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Medical Imaging Quiz – Case 69

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A 55-year-old male patient was referred to the Interventional Radiology Department for a computed tomography guided (CT) percutaneous biopsy of a large mass of the right thigh. He mentioned a painless mass for at least 6 months. A magnetic resonance imaging (MRI) was performed and revealed a large well-circumscribed mass in the medial compartment of the right thigh (fig. 1). CT guided biopsy was performed and confirmed the diagnosis.

Comment

Soft tissue sarcomas are a heterogeneous group of malignant tumors of mesenchymal origin that originate mainly from soft tissues rather than bone. They are classified on the basis of tissue seen on histology. The commoner sarcomas in the adult are undifferentiated pleomorphic sarcoma, liposarcoma, rhabdomyosarcoma, fibrosarcoma, dermatofibrosarcoma, leiomyosarcoma, hemangiosarcoma, Kaposi sarcoma, lymphangiosarcoma, synovial sarcoma, malignant peripheral nerve sheath tumor, extraskeletal chondrosarcoma, extraskeletal osteosarcoma, alveolar soft-part sarcoma.

Undifferentiated pleomorphic sarcoma (UPS), previously known as malignant fibrous histiocytoma (MFH), is considered the most common type of soft tissue sarcoma (25–40% of all adult soft tissue sarcomas). It has an aggressive biological behavior and a poor prognosis. They frequently metastasize and locally recur despite aggressive treatment. Typically UPS affects the extremities and occurs in adults (range 32–80; mean 59 years) with a slight male predilection with an M:F ratio of 1.2:1. Usual presentation is a painless, enlarging palpable mass.

They are the most frequent soft tissue sarcoma to occur as a result of radiotherapy and are also seen on a background of Paget disease. Secondary transformation into the malignant sarcomas like UPS, have also been reported in fibrous dysplasia, giant cell tumor, enchondroma, chronic osteomyelitis, and osteonecrosis.

They are usually confined to the soft tissues but occasionally may arise in or from bone (1–5%). Although UPS can occur almost anywhere in the body, they have a predilection for extremities: Lower and upper extremities corresponding to approximately 50%

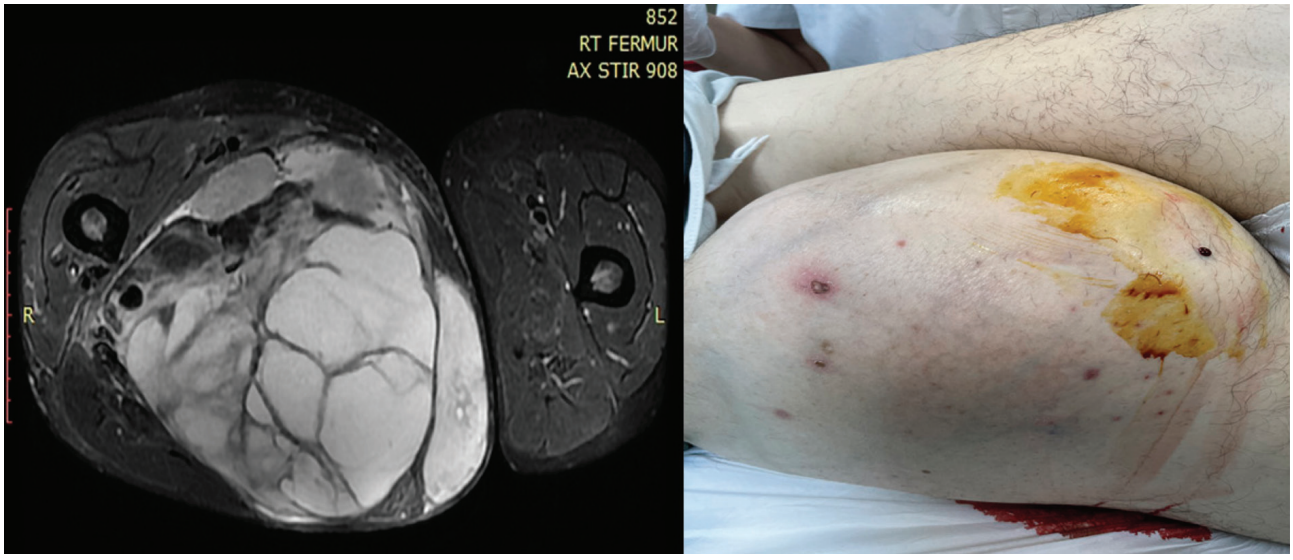

Figure 1. Large well-circumscribed mass in the medial compartment of the left thigh is present with high T2 signal and low T1 signal. It does not invade bone or subcutaneous fat.

and approximately 20% of the cases, respectively. Retroperitoneal UPS represent about 16% of the cases and, those involving the peritoneal cavity, about 5–10%. Involvement of the abdominal viscera is described as extremely rare.

Macroscopically, these tumors are typically large (6–20 cm) well circumscribed but unencapsulated with a firm heterogeneous cut surface with areas of necrosis. Microscopically they are heterogeneous fibroblastic tumors made up of poorly differentiated fibroblasts, myofibroblasts, histiocyte-like cells with significant cellular pleomorphism, storiform architecture and also demonstrate bizarre multi-nucleated giant cells. They are sometimes difficult to distinguish from other high-grade sarcomas. Radiological evaluation of UPS occurs with plain radiographs, CT and MRI. Plain radiographs demonstrate a soft tissue mass and if arising from bone, then appear as an aggressive destructive bony lesion. In some cases regions of calcification may be demonstrated. CT reveals similar density to adjacent muscle, with heterogeneous lower density areas if hemorrhage, necrosis or myxoid material is abundant. The soft tissue component enhances. In up to 15–20% of cases, some mineralization is present. MRI is the modality of choice for assessing soft tissue sarcomas, as it is best able to locally stage a tumor. These tumors are typically relatively well-circumscribed, located within or adjacent to muscle, exerting a positive mass effect on surrounding structures due to their large size at presentation. Reveals T1 intermediate (to low) signal intensity similar to adjacent muscle, T2 intermediate to high signal intensity and heterogeneity if hemorrhage, calcification, necrosis or myxoid material present.

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