

Focal Hematopoietic Hyperplasia of Rib, Mimicking to Fibrous Dysplasia

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ABSTRACT

Focal haematopoietic hyperplasia is a rare and localized proliferation of the bone marrow to such an extent that it produces a tumor-like expansion hence called as pseudotumor. Radiologically it presents as an osteolytic expansile mass and may mimics with certain bony tumors. We report a case of Focal haematopoietic hyperplasia in the 3rd anterior rib of a 45 year old man clinically presenting as a bone tumor and radiologically mimicking to fibrous dysplasia.

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Introduction

Focal haematopoietic hyperplasia (FHH) of bone is a very rare lesion and only few cases have been reported till date [1-3]. It is characterized by the presence of a tumor-like mass in the bone which on radiology present as an osteolytic and expansile bony lesion with or without cortical destruction. Histologic examination of the lesion reveals only hypercellular red marrow elements merging with fatty marrow and without any other associated significant histopathologic lesion. It involves the rib in most of the cases however two cases in vertebral bodies are also been reported [1-5]. This lesion is considered as a form of pseudo tumor as the process is benign proliferative presenting as a mass lesion. Here we report a case of focal hematopoietic hyperplasia occurring in the 3rd anterior rib and clinically presenting as a bony tumor.

Case Report

A 45-year-old man presented with a mass over right lateral aspect of chest wall gradually increasing in size for the last 4 months. Initially it was painless but now from last 20 days there is continuous dull aching pain. On local examination there was hard bony mass of approx 7x6cm on right lateral aspect of chest wall in relation with the shaft of right 3rd rib. It was regular with smooth surface and mildly tender.

Chest X-ray revealed an expansile bony lesion in the anterior part of right third rib with ground glass matrix (Fig 1A) Computed tomography (CT) revealed that it was arising from the shaft of the rib. The cortex was ballooned and thin with internal calcification within it. There was no evidence of cortical destruction, soft tissue mass or periosteal

reaction (Fig 1B). Based on the above radiological findings a diagnosis of fibrous dysplasia was suggested. He had no history of hypertension, diabetes mellitus, hematologic malignancy or other solid malignancies, or any trauma involving ribs. There was no other abnormal finding on general physical examination. Hematologic examinations at admission were within normal limits, with hemoglobin of 13.4 g/dl, hematocrit of 44.5%, white blood cell count of 7400/mm³ with normal morphology. Other laboratory tests including all biochemical investigations showed normal values. Wide marginal excision of the rib was performed.

Gross examination showed a resected rib containing a globular expansile mass 7x6cm in size, having a smooth outer surface. Cross section revealed a soft grey brown mass with yellow white areas in between. Cortex was thinned out and bulging, however there was no cortical breach. There was no separate demarcation between the lesion and the adjacent normal marrow. (Fig 2A & B) Microscopic examination revealed a hypercellular marrow with a cellularity >70% merging with fatty marrow. Trilineage haematopoiesis was seen with normal maturation of the erythroid, myeloid as well as the megakaryocytic series cells.

The myeloid and erythroid ratio was normal. Megakaryocytes were adequate in number with a normal morphology. Both thin and thick types of bony trabeculae with extensive areas of calcification were seen. There was no osteoclastic activity and no atypical or abnormal cell collection was identified. Focal area of fibrosis was also evident. (Fig 3 A to D)

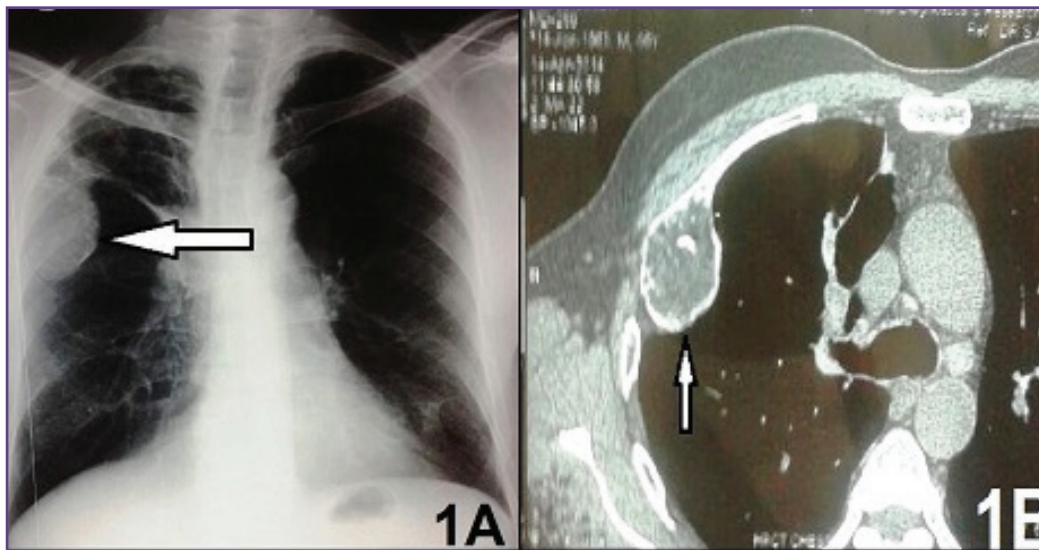


Fig. 1 A. Chest radiograph reveals an expanding bony mass in the anterior part of right third rib with ground glass matrix B. Plain CT scan thorax a well corticated expansile rib lesion with soft tissue density and foci of calcification within it

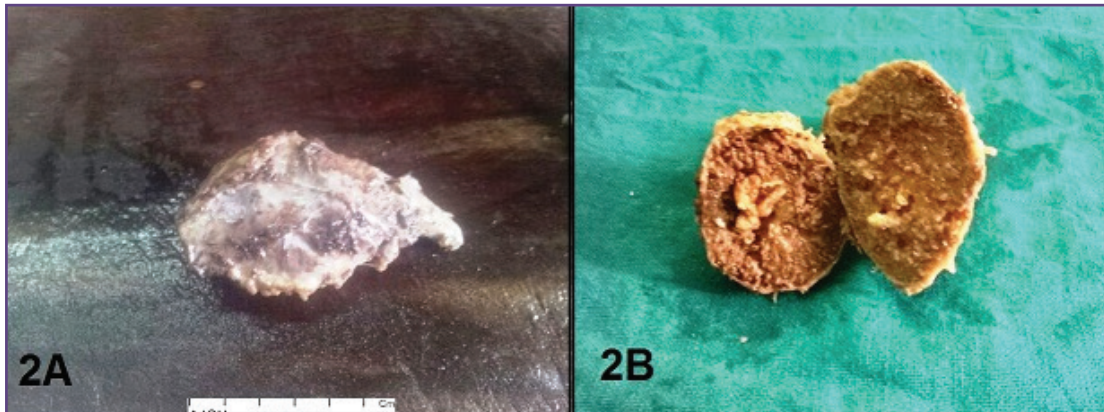


Fig. 2 A. A globular well demarcated mass lesion B. Cross-section of the tumor demonstrating grey brown areas with a thin and bulging cortex

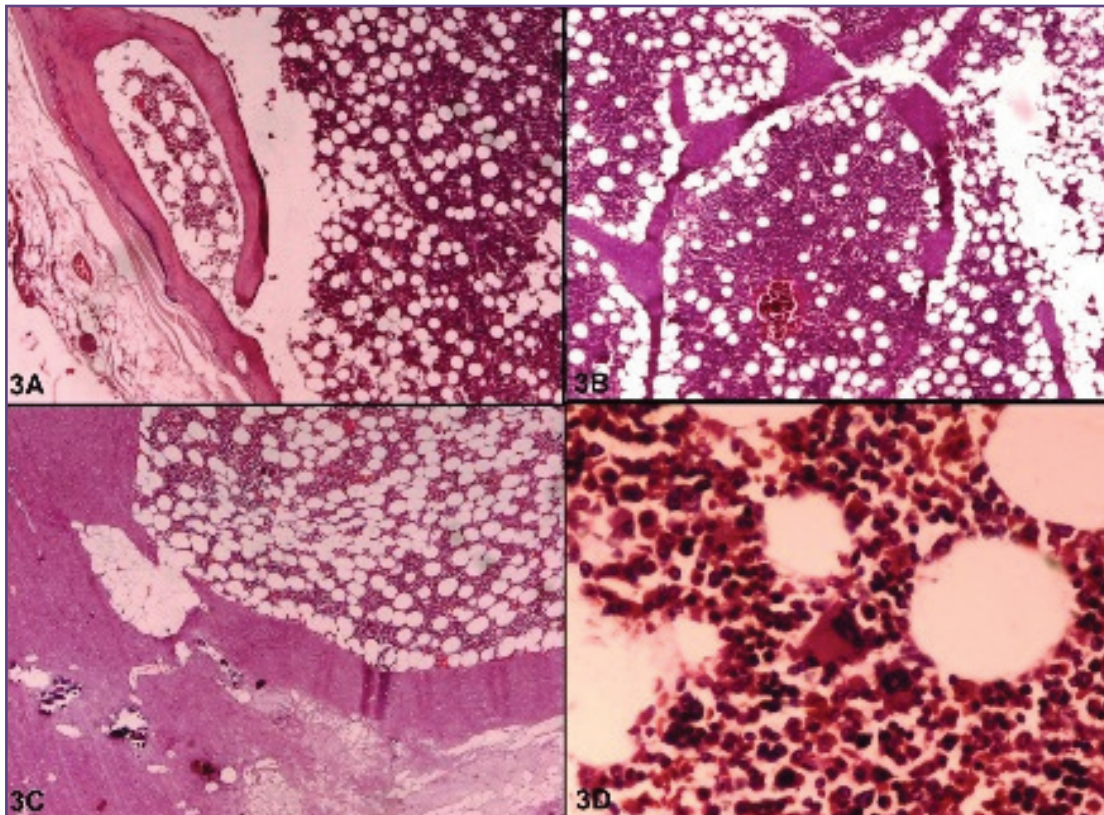


Fig. 3. A Low magnification showing well demarcated tumor tissue composed of hematopoietic bone marrow with interspersed adipocytes. Note outer thinned out cortical bone (H&E, x100) B. Hypercellular marrow merging with fatty marrow with intervening woven bone trabeculae. The cellularity is different from area to area. (H&E, x100) C. Thick fibrous band with area of calcification (H&E, x100) D. high magnification showing normal trilineage haematopoiesis. (H&E, x400)

Discussion

Focal haematopoietic hyperplasia is a rare and localized proliferation of the bone marrow to such a degree that it produces a tumor-like expansion and hence called a

pseudotumor. From the clinical course and follow-up information, the process is considered as nonneoplastic benign proliferative lesion in response to trauma or any other stressful condition. Some other author

considers it a deviation from the adult pattern of red-yellow marrow distribution, with an increase of the red marrow components. Edelman proposed two mechanisms regarding pathogenesis of marrow hyperplasia [1] one could be in response to trauma, inflammation or other stressful condition and other could be preexisting developmental anomaly.

However we could not find any history of trauma, stressful condition or any detectable developmental anomaly in our case similar to other reported cases. Our case is clinically, radiologically and histologically similar to previous four reported cases where patient was presenting as expansile lytic mass lesion involving the rib [1-3]. Radiologically these lesions are characterized by an expansile and radiolucent lesion and contained ill defined linear areas of increased density or calcification. The cortex was thinned but intact, and no soft tissue extension was noted [3]. Radiologic differential diagnosis should have included fibrous dysplasia, aneurysmal bone cyst, plasmacytoma, osteomyelitis and osteochondroma. Our case on radiology has a diagnosis of fibrous dysplasia, a benign lesion. PET scan or bone scan was not done in this case as clinical and radiological findings were not suggestive of any malignancy or other disease with systemic involvement. Histologically the marrow consisted of a proliferation of all three lineage hematopoietic components including erythroid, myeloid and megakaryocytic lineage. The cellularity was increased by approximately 50-95%. The trabeculae of the cancellous bone were thin and disrupted, which resulted in severe osteoporosis, osteoclastic activity did not appear to be increased [3]. Apart from these aforementioned features we found thick, distorted bony trabeculae, thick bands of fibrosis and extensive calcification which was seen as a feature of increased bone density and calcification on the radiographs. There were no abnormal or atypical cells found in our case similar to the other cases reported in literature. Other bony lesions which may show similar histologic features include myelolipoma and some hematologic disorders showing increased hematopoiesis such as chronic anemia, thalassemia [6], leukemia and myelofibrosis [7]. Two cases of intraosseous myelolipoma associated with prostate and colon cancer has also been reported in the literature [8, 9].

Myelolipoma is defined as a tumor like lesion composed of admixture of mature adipose tissue and hematopoietic elements in a variable proportion, most commonly involving the adrenal gland. Most of the cases encountered as extra adrenal myelolipomas have occurred in soft tissue especially in retroperitoneum[10]. Occasional cases of

intraosseous myelolipomas with the similar microscopic findings as in FHH have also been reported, however clinical and radiological findings in these cases are entirely different [8, 9]. Intraosseous myelolipoma are sclerotic lesion, do not contain reticular sinusoids or bone spicules and are normocellular for that age. Contrast to it our case has osteolytic expansile mass lesion containing thin and thick bony trabeculae along with hypercellular marrow. Thalassemia patients may show expansion of marrow cavity, cortical thinning and marrow hyperplasia. Cortical expansion is prominent in the posterior aspect of the rib [6]. However, the bony involvement of thalassemia is usually multiple and frequently involves the skull. Also, the clinical features of thalassemia of anemia and hepatosplenomegaly were absent in our case.

Myelofibrosis is one of the myeloproliferative disorders manifested by a trilineage proliferation of cells including normoblasts, granulocyte precursors and megakaryocytes [7]. It causes a varying degree of marrow fibrosis and diffuse osteosclerosis. Most patients present with anemia and hepatosplenomegaly. Our case had some features of myelofibrosis, such as hypercellularity and focal fibrosis, but the fibrosis was present focally and most bony trabeculae were thin and osteoporotic rather than sclerotic. There have been no reports of myelofibrosis presenting with an expanding tumor mass of bone.

Other findings reported to be associated with benign hyperplasia of bone marrow such as obesity, cigarette smoking with peripheral leucocytosis, administration of granulocyte colony stimulating factors or past history of malignancy were absent in our case.

Conclusion

FHH is although a rare entity, a clinician and radiologist should aware about this as it mimics and also found to be associated with a number of lesions as described above. There is no set algorithm or recommendations to diagnose FHH due to its extreme rarity. Awareness of this entity will avoid unnecessary whole body scans, repeated biopsy procedures and potentially large, complicated surgical procedures. These types of lesions should be managed conservatively.

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Competing Interests

None declared

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