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TEST YOURSELF: ANSWER

Heavily calcified synovial sarcoma leading to chronic thigh pain and swelling

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This is a case of heavily calcified synovial sarcoma leading to chronic thigh pain and swelling.

The pelvic radiograph shows a cortical-based, lobulated lesion at the medial aspect of the proximal right femoral shaft without obvious extension into the medullary cavity (Fig. 1). MRI shows a lesion along the anteromedial aspect of the right thigh. Nearly the entire lesion shows large areas of T1 and T2 hypointensity suggestive of calcification/ossification (Fig. 2). Post-contrast study shows patchy enhancement in between the hypointense portions (Fig. 3). On CT, there is continuity with the medial femoral cortex, without evident medullary continuity (Fig. 4). Equivocal imaging features prompted a Jamshidi needle biopsy predominantly from the enhancing component of the lesion. Tissue sections demonstrated bony trabeculae along with areas of calcification and spindle cell proliferation with elongated nuclei and a moderate amount of cytoplasm, leading to a provisional diagnosis of myositis ossificans. Since the patient was symptomatic, a decision was taken for wide local excision of the lesion with adjacent marginal femoral resection. The final histopathological specimen showed scanty viable tumor cells composed of uniform round cells having mild pleomorphism and arranged in sheets with large areas of dense calcifications and sclerosis (Fig. 5). Tumor cells show diffuse positivity for MiC-2 (CD99), Bcl-2 protein, and TLE-1 and showed focal positivity for EMA (Fig. 6). This combination of IHC markers is quite specific for synovial sarcoma and it rules out MO as well as osteogenic sarcoma. Molecular confirmation with SS18-SSX1/2 is ideal, however could not be done due to the very scanty volume of viable tumor cells. These findings lead to the diagnosis of poorly differentiated heavily calcified synovial sarcoma.

Synovial sarcomas are one of the most common soft tissue tumors occurring in adults described in the 5th edition of the WHO classification of soft tissue tumors as a category of "tumors of uncertain differentiation." Conventionally four histological subtypes biphasic, monophasic fibrous, monophasic epithelial, and poorly differentiated are described [1]. Radiological detection of stippled calcifications is a commonly known feature but the presence of marked chunks of calcifications involving almost the entire lesion is rare [2]. This case is a prime example of such a lesion. The lack of characteristic clinical manifestations often leads to the reliance on imaging to pave a path towards a probable diagnosis [3]. The imaging findings though equivocal can be useful in ruling out certain pathologies and paving a path towards diagnosis. The absence of a history of trauma and lack of medullary continuity make the diagnosis of myositis ossifications and osteochondroma remote [4].

Tumoral calcinosis on CT images has the classical description of a cystic lesion showing the so-called sedimentation sign, occurring due to fluid-fluid levels caused by calcium layering. On MR imaging, the appearance is variable seen either as a near homogeneous diffuse low signal intensity pattern or a nodular pattern with alternating areas of signal void and high signal intensity [5]. Periosteal osteosarcoma is very difficult to differentiate from a heavily calcified synovial sarcoma [6]. They both present with unmineralized/non-calcified soft tissue components and a

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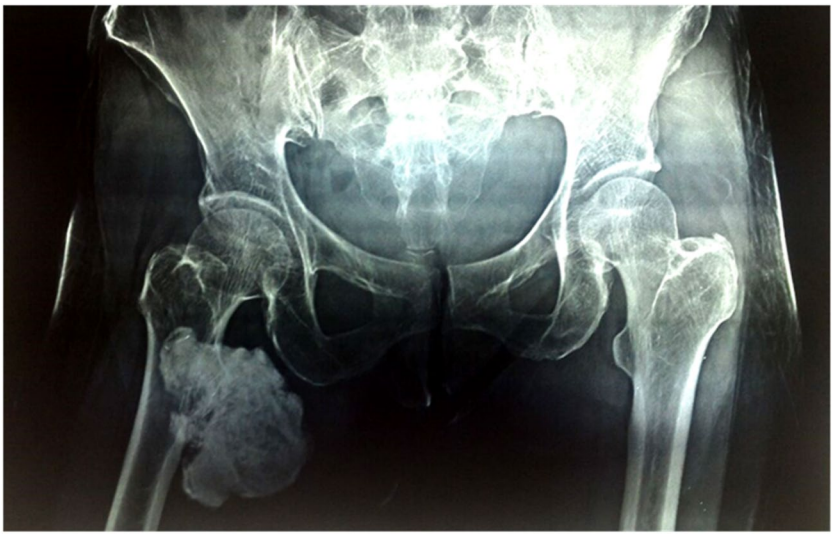

Fig. 1 The pelvic radiograph shows a cortical-based, lobulated lesion at the medial aspect of the proximal right femoral shaft without obvious extension into medullary cavity

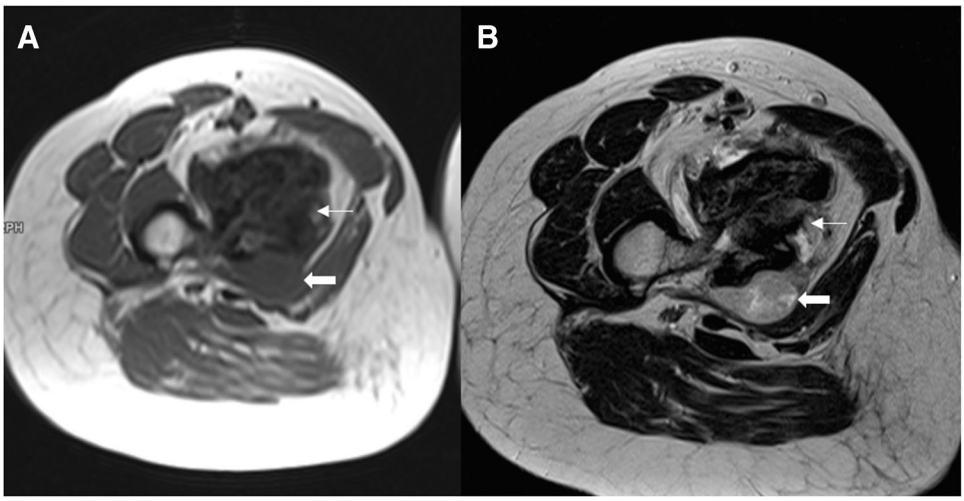

Fig. 2 **A** (Axial T1) and **B** (axial T2): nearly the entire lesion shows large areas of T1 and T2 hypointensity suggestive of calcification/ossification (thin arrows). Unmineralized T1 hypointense and T2 hyperintense components seen in the posteromedial aspect of the lesion (thick arrows)

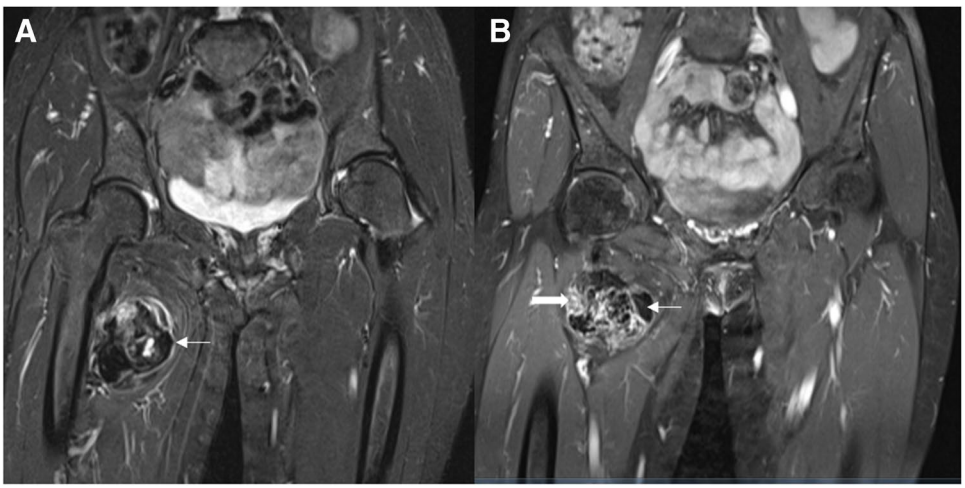

Fig. 3 **A** (Coronal T2 with fat suppression) and **B** (coronal post-contrast with fat suppression): the mineralized component does not show any post-contrast enhancement (thin arrows). Patchy enhancement is seen in the soft tissue component (thick arrow)

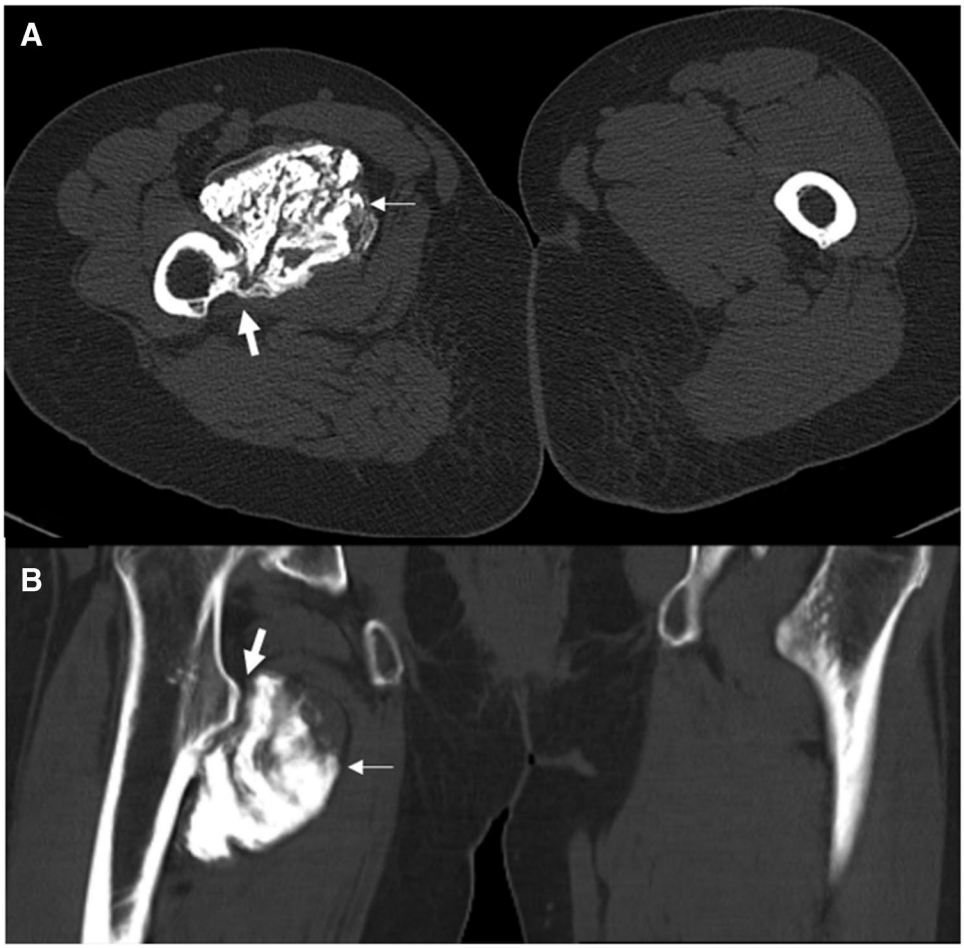

Fig. 4 **A** (Axial CT image in bone window) and **B** (coronal CT image in bone window): on CT, the lesion (thin arrows) shows continuity with the medial femoral cortex (thick arrows), without evident medullary continuity

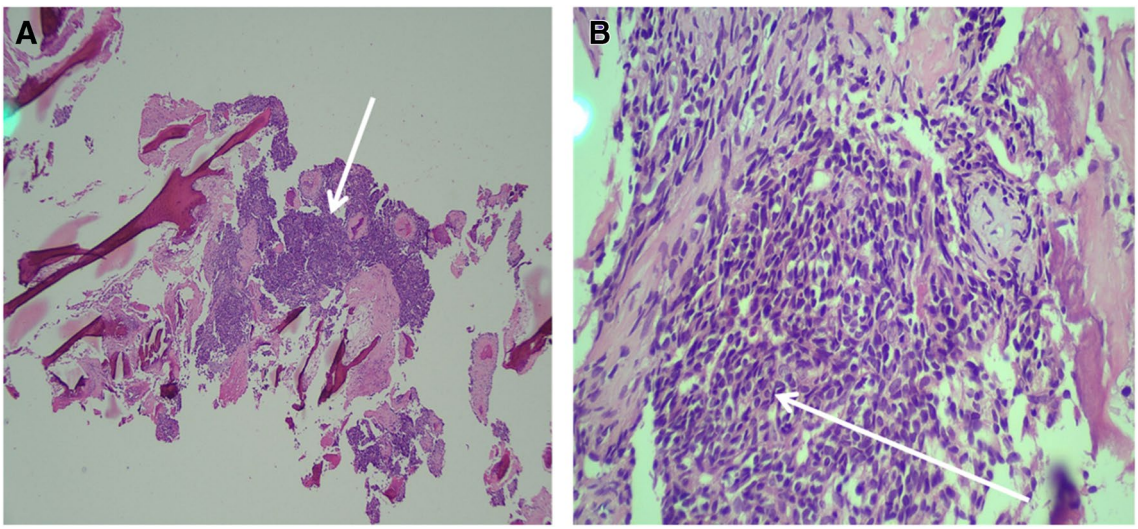

Fig. 5 **A** (HE 4X) and **B** (20X): solid nests of blue-appearing tumor cells along with irregular bone formation (white arrow in **A**). Tumor cells are round and oval with slight spindling and have scant cytoplasm. Irregular foci of calcifications are present (white arrow in **B**)

synovial sarcoma close to a bone can lead to a periosteal reaction that can mimic periosteal osteogenic sarcoma [7]. In such cases, a biopsy would be a prudent step. Myositis ossificans progressiva is an autosomal dominant condition where ossification of the muscles, tendons, and ligaments occurs. It is caused by a mutation in the ACVR1 gene. Thus,

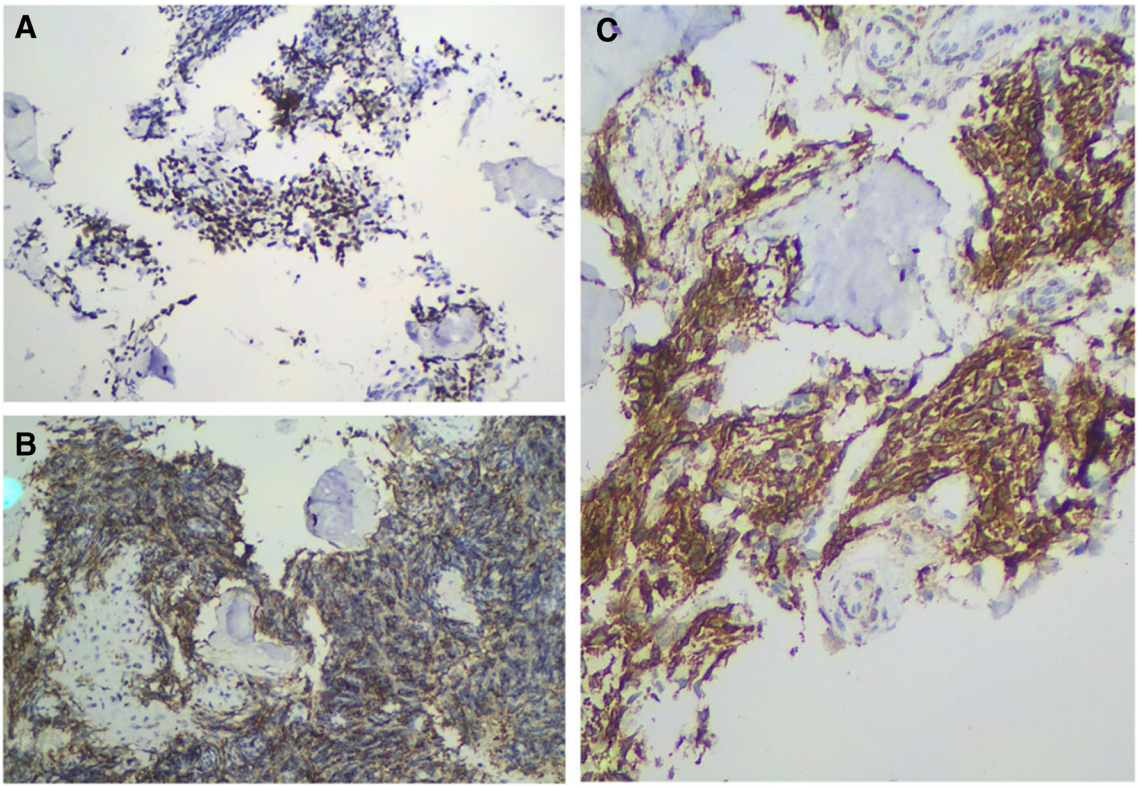

Fig. 6 Immunohistochemistry (IHC) staining pattern TLE 1 (**A**), MiC 2 (**B**), and Bcl-2Protein (**C**) consistent with diagnosis of a synovial sarcoma

if a seemingly benign lesion is encountered with chunks of calcifications and unmineralized soft tissue components, it is important to keep heavily calcified synovial sarcoma in the differentials before labelling it as a benign entity. This case highlights the importance of considering synovial sarcoma as a radiological possibility even if the calcifications are extensive and highlighted.

Declarations

Conflict of interest The authors declare no competing interests.

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