

Case report

Primary Xanthoma of Femur: A Case Report

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Abstract:

Xanthoma of bone is a very rare benign bone disorder with features mimicking primary bone tumours or metastatic lesions and their definitive diagnosis is very difficult. This condition can occur in patients with both normal and abnormal lipid profiles. Xanthoma of femur in a normolipidemic patient is very rare. Histopathological study gives a definitive diagnosis. We report a case of a 17 year old patient who was hospitalized in orthopedic department for fracture of the right femur with a lytic lesion at the fracture site. The patient was normolipidemic and hypothyroid. The diagnosis of xanthoma was selected histomorphologically. The fracture was treated surgically by nailing and bone graft.

Keywords: bone; femur; xanthoma; benign; normolipidic

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Introduction:

Xanthoma derived from the Greek “Xanthos” meaning yellow¹. Xanthomas are benign lesions which may originate from abnormal deposition of cholesterol and lipid into tissues leading to inflammatory cell infiltration. They affect various parts of the body; skin, tendons, flat bones, cerebral paranchyma etc.². The most common sites of solitary xanthomas are areas subjected to repeated minor trauma, such as elbows, buttocks, and patellar and Achilles tendons³. Primary

bone invasion by xanthoma is rare⁴. Intraosseous xanthomas are lytic, expansile lesions, histologically characterized by mononuclear macrophage-like cells, abundant foam cells, and multi-nucleated giant cells. Occasionally, spindle cells are pre-sent, which has led investigators to include this lesion as a subset of benign fibrous histiocytoma (BFH) of bone⁵. These lesions usually occur in the appendicular skeleton, especially the long bones such as the tibia. No bone is spared, however, and lesions can occur anywhere



Fig 1: Fracture and lytic lesion of femur [X-Ray]



Fig 2: After bone graft and nailing [X-Ray]

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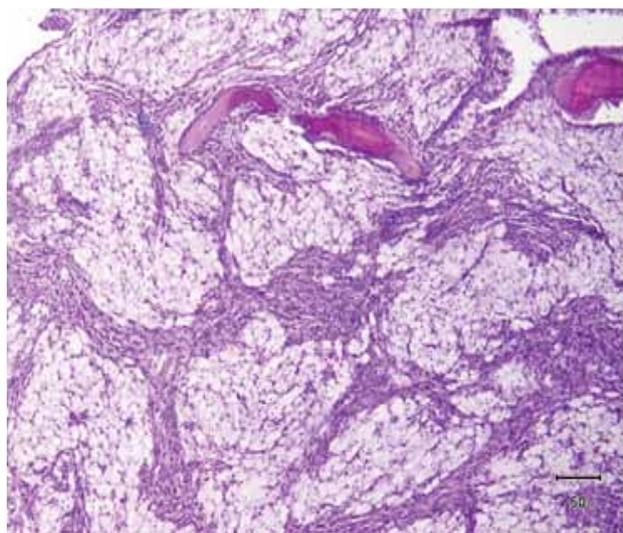


Fig 3: Lobules of foamy cells[40X, H&E stain]

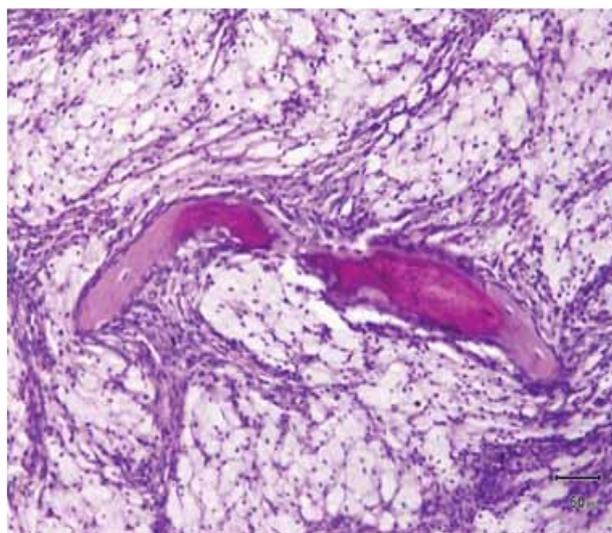


Fig 4: Foamy histiocytes with bony chips[200X]

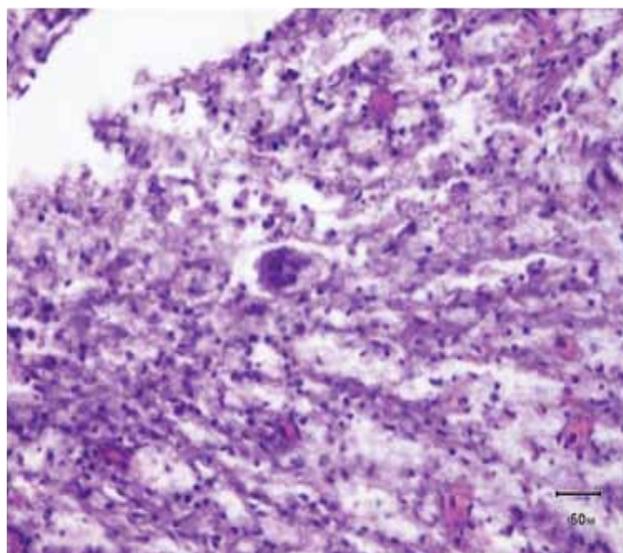


Fig 5: Foam cells, tufton giant cells [400X]

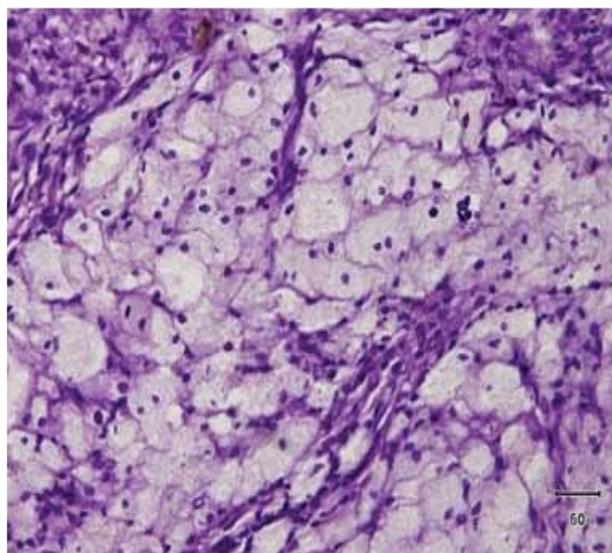


Fig 6: Foam cells with fibrous proliferation

in the appendicular or axial skeleton including the bony pelvis, spine and skull base⁶. We present an unusual case of pathological fracture of lower end of femur following a slip and fall, and were found to have primary xanthoma of femur.

Case report:

A 17 years boy presented with the history of swelling and pain around right knee, difficulty in walking and shortening of his right leg, following a slip and fall. On examination there was a diffuse swelling, tenderness and shortening of right leg. The radiography showed fracture of the lower end of the right femur with osteolytic lesion at the fracture site [Figure 1].

No obvious extracortical tissue extension was seen, with normal joint space and articular surface maintained. Biochemical investigation revealed that serum calcium, alkaline phosphatase and lipid profile within normal limit. He is a known case of

hypothyroidism since 2 years for which he is under treatment of endocrinologist. No abnormality was noted in remaining other system. Following this, curetting biopsy of the lesion with bone graft and nailing was done. Biopsy material was sent to private histopathological laboratory for a definitive diagnosis of the lesion. Macroscopically biopsied material was yellow in color, soft to firm in consistency. Histopathological examination of the tissue showed sheets of lipid laden foam cells interspersed in to the trabeculae embedded in a variable background of other cells, including tufton giant cells and fibrosis also noted [Figure: 3-6].

Immunohistochemical examination was not possible due to unavailability in our setup. So, histomorphologically this case was diagnosed as xanthoma of bone. Histologically benign fibrous histiocytoma and non ossifying fibroma were

considered as differential diagnosis. The benign fibrous histiocytoma shows spindle cells arranged in storiform pattern which was lacking in our case, where as non ossifying fibroma shows cellular fibrous tissue arranged in storiform patterns, scattered osteoclasts, foamy & hemosiderin laden macrophages and sometimes bizarre nuclei, and all these features were lacking in our case. And thus depending on the above findings and radiological and biochemical findings the final diagnosis was rendered as primary xanthoma of the right femoral bone. The case was treated surgically by bone graft and nailing [Figure: 2].

Discussion:

Bone xanthoma is a rare disorder due to the presence of cholesterol deposits in the bone. It is usually found in hyperlipidemia and hyperlipoproteinemia family. These are characterized by elevated levels of cholesterol that form deposits in soft tissue and bone. These are also reported in nonhyperlipidemic states. The most frequent location of bone xanthoma is the diaphysis of long bones, especially the tibia. Other locations may be particularly in the facial skeleton, mastoid air cells, and mandibular bone. The skull can be affected (temporal or frontal) which may cause cerebellar compression⁷. The pathogenesis of the xanthomas consists of lipid leakage from the blood vessels in the site of the lesion, with subsequent phagocytosis of this material by the macrophages. The non-degraded cholesterol

accumulates within the cytoplasm, leading to the presence of foamy macrophages. Minor trauma is also involved in the development of xanthomas. Particularly in primary bone lesions, the xanthomas can apparently occur in a pre-existing lesion, such as a simple bone cyst, aneurysmal bone cyst and fibrous dysplasia⁸. Typically, osseous xanthoma appears lytic with a narrow zone of transition. A sclerotic rim is often completely or partly present, making it difficult to differentiate from more common benign lesions such as nonossifying fibroma, fibrous dysplasia, simple bone cyst or brown tumor of hyperparathyroidism⁶. Histologic evaluation usually demonstrates multinucleated giant cells with a fine, ground-glass appearance, and in some cases, foamy or vacuolated histiocytes, as in our patient's case. Immunohistochemistry is variable, but in most cases, staining is positive for CD68, CD4, CD45, HLA-DR, and lysozyme, but negative for S100, CD20, and factor XIIIa⁹. Oil Red O and scarlet red stains confirmed the presence of neutral fat¹⁰.

Skeletal xanthomas are very rare lytic lesion of bone whose prognosis is good. The xanthomatous degeneration occurring in other bone tumors need to be evaluated carefully in whole of the specimen to avoid erroneous diagnosis of primary xanthoma. In view of the rare intraosseous location in normolipidemic individual and with the histology corresponding to a tuberous xanthoma, our case is unique localization involving femur and hence this is being presented.

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