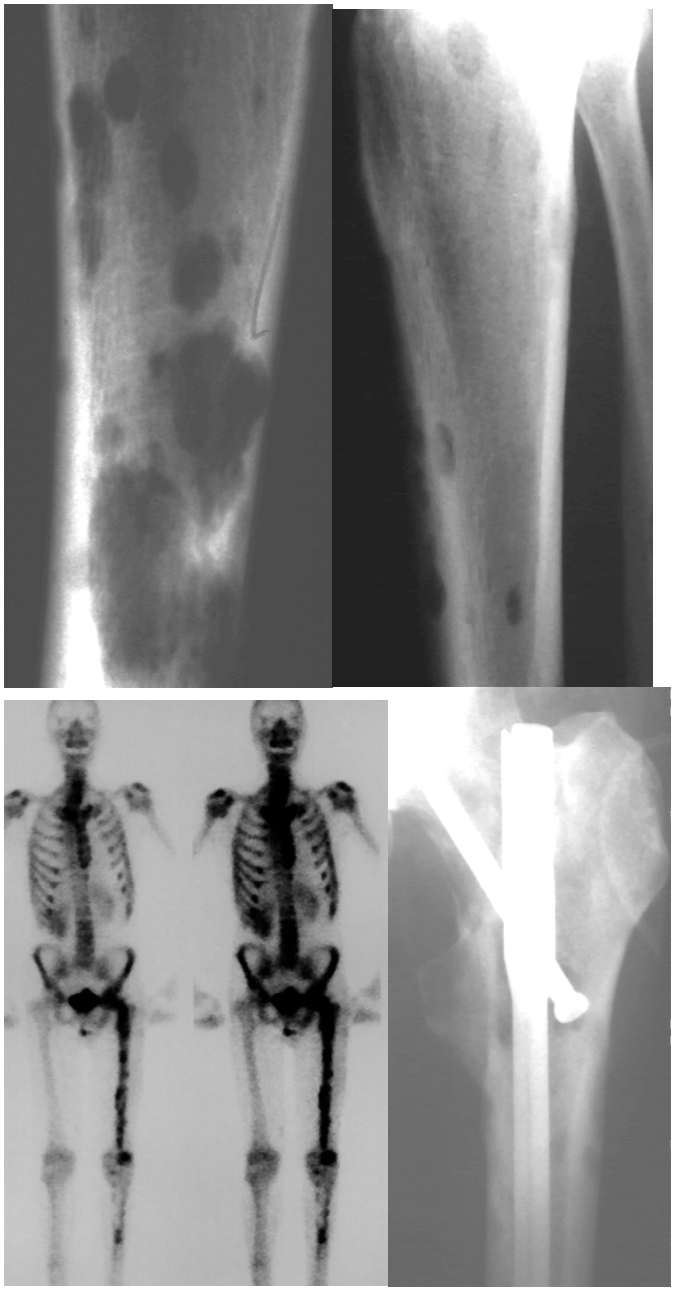


Case 6: A 64-year-old man was referred for a pain in the left groin and left thigh; insidious; worse in last 3 months. The pain radiated to the left knee, became worse after long periods of walking or standing, and was not relieved with common analgesics.

He had deep tenderness in the left groin and over the greater trochanter. There was no swelling, deformity, varicosity, or redness. The range of motion of the left hip was limited/

The ESR was elevated. Plain radiographs showed multiple osteolytic lesion on one side of the body [left]. **? Diagnosis**

Diagnosis: Angiosarcoma

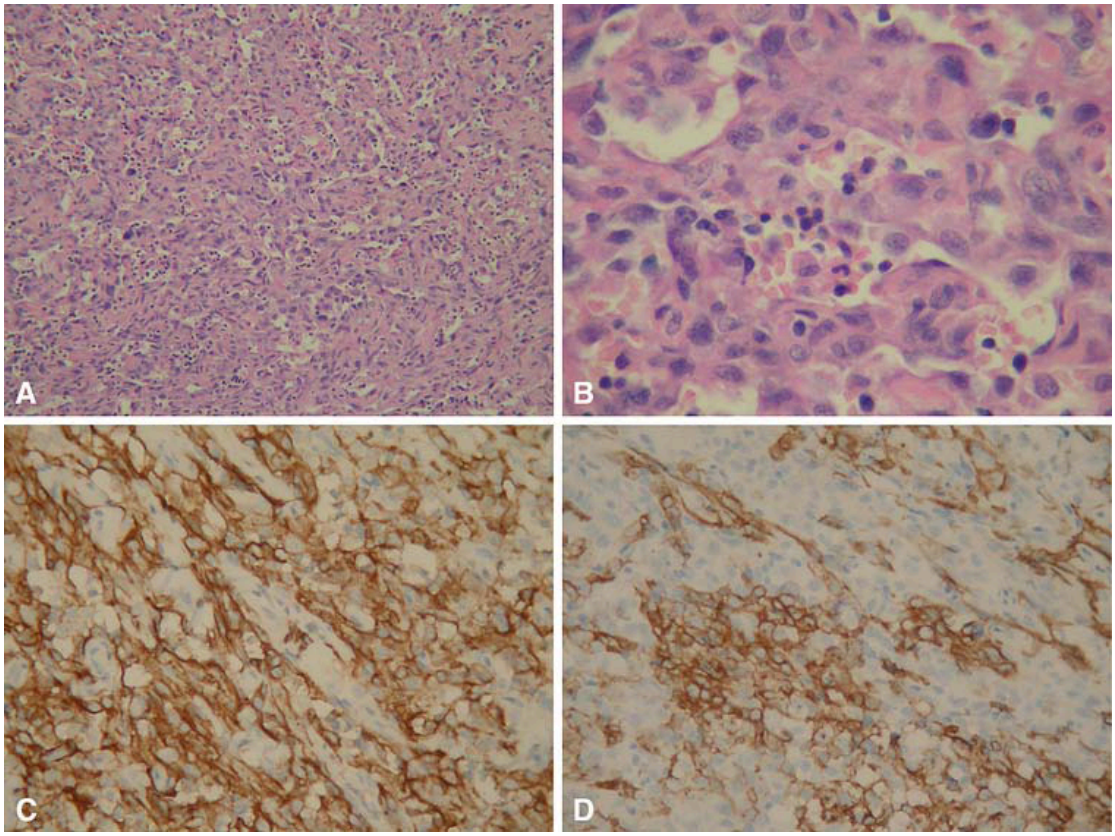
X rays
showed irregularly expansive, irregular lytic bone destruction with no surrounding sclerosis or any periosteal reaction.

Bone Scan
Multiple lesions of increased uptake was localized in the left side of the body.

Differential Diagnosis
Multiple myeloma
Metastasis
Fibrous dysplasia
Paget's disease
Multicentric osteolytic osteosarcomaHistology

Pathology
Lesion consisted of grayish-white, soft, rubbery, hypervascular soft tissue
Necrotic and cystic areas were interspersed in the lesion.
Tissue were stained with hematoxylin and eosin and antibodies specific for vimentin, CD31, CD34, CD68, and epithelial membrane antigen. Hematoxylin and eosin staining of the tumor showed a large amount of necrotic tissue, fibrous hyperplasia and significant vessel structures also were seen. The tumor consisted of solid nests or large sheets of spindle or epithelioid cells interspersed between and around preexisting vessels with flat endothelium.
Mitotic figures were numerous and frequently abnormal.

(A) Solid nests or large sheets of spindle or epithelioid cells are interspersed between and around preexisting vessels with flat endothelium (Stain hematoxylin and eosin)
(B) Tumor cells are large and pleomorphic and show a moderately abundant eosinophilic cytoplasm and a round-to-oval nucleus with one or two prominent nucleoli. Mitotic figures were numerous.
(C) Immunohistochemical analysis revealed strong staining of tumor cells for CD31
(D) Immunostaining for CD34 was partially positive



Discussion

Fibrous dysplasia of bone starts in early childhood but is usually mild and asymptomatic, often being discovered at the onset of symptoms by pathologic fracture. Sarcomatous degeneration of fibrous dysplasia is rare .

Malignant degeneration in Pagetic bone is a well-recognized but rare complication. Imaging typically shows an aggressive bone-changes such as deformed widened bone with coarse trabeculae. The radiographs showed areas of aggressive biologic behavior as there was no sclerosis at the lesion-bone junction [transition].

For osteosarcoma, its peak incidence is the second decade of life and it rarely is seen in patients older than 50 years. Its clinical and radiographic presentations vary in different types. Typically, periosteal reaction such as the sunburst phenomena or Codman's triangle is common in osteosarcoma; however, our patient lacked these features.

Angiosarcoma is a malignant neoplasm of mesenchymal cell origin leading to formation of blood vessels. Angiosarcoma of bone also is a rare vasoformative tumor and occurs exclusively in adults.

These tumors usually occur in long tubular bones and much less frequently in the ribs, pelvis, and vertebrae and seen in multiple bones and seen on one side of the body.
The lesion is solitary or multiple greater than 5 cm. Diagnosis is particularly difficult as vascular tumors of the bone often show heterogeneous differentiation .

Multiple lesions can develop in a single bone or involve multiple bones with lesions randomly distributed throughout the skeleton or clustered in an anatomic region, such as a single extremity. The presence of multicentric lesions may be the only clue that suggests the diagnosis of a vascular tumor, whereas the solitary lesion might have numerous differential diagnoses.

When an angiosarcoma of bone is identified, a skeletal survey is recommended to evaluate whether the patient has multicentric disease or to show areas of increased activity.

MRI changes are not specific. The diagnosis is established by the characteristic histologic features. Histologically, it is composed mainly of a mass of anastomosing vascular channels. Its diagnostic feature is always the formation of new blood vessels, and typical endothelial cells should be identified
The absence of cytokeratin reactivity and strong reactions to antibodies for vimentin help to distinguish these tumors from spindle cell type carcinomas .
Antibodies currently used are Factor VIII-related antigen and CD31, which are specific markers for endothelial cells . CD31 is considered the most sensitive and specific routine marker for all types of Angiosarcoma.

An angiosarcoma is an extremely aggressive malignancy that often has spread hematogenously before it is .recognized and exhibits variable malignant behavior.
Course is always is characterized by the rapid onset of symptoms and high frequency of local recurrence or metastasis. The most effective treatment is complete surgical removal of the tumor, however obtaining adequate surgical margins often is difficult.

The role of adjuvant treatment is not well defined. Radiotherapy has been given as an adjunct to surgical therapy or as palliative surgical stabilization in numerous cases of angiosarcoma.

By 3 months postoperatively the tumor had metastasized to the lung and he died 4 months after surgery.