CYSTIC ANGIOMATOSIS: A RARE CAUSE OF OSTEOLYTIC LESIONS AND PATHOLOGICAL FRACTURE- A CASE REPORT

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ABSTRACT:

Cystic angiomatosis is an uncommon benign multifocal disorder in which there is proliferation of endothelial lined vessels in bone. Angiomatous malformation results in progressive loss of bone. Bony cystic lesion affects the axial and proximal appendicular skeleton. Patients usually presents with pain. We report a case of 58 year old man who presented with pain and pathological fracture. X-Ray imagery showed multicystic lesions involving the whole length of multiple ribs. Rib biopsy showed the proliferating capillary network growing in to the bone marrow spaces associated with destruction of bone. Cystic angiomatosis should be kept as one of the rare differential diagnosis in a case presenting with multiple lytic lesions of bone.

INTRODUCTION

Cystic angiomatosis is rare clinical entity characterized histologically by vascular malformation within bone leading to destruction of bone.[1,2] The exact cause of cystic angiomatosis is not clear. However, it is believed that it is congenital vascular hamartoma. Extensive search of literature revealed that less than 2000 cases have been described in world literature.[2] There are few cases of cystic angiomatosis which presented with pathological fracture. This case has been presented as it is a rare cause of multiple osteolytic lesions and pathological fractures.

CASE REPORT

A 58-year-old man presented in the orthopedic clinic with a pain in the right anterior thoracic wall for more than a month. The pain was constant and worsened by any movement. There was no paresthesia. There was no history of anorexia, weight lost or fever. Hematological investigations showed total leukocyte count, differential leucocyte count, hemoglobin and platelets to be in normal range. Erythrocyte sedimentation rate was 52 mm/hour. Biochemical investigations revealed lactate dehydrogenase, alkaline phosphatase and ionized calcium in the normal range. Renal and hepatic function test were within normal limits. X-ray imaging showed multicystic expansive lytic areas involving almost the whole of length of all the ribs of the chest. Pathological fractures were seen in 5th, 6ths nf 7th ribs on right side. [Fig.1]. Proximal humerus was also involved.

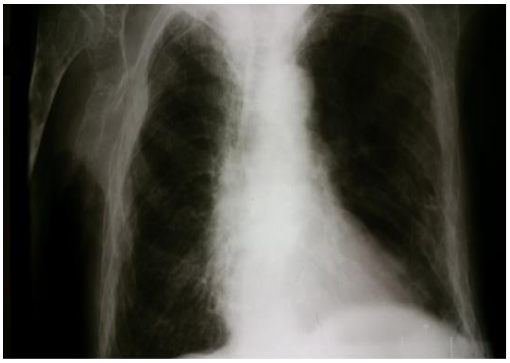

Fig. 1: showing osteolytic lesions involving the whole length of ribs with fractures of 5th, 6th and 7th ribs in right hemithorax

A biopsy was taken from one of the fractured ribs. The histological section showed replacement of bone by proliferating capillaries [Fig. 2].

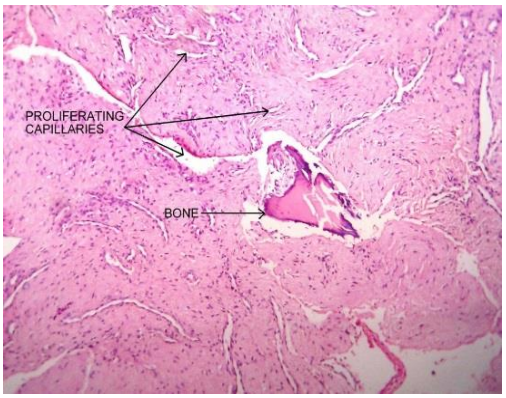

Fig. 2: Histopathological section showing piece of bone that is surrounded by proliferating endothelial lined capillaries. (hematoxylin and eosin stain x 200)

Patients was put on bisphosphonates and started on radiotherapy. The patient was asked to report monthly for follow up for progression of disease.

DISCUSSION

J.B.S. Jackson published the first case of spontaneously dissolving bone in 1838 [3]. Skeletal angiomatosis is an uncommon disease of unknown cause characterized by the growth of endothelial-lined blood/lymphatic vessels in bone leading to distruction of bone [2,4]. Skeletal angiomatosis is most commonly seen below 25 years of age but few cases have been documented in older individual.

Skeletal angiomatosis commonly affects the maxilla, mandible, ribs, clavicle, cervical vertebrae, femur and pelvis. Mandible is the commonest site of bone distruction. The earliest changes on x- ray imagery are the foci of subcortical and intramedullary lucency mimicking osteoporosis.[5,6] These foci usually become large and coalesce with advancing disease . [7] These lesions can cause the bones to shrink leading them to what is called as "sucked candy" appearance. On the other hand this can lead to resorption of entire bone. This whole process can also undergo spontaneous arrest and get stabilized. [4]

The commonest symptom of skeletal angiomatosis is pain. Bone fracture may be presenting complaint with or without trauma. Patients in whom thoracic rib is involved may present with chylothorax. Our patient presented with localized pain and fracture.[8] Histological changes were comprehensively described by Gorham et. Al. In 1954.[9] They described the histological changes in the affected bones as extensive proliferation of endothelial-lined channels accompanied by osteolysis.[9]

The etiology of skeletal angiomatosis remains obscure. It is now thought that the vascular endothelial growth factor (VEGF) group and cytokine IL-6 are the principal molecules responsible for distruction of bone. [10-11] VEGF-C which acts as ligand of the tyrosine kinases receptor VEGFR2 and VEGFR3, is beleived to be the most critical lymphangectatic stimulus leading to proliferation and growth of vessels [12,13].

Treatment of Skeletal angiomatosis is not well established. Once the disease stabilizes surgery can be performed [14]. Radiotherapy can be used alone or it can be used along with surgery [15-17]. Success rate of radiotherapy is about 75% if only local lesions is present [15]. The most commonly prescribed pharmacological agent to treat skeletal angiomatosis are interferon and alpha bisphosphonates[18--20]. Differential diagnosis of cystic angiomatosis presenting as multiple lytic bone lesions Include; Multiple myeloma, Bone Metastasis, Fibrous dysplasia, Lymphoma, Hyperthyroidism, Hystiocytosis X, Gaucher's disease, Amyloidosis Gorham disease and Enchondromatosis. In this case the clinical course, the physical examination, the laboratory findings and the histopathological examination lead to exclusion of all diffrential diagnosis but exclusion of histiocytosis X is always difficult. In Histiocytosis X, the skull is involved more frequently than other sites. Moreover the "punched out lesions" do not usually have a peripheral ring of increased density as well as they tend to coalesce to produce map-like areas of bone destruction. In contrast with cystic angiomatosis, lesions near the cortex of long bones in cases of Histiocytosis X stimulate a periosteal reaction.

Clinician, radiologist and pathologist should be aware of this rare entity and despite the rarity of the disease; skeletal angiomatosis should always be considered as a differential diagnosis in a patient presenting with skeletal lytic lesions and minimal or no associated laboratory findings.

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