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Solid Tumour Section

Short Communication

Bone: Fibrous dysplasia of the bone

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Identity

Note: In the major text-books on bone pathology, fibrous dysplasia (FD) is regarded as a non-neoplastic process. It is included in the chapter on "conditions that simulate a bone neoplasm" and it is defined elsewhere as a dysplastic disorder of bone.

Clinics and pathology

Etiology

Unknown.

Clinics

It is characterized by a broad clinical spectrum, varying from the more frequent solitary (monostatic) asymptomatic lesion to extensive and severe multifocal (polyostatic) lesions. The monostotic variant most commonly affects the ribs, femur and tibia of older children and young adults. The less frequent polyostotic type may be associated with endocrine abnormalities, skin hyperpigmentation and soft tissue myxoma (so-called Albright syndrome).
On radiographs, fibrous dysplasia presents as a well defined osteolytic lesion with benign features, centered within the medulla, and frequently bordered by a shell of reactive sclerosis. Depending on the relative proportion of osseous to fibrous tissue, the lesion can be completely lytic or, most typically, show a relatively high density, the so-called Œground-glass' appearance in a least part of it. Especially in bones with small diameter and flat bones, fibrous dysplasia frequently expands the bone.

Pathology

Microscopically, irregular trabeculae of woven bone are embedded in a moderately cellular fibrous matrix. The bony trabeculae have variable and irregular shapes. Osteoblastic rimming of the trabeculae is characteristically inconspicuous and some of the trabeculae seem to emerge from the surrounding fibrous background, suggesting a 'metaplastic' process. Benign lesions that histologically can be confused with FD are osteofibrous dysplasia of long bones and desmoplastic fibroma. Cases of FD with extensive cartilaginous differentiation may be mistaken histologically or radiologically for a benign chondroblastic tumor, although the adjacent fibro-osseous component allows this distinction. The most important differential diagnosis of FD is with low grade (well-differentiated) central osteosarcoma. Rarely sarcomas can arise in FD and some of them occur in patients that had received radiation therapy (post-radiation sarcomas).

Treatment

It should be conservative.

Cytogenetics

Cytogenetics Morphological

Clonal chromosome aberrations have been reported in 8 out of eleven cases of FD in which chromosomal analysis has been performed, suggesting that this entity is of a neoplastic nature. The only recurrent changes described so far in FD have been structural 12p13 aberrations (3 cases) and trisomy 2 (3 cases).

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