Soft-Tissue Mass Lesion of the Foot - Synovial Sarcoma

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Section 2 – Answer

Case

A 46-year-old male presented to the orthopedic outpatient clinic with a history of gradually progressive swelling and pain in the left foot for 2 years. The patient denied any history of trauma and any significant past medical or surgical history. On examination, the swelling was seen in the left foot involving the dorsal and medial aspect with tenderness on palpation. The overlying skin was stretched and showed hyperpigmentation with minimal scales and central depigmentation [Figure 1].

Radiograph and ultrasound images of the foot are shown in Figures 2 and 3. An ultrasound video clip is also shown (video file :1). What is your impression?

Interpretation

Conventional radiograph of the foot showed soft-tissue swelling involving the first intermetatarsal space with scalloping of the first and second metatarsals. There was no sclerosis or lysis of adjacent bone, and there was no calcification seen [Figure 2].

High-frequency ultrasound of the foot showed heterogeneously hypoechoic predominantly solid lobulated mass lesion measuring approximately 6 cm × 4 cm with ill-defined margins in the first and second intermetatarsal spaces with high vascularity on Power Doppler [Figure 3 and Video File 1]. A provisional diagnosis of soft-tissue sarcoma of the left foot was made, and for further confirmation of the diagnosis and to see the extent of mass, magnetic resonance imaging (MRI) of the foot was performed.

MRI of the left foot showed large lobulated solid cystic mass lesion measuring approximately 6 cm × 4 cm × 2 cm in the first and second intermetatarsal spaces. The lesion showed mixed signal intensity appearing predominantly hypointense on T1-weighted sequence and hyperintense on T2-weighted sequence. The cystic component of the lesion showed fluid-fluid level within it. The lesion was encasing the second metatarsal bone with infiltration; and, the lesion showed significant postcontrast enhancement [Figure 4].

The patient underwent an excisional biopsy, which showed features of biphasic synovial sarcoma [Figure 5]. The metastatic workup of the patient was negative. The patient is kept on neoadjuvant chemotherapy, on the completion of which a radical surgical excision is planned.

Discussion

Synovial sarcoma arises from mesenchymal tissue and then undergoes differentiation to exhibit the histological appearance of the synovium. It is the third most common soft-tissue sarcoma in adults constituting approximately 10% of all soft-tissue sarcomas.[1] Men and women are equally affected. It is seen in adolescents and adults with ages ranging from 15 to 40 years. Three main histological subtypes of synovial sarcoma have been identified: biphasic, monophasic, and poorly differentiated.[2] Monophasic synovial sarcoma represents 50%–60% of cases (the most common subtype), and biphasic sarcoma constitutes about 20%–30% of cases. Translocation (X;18) is highly specific and is seen in approximately 90% of the cases.[3]

Most of the patients present with deep-seated swelling surrounding a large joint, most commonly the knee. Mostly, tumors are larger than 5 cm at the time of presentation. Some patients may present with pain, tenderness, and restricted movement of the adjacent joint. The lesions are most commonly seen in the extremities (lower >upper), in periarticular locations close to a bursa or tendon sheath instead of involving the joint

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proper. Approximately 18% of all synovial sarcomas occur in the foot and ankle.[3] Very rarely, the lesions are seen in an intra-articular location (<10%).[4]

Initial radiographs will be normal in the majority of cases. Larger lesions may appear as nonspecific round or oval soft-tissue masses.[3] Calcification can be identified in up to 30% of synovial sarcomas.[5,6] The calcifications are often nonspecific and located in the periphery. Chondroid and osteoid mineralization has been reported in few cases.[7] Extensive calcification is associated with better prognosis.[7,8]

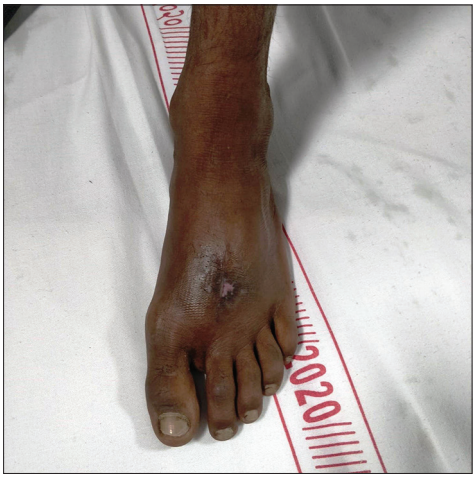

Figure 1: Photograph of the left foot showing swelling in dorsum with hyperpigmentation of the overlying skin with minimal scales and central depigmentation

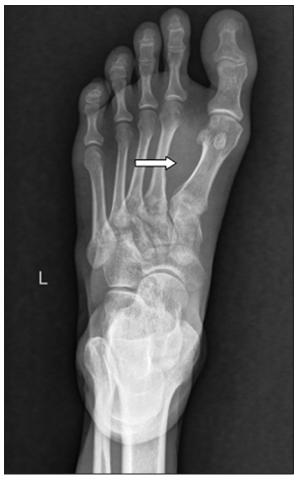

Figure 2: Radiograph of the left foot in dorsoplantar view showing soft-tissue swelling (white arrow) in the first metatarsal space with scalloping of the first and second metatarsals

Only one study[9] has been published so far on the imaging features of synovial sarcoma on ultrasonography. According to that study done by Marzano *et al*.[9] on 35 cases of synovial sarcoma, 66% of cases revealed a mass lesion with well-defined rounded and lobulated margins suggestive of less aggressive process, 14% of cases revealed prominent heterogeneity with irregular margins, and 20% of cases revealed a complex sonographic appearance. The Doppler study almost always revealed high vascularity.

MRI is the imaging study of choice for evaluating tumor size, texture, and local extent. Synovial sarcoma generally appears as a multilobulated mass lesion with a "bowl-of-grapes" appearance; with areas of hyper, iso, and hypointensity on T2-weighted sequence known as triple signal intensity and fluid-fluid levels with intervening septa.[10] Triple sign on T2-weighted MRI is due to the mixture of solid cellular elements (intermediate signal intensity), hemorrhage or necrosis (high signal intensity), and calcified or fibrotic collagenized regions (low-signal-intensity).[3]

Differential Diagnosis

Clinical and radiological differentials include both benign and malignant entities. Benign entities include hemangiomas, neurofibroma, tenosynovial giant cell tumor, whereas malignant entities include malignant fibrous histiocytoma, epithelioid sarcoma, and liposarcoma.

Hemangioma appears as multiple high signal intensity lobules on T2-weighted images due to cavernous or cystic vascular spaces containing stagnant blood. Punctate or reticular low-signal-intensity areas may be present, representing fibrous tissue, fast flow within vessels, or foci of calcification.[11] Lack of vascular channels and phleboliths ruled out hemangioma in our case.

Neurofibromas are characterized by fusiform enlargement of the nerve, with the tapered ends of the lesion toward the parent nerve. Neurofibromas show few characteristic signs on imaging such as target sign, fascicular sign and split fat sign, and thin

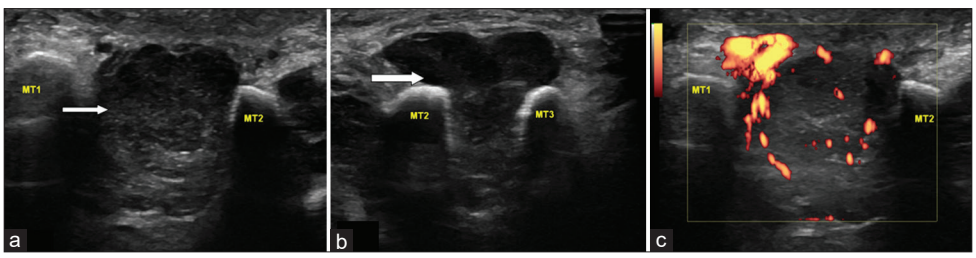

Figure 3: Ultrasound of the foot using a high-frequency probe in transverse plane showing a heterogeneously hypoechoic predominantly solid lobulated mass lesion in the first (arrow in a) and second (arrow in b) intermetatarsal space. Power Doppler images of the lesion showed high vascularity (c). MT1: First metatarsal, MT2: Second metatarsal, MT3: Third metatarsal

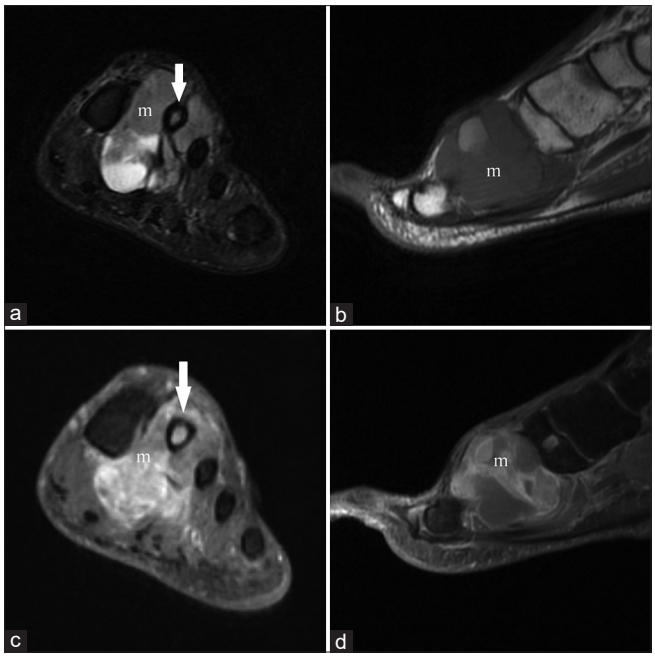

Figure 4: Magnetic resonance imaging of the foot showing heterogeneous mass lesion (m) giving mixed signal intensity (triple sign) on T2-weighted sequence (a), hypointense signal intensity on T1-weighted sequence (b), and heterogeneous postcontrast enhancement (c and d). The second metatarsal (solid arrow) is infiltrated by the mass

T2-weighted hyperintense rim. No continuity with any nerve and absence of any of the abovementioned signs ruled out the diagnosis of neurofibroma.

Giant cell tumor of tendon sheath presents as a well-defined mass eccentrically located in association with or partially/completely enveloping a tendon. They characteristically exhibit a low signal on T1-weighted images and T2-weighted images due to the presence of hemosiderin and exhibit homogeneous enhancement on postcontrast scan.[12] The absence of low signal on T2-weighted sequence pointed against the diagnosis of tenosynovial giant cell tumor in this case.

Among malignant tumor, liposarcoma can be a differential; however, the absence of fat ruled out liposarcoma in our case.

Radical surgical excision of the tumor is the treatment of choice. It has a good prognosis in the absence of metastasis.[1] Ultrasonography is a very useful modality for the evaluation of mass lesions located in periarticular locations. It can help in seeing the extent of the disease and guide biopsy from the solid component for better yield.[9]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

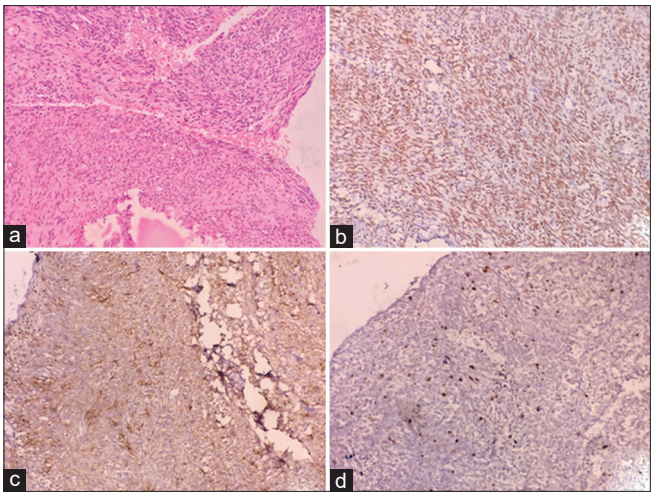

Figure 5: H and E images at ×10 (a) shows infiltrating tumor with predominantly spindle cells and round to oval cells (biphasic pattern). Immunohistochemical image showing TLE nuclear positivity (b), epithelial membrane antigen focal positivity in round-to-oval cells (c), and Ki67 of 2% positivity (d)

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Conflicts of interest

There are no conflicts of interest.

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