

Fibrocartilagenous dysplasia: a variant of fibrous dysplasia

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Abstract

Eight cases of fibrous dysplasia having enchondroma-like areas (fibrocartilagenous dysplasia) are reported. Four patients were male, four were female. The patients' ages ranged from 8 to 18 years (mean = 11.3 years). In seven cases, the lesions were located at the proximal femur while one was at the tibial diaphysis. Histologically, large, well-defined, sharply demarcated islands of hyaline cartilage tissue were seen within the typical fibro-osseous stroma of fibrous dysplasia. Some of the cartilage islands showed growth plate-like enchondral ossification at the periphery. Cartilage islands were mostly low in cellularity but infrequently, hypercellular areas were detected. Prolonged growth plate columns or developmental cartilage nests are considered in the origin of the cartilagenous component. In the differential diagnosis from benign and malignant chondroid tumours, the lack of atypia in the surrounding fibrous stroma is important.

Key words: fibrous dysplasia, fibrocartilagenous dysplasia

INTRODUCTION

Fibrous dysplasia (FD) is a benign monostotic or polyostotic fibro-osseous lesion of bone. Besides its classical fibrous and bony components, islands of hyaline cartilage are not uncommon in fibrous dysplasia.¹⁻⁴ Small irregular foci of cartilage are believed to be remnants of a previous fracture callus.⁵ However, large, tumour-like nodules of cartilage may be mistaken as a chondromatous neoplasm and even a chondrosarcoma.^{6,7} Fibrocartilagenous dysplasia is a term used to describe fibrous dysplasia with an extensive cartilagenous component.¹ We report eight cases of fibrocartilagenous dysplasia and discuss the origin of the cartilagenous component and its differential diagnosis from true cartilagenous neoplasms.

MATERIALS AND METHODS

In this study, specimens were identified from the files of Ege University Department of Pathology, Izmir. Paraffin-embedded and haematoxylin-eosin stained sections of 71 cases diagnosed as monostotic or polyostotic fibrous dysplasia during the 28 year period (1967-1995) were reviewed. Eight cases had prominent cartilage islands. These had been diagnosed as fibrous dysplasia with benign cartilage or fibrous dysplasia with enchondroma. Clinical and radiographical findings of these cases were

obtained from case records and analysed.

RESULTS

In 8 cases of FD, prominent areas of hyaline cartilage were identified. These patients' ages ranged from 8 to 18 years with a mean age of 11.3 years. Four were male and four were female. All had monostotic type fibrous dysplasia. The patients had either pain or pathological fracture. The duration of symptoms were variable, ranging from a few weeks to a few years. No other abnormalities were recorded.

Seven of the lesions were located in femoral neck region. One was in the tibial diaphysis. Radiographically, typical, well-demarcated lesions were seen in all cases.

Microscopically, in addition to the typical, fibro-osseous pattern of fibrous dysplasia, large, irregular areas of hyaline cartilage islands showing lobulation were encountered. Most were sharply demarcated from surrounding benign fibrous stroma (Fig. 1). Infrequent interdigitating extensions of cartilage islands were present within the stroma. The cartilagenous tissue was hypocellular in general, but in a few foci cellularity was increased and even binuclear, enlarged chondrocytes were noted (Fig. 2). At the periphery of some islands, enchondral ossification was striking (Fig. 3). Chondrocytes were arranged in columns simulating epiphyseal

growth plate. Newly-formed bone trabeculae had osteoblastic activity contrasting with the classical woven trabeculae of fibrous dysplasia. No anaplasia or prominent mitotic activity was present in the **fibro-osseous** component. Bone trabeculae within the loose fibrous stroma were woven, irregular, curved and in "letter soup" pattern, without osteoblastic rimming. The fibrous stroma was moderately cellular having no evidence of **storiform** pattern and anaplasia. Clinical follow-up did not detect recurrence in any of the cases.

DISCUSSION

Fibrous dysplasia is well known to have infrequent cartilaginous areas within its **fibro-osseous stroma**.^{1-5,9} The amount of this cartilage tissue is widely variable, ranging from small microscopical foci to radiographically evident extensive areas of hyaline cartilage sometimes even resembling a primary cartilage neoplasm. Some studies have reported polyostotic fibrous dysplasia cases with cartilage **areas**.^{8,10} but all our cases were monostotic.

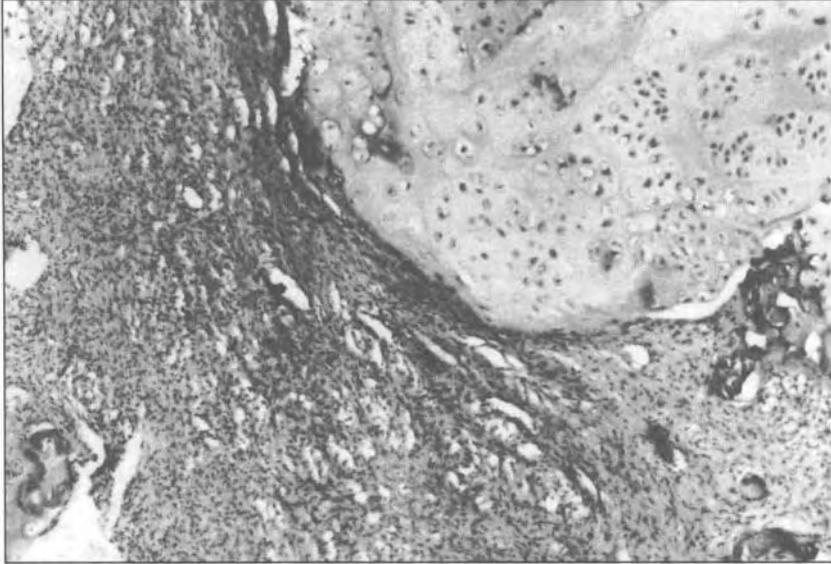


FIG. 1: Lobular cartilage islands with sharp demarcation from surrounding typical fibrous dysplasia stroma. (H&E x100)

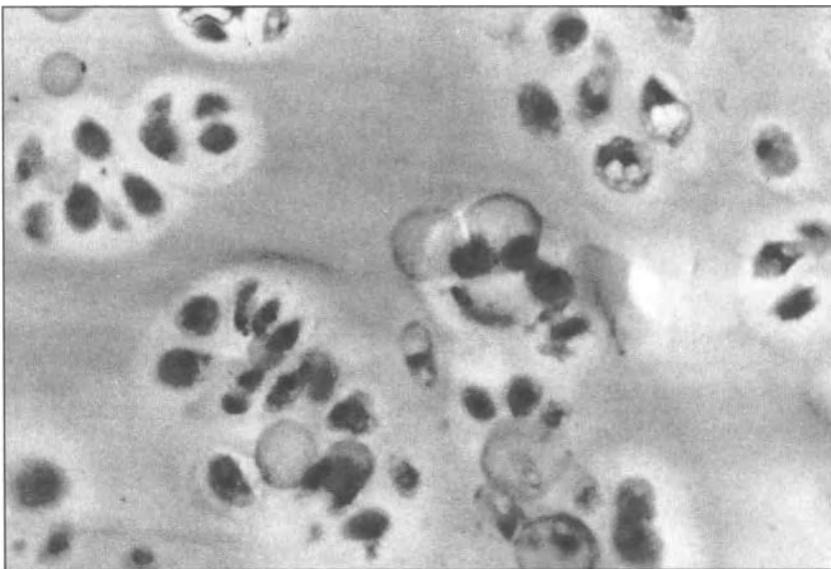


FIG. 2: Occasional enlarged **binuclear** chondrocytes were present within the islands. (H&E x 400)



FIG. 3: Cartilage islands with peripheral enchondral ossification and occasional growth plate-like columnar chondrocytes. (H&E x 40)

Histologically, some of the cartilage areas have high **cellularity** and atypical chondrocytes. These findings may easily mislead a pathologist, especially one with little experience in fibro-osseous lesions, towards the diagnosis of a chondromatous neoplasm even a chondrosarcoma. To avoid this, radiographical analysis and the histology of the benign fibro-osseous component have to be considered.

Radiologically all the lesions were well-demarcated. Some had stippled calcifications interpreted as cartilaginous areas. Annular or ring calcifications in fibrous dysplasia have been reported to be the result of enchondral calcification and ossification within the cartilage islands or chondromata present in the lesion." Most of the cases were located in trochanteric area of the proximal femur. This site is the most frequent site for fibrocartilaginous **dysplasia**.^{8,12} In one case, the tibial diaphysis was involved. Neither our cases nor any in the literature was located in the craniofacial **bones**.^{6,8,10,12} This was in contrast to typical fibrous dysplasia, which has a predilection for craniofacial bones. A suggested origin from prolonged columns of epiphyseal growth **plate**.^{5,13} was consistent with long bone localization. Since craniofacial bones mostly show intramembraneous ossification, the infrequency of fibrocartilaginous dysplasia in this site is not surprising. Focally prolonged columns of chondrocytes may extend deep into the metaphyseal **bone**.¹³ **Sanerkin**¹¹ proposed that the cartilaginous islands arose as

developmental rests from epiphyseal cartilage and that these cartilage tissues developed simultaneously with fibrous dysplasia. Peripheral enchondral ossification is frequently encountered in cartilage islands. Newly formed bony trabeculae have osteoblastic activity in contrast to typical bony components of fibrous dysplasia which lack osteoblastic rimming. This also supports the 'rest' nature of cartilage islands rather than their occurrence as a true component of the lesion. Also, **Scherer**¹³ found nests of benign cartilage as incidental findings in 20 of 1125 femora he examined at autopsy. The process of mineralisation of both bony and cartilaginous tissues appeared similar to normal bone and **cartilage**.¹⁴ In the femoral neck region, trochanteric growth plates also provide a setting for the cartilaginous component in fibrous dysplasia. The young age of the patients is in favour of a growth plate origin. In our cases the mean age of fibrocartilaginous dysplasia was 11.3 years. For the pure fibrous dysplasias, this was 21.8 years. Skeletal immaturity and open epiphyseal growth plate may contribute to the development of fibrocartilaginous dysplasia which occurs even earlier than fibrous dysplasia.

In terms of cellularity, cartilage nodules are usually hypocellular with small chondrocytes. However not uncommonly hypercellular areas with slightly atypical, binuclear, enlarged chondrocytes are seen.' This may be misinterpreted as chondroid neoplasia. A well-demarcated nodular structure with an adjacent

typical benign, fibro-osseous stroma should warn against a diagnosis of chondroid neoplasm. Besides, frequent peripheral enchondral ossification similar to epiphyseal growth plate is an important diagnostic clue.

Low-grade tumorous cartilage tissue juxtaposed against fibrous stroma may be misinterpreted as dedifferentiated chondrosarcoma. Fibrous stromal elements should be carefully studied. In dedifferentiated chondrosarcoma, high-grade spindle cells are frequently arranged in storiform pattern. Fibrosarcoma and malignant fibrous histiocytoma are frequent non-cartilaginous components of dedifferentiated chondrosarcoma.^{15,16} Mesenchymal chondrosarcoma has its cartilage speckled throughout the tumour and mixed with a spindle- or small-cell mesenchymal component.⁷ In fibrocartilaginous dysplasia, cellular areas of fibro-osseous stroma has to be closely searched for anaplasia before chondrosarcoma can be excluded from the differential diagnosis.

Other infrequent lesions that need to be considered in the differential diagnosis are desmoid tumour with enchondroma-like nodules and low grade fibrosarcoma with enchondroma-like nodules.¹⁷ These lesions have been termed as fibrocartilaginous mesenchymoma with low grade malignancy⁷ and fibrocartilaginous mesenchymoma.⁴ In these lesions, the histological appearance of the fibrous component is either an aggressive fibromatosis or a cellular fibroblastic lesion with minimal anaplasia that is consistent with low-grade fibrosarcoma.^{4,5} The typical benign bony component of fibrous dysplasia is lacking.

Malignant change in fibrous dysplasia is rare. Osteosarcoma and fibrosarcoma are more common than chondrosarcoma.¹⁸ In reported cases, a late age of onset, previous irradiation, cortical and soft tissue invasion, and a malignant course are noted.¹⁷⁻²⁰ However, cases previously diagnosed as chondrosarcoma lacking a typical onset and malignant course might have been fibrocartilaginous dysplasias. Ishida^x mentioned several cases in the literature exhibiting a benign course but were diagnosed as chondrosarcoma derived from fibrous dysplasia.

In conclusion, fibrous dysplasia is a common benign fibro-osseous entity having a spectrum of histological appearances. Differing ratios of bone to fibrous tissue, varying cellularity of the fibrous component, aneurysmal or simple bone cysts, acellular cement-like material are the histological variations that one may encounter."

Enchondroma-like areas further complicate the picture. Though the femoral neck region is the most frequent location for this latter variant, other bony areas close to the epiphyseal plate can theoretically contribute to fibrocartilaginous dysplasia.

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