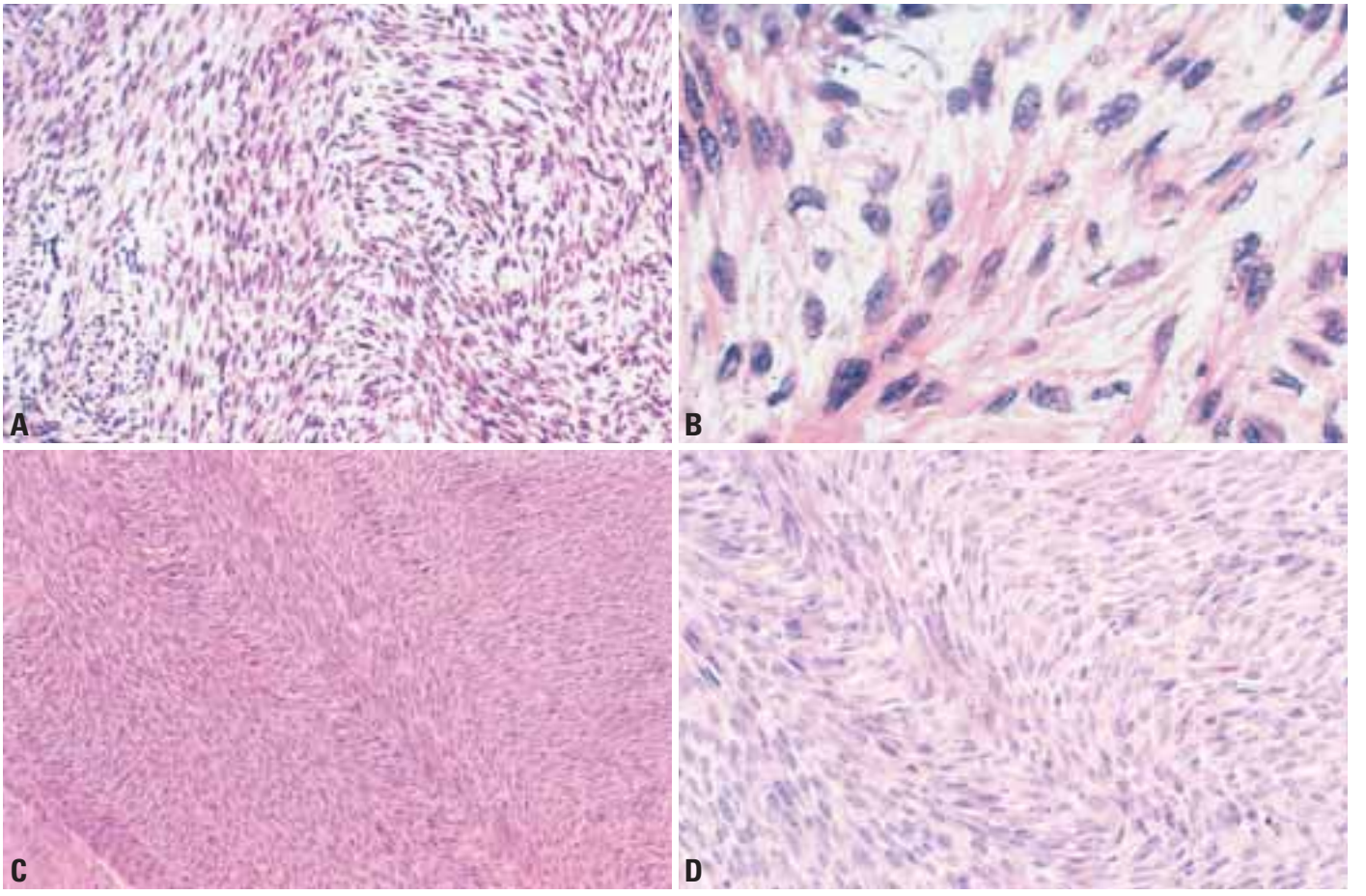

Fig. 12.05 Fibrosarcoma of tibia. **A** The fibrocytic cells are arranged in a haphazard fascicular rather than in the more typical "herring bone" pattern. **B** High power photomicrograph reveals a fairly uniform appearance of the neoplastic cells. The nuclei are ovoid, blunt-ended and have single small nucleoli and finely dispersed chromatin. Collagen fibres appear to emanate from the nuclear poles. **C** Fibrosarcoma illustrating the characteristic "herringbone" pattern. **D** High power appearance of the previous photomicrograph.

radiological and even prognostic features of these two lesions, some investigators have chosen to include them within the category of fibrosarcomas {2075}. Well differentiated fibrosarcoma is distinguished from desmoplastic fibroma by the presence of readily identifiable mitoses and high cellularity in the former and their extreme paucity or absence in the latter.

Prognostic factors

Two series have reported an overall 5-year survival approximating 34% {1647, 2075}. The most important prognostic factor is histological tumour grade. In one series, the 10 year survival was 83% in low grade and 34% in high grade fibrosarcoma {181}. Another series reported an overall 10-year survival rate of 28%, but there was a higher chance of survival (48%) in primary tumours originating from the cortical surface {991}. In the latter series, metastases occurred in 59/130 patients (45%), most frequently involving lung and other bones. In addition to poor histological differentiation, other ad- verse prognostic factors included age over 40 years and axial skeletal location {1647}.