Case Report

Fibrocartilagenous dysplasia: A rare variant of fibrous dysplasia

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ABSTRACT

Fibrous dysplasia (FD) is a benign disorder affecting one or more bones seen in all age groups and shows no geographical, racial and sex predilection. Fibrocartilaginous dysplasia (FCD) is a type of fibrous dysplasia where there is extensive cartilaginous differentiation. Our patient was a young girl with a 2 year history of pain in the right forearm and hip. Imaging studies showed multiple lucent lesions in the femur, humerus and radius. Microscopic examination of the curetted fragments from the lesion showed predominant cartilaginous components with focal fibro-osseous areas and areas of enchondral ossification. We present this case due to its rarity and the importance of distinguishing FCD from other benign and malignant cartilaginous tumors as it mimics these in clinical and histological features.

Keywords: *Fibrous dysplasia, fibrocartilaginous dysplasia, femur*

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Introduction

Fibrous dysplasia (FD) is a benign localized intramedullary proliferation of trabecular woven bone admixed with fibrous tissue. It may be monostotic, involving one bone or polyostotic involving several bones.[1] In addition to fibrous and bony components, the lesion may include benign cartilage. This is usually seen as small and irregular foci. Massive chondroid differentiation in FD is extremely rare and is known as fibrocartilaginous dysplasia.[2] This case report highlights the clinicopathological aspect of fibrocartilaginous dysplasia.

Case report

An 18-year-old female, presented with pain in the right forearm and hip of 2 years duration. Two weeks before she had sustained a fall following which the pain in the hip region has worsened. She also developed a limp during walking. She had a fracture of right forearm about 2 years back and was treated conservatively at a local hospital. X-ray showed extensive lytic lesions in the medullary cavity of shaft of right femur, proximal right humerus and radius [Figure 1a]. There was also an old united fracture of the proximal third of right radius.

She underwent curettage of right femur lesion followed by bone grafting and intramedullary nail fixation. Histopathology of the lesion showed predominantly lobules of benign hyaline cartilage with chondrocytes showing focal increase in cellularity with few binucleate and multinucleate cells resembling the enchondroma like areas [Figure 1b and c]. However, extensive sampling of the tissue revealed focal fibroosseous component comprising spindly fibroblasts and irregular trabeculae of immature bone, showing occasional osteoblastic rimming [Figures 1d, 2c and d]. Also noted were areas of enchondral ossification [Figure 2a and b]. Hence, the final diagnosis of polyostotic fibrocartilaginous dysplasia was made. Follow up of the patient over a period of 2 years showed considerable symptomatic relief and no further progress of the other lesions.

Discussion

Fibrous dysplasia is a genetic noninherited condition caused by missense mutation in the GNAS1 gene on chromosome 20. The polyostotic variety may be associated with endocrine dysfunction, abnormal pigmentation, and precocious puberty in girls (McCune Albright syndrome).[2,3] Patients with FD may be asymptomatic, or may present with pain or pathological fractures. The presence of cartilage in FD is small and irregular. Cartilage in the FD is either intrinsic to the lesion or secondary

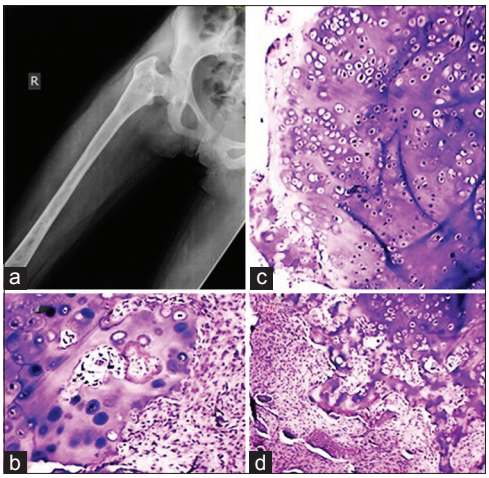

Figure 1: (a) X- ray showing lytic lesion in the medullary cavity of right femur (b) Histopathology showing lubules of cartilage with increased cellularity [H and E, × 100] (c) Histopathology showing cartilage tissue with focal fibroblastic proliferation [H and E, × 100] (d) Histopathology showing enchondral ossification [H and E, × 100]

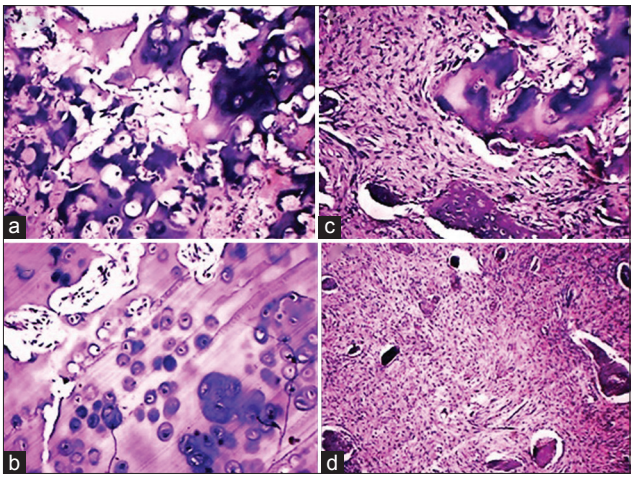

Figure 2: (a and b) Cartilage with fibro osseous lesion and enchondral ossification [H and E, × 100] (c and d) Typical fibrous dysplasia areas with bony trabeculae with fibrous stroma [H and E, × 100]

to fracture, or may result from disruption of an affected growth plate during childhood.[4] These are commonly seen with the polyostotic variety of FD.[5] Cartilaginous foci in FD of the jaws or skull is very rare.[1] When cartilage tissue dominates in the histopathology of FD, the term fibrocartilaginous dysplasia is used. It is a benign condition that occurs in patients between the ages of 4 and 26 years and is mostly located in the proximal femur.[2] Our patient had multiple lytic lesions in the femur, humerus and radius. There were no endocrine abnormalities or any other lesions.

In our case, histopathological finding was consistent with fibrocartilaginous dysplasia. The cartilaginous tissue with focal high cellularity and atypical chondrocytes may lead to confusion in diagnosis and the lesion may be mistaken for a chondrosarcoma.[6] The differential diagnosis includes enchondroma, dedifferentiated chondrosarcoma, intramedullary osteosarcoma and fibrocartilaginous mesenchymoma.[2,3,5] Radiological findings and histology of the benign fibro-osseous areas in addition to clinical correlation are vital to the diagnosis. Enchondroma is a common benign cartilaginous tumor that occurs most frequently in small bones of the hands and feet, particularly the proximal phalanges. Histologically, it is composed of mature lobules of hyaline cartilage. Foci of myxoid degeneration, calcification, and endochondral ossification are common. The common malignant bone tumor in young age group exhibiting cartilage formation is osteosarcoma with predominant cartilaginous component. Chondrosarcoma comprises 5% or fewer of all primary malignant skeletal tumors in the first two decades of life.[7] Three variants of chondrosarcoma are recognized in children: Mesenchymal, clear cell, and myxoid. Histologically, these variants of chondrosarcoma lack the fibroosseous component characteristic of FD. The presence of frequent areas of enchondral ossification helps to differentiate it from conventional chondrosarcoma. Dedifferentiated chondrosarcomas are composed of malignant cartilage and the fibrous stromal elements are high-grade spindle cells arranged in storiform pattern.[2,6] In the case of well-differentiated intramedullary osteosarcoma most patients are adults, the femur, and tibia being the most commonly affected sites. Histological features simulating FD and small foci of atypical cartilage may be present. However, areas of high-grade pleomorphic osteosarcoma will be seen. Fibrocartilaginous mesenchymoma is an extremely rare lesion that tends to affect the metaphyseal region of bones, particularly the proximal fibula. The lesion contains fibrous areas, islands of cartilage and bony trabeculae that show a pattern of organization reminiscent of epiphyseal growth plates.[8] These tumors show an exuberant spindle cell component with elongated and hyperchromatic spindle cells whereas in FCD, spindle cells are short and without hyperchromasia.[2,9] It is locally aggressive with a high frequency of recurrence, especially when resection is incomplete.

The amount of cartilage has been made to consider the diagnosis of FCD, but in our case, 85% of the lesion was composed by hyaline cartilage. Treatment consists of curettage, correction of deformities and fractures. Spontaneous resolution of the lesion has been reported in the tibia.[10]

In conclusion, it is important to recognize this rare variant of FD with benign nature, to avoid misdiagnosis as a cartilage neoplasm and its unnecessary treatment.

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