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Case Report

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A Case of Unusual Swelling in a Young Child: Myelosarcoma

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Introduction

A previously healthy, two-year old male presented to the Emergency Department for the evaluation of a painless left thigh mass noticed earlier that day by family members. Review of systems was negative for fever, trauma, and recent illness. Per father, the patient "was acting like himself, running and playing." The patient was alert and well-appearing with vital signs as follows: heart rate 94/min, blood pressure 95/44 mmHG, temperature 36.7 °C, and respiratory rate 22 breaths per minute. Examination of the left lower extremity revealed an ill-defined, non-tender, firm, immobile swelling measuring around 7 x 4 cm in the left anterolateral thigh region. There were no cutaneous signs of inflammation or trauma. Range of motion and gait testing was normal. Rest of the examination was unremarkable.

Initial diagnostic work up including X-ray, complete blood count, complete metabolic panel, and C-reactive protein was normal. Lactate dehydrogenase was mildly elevated at 733 U/L. Further work up consisted of an ultrasound and magnetic resonance imaging (MRI of the left lower extremity. Ultrasound showed a 7 x 3 x 4 cm hyperechoic focus in the left thigh region suggestive of focal myositis versus hematoma (Figure 1). MRI of the left femur revealed a large anterior infiltrating intramuscular mass measuring 6.7 cm in greatest diameter, concerning for an aggressive soft tissue growth (Figure 2). Incisional biopsy and flow cytometry analysis of the left upper thigh mass demonstrated abnormal myeloid population with CD56 positivity, negative for CD4 and 123, thus enabling the diagnosis of myeloid sarcoma. Bone marrow biopsy revealed no involvement or suspicion for leukemia. A whole-body Positron Emission Tomography (PET) scan did not reveal any other lesions. As recommended, patient was started on systemic chemotherapy per standard AML protocol. He completed induction chemotherapy with four rounds of cytarabine, and consolidation II therapy with nine rounds of pegasparagase. He responded well and his post treatment MRI showed complete resolution of the mass.

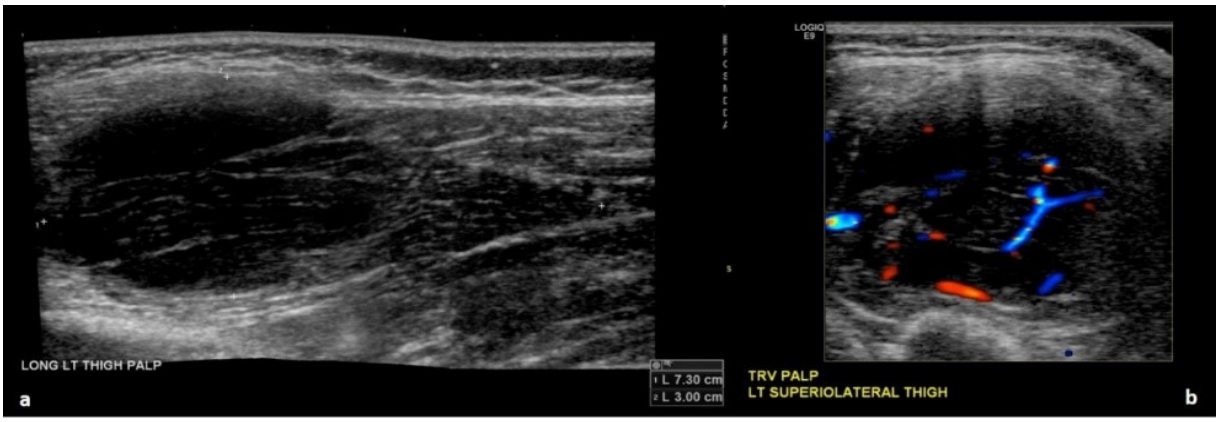

Figure 1: Ultrasound (a,b) showed a 7 x 3 x 4 cm hyperechoic focus (denoted by white arrows) along the lateral extensor muscle of left thigh suggestive of either a hematoma, myositis, or sarcoma.

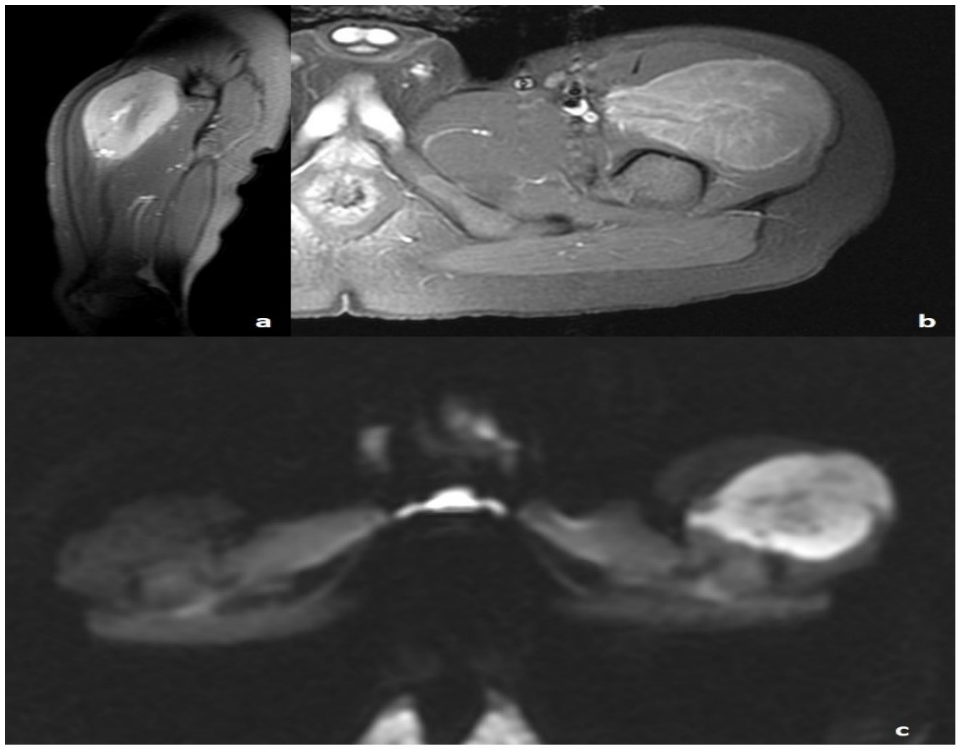

Figure 2: MRI of the left femur (a,b,c) revealed a large anterior hyperintense infiltrating intramuscular mass (denoted by white arrow) with enhancement and diffusion restriction, measuring 6.7 cm in greatest diameter, without evidence for osseous involvement.

Discussion

Myeloid Sarcoma

Myeloid sarcoma (also known as granulocytic sarcoma, myeloblastoma, or chloroma) is a rare form of extramedullary Acute Myeloid Leukemia (AML) presenting without bone marrow involvement. According to the World Health Organization, Myeloid Sarcoma (MS) is classified as a subcategory of AML [1]. It can present by itself months or years before the diagnosis of AML. Otherwise, it may appear concomitantly or with AML relapse. It is termed isolated or primary MS, which occurs in the absence of a history of leukemia, myelodysplastic syndrome or myeloproliferative neoplasm. The exact pathogenesis is unknown. Extramedullary presentation suggests there might be an aberrant homing signal for the leukemic blasts precluding the more common bone marrow localization [2]. In MS there is rapid extramedullary proliferation of immune cells of myeloid lineage. Onset can be acute or subacute and is location dependent. MS can occur at any body site, including central nervous system, bone, skin, lymphoid tissue, GI tract, and orbits. In children, MS most frequently occurs in the skin, gingival tissue and orbits [3]. According to Dores, et al. [4], less than one percent of AML cases present as extramedullary myeloid sarcoma. Age of diagnosis may be highly variable, with cases being reported in patients 1-81 years old; a majority of MS cases occur in individuals 55 years and older.

Diagnosis

Pediatric soft tissue masses have varied underlying etiology and presentations. Though mostly benign (e.g. lipoma, hemangioma, hematoma), they can also be malignant (e.g. liposarcoma, malignant melanoma) in nature. They can occur in patients who appear well and lack constitutional symptoms, delaying the correct diagnosis or causing it to be missed altogether. A systematic approach is key towards making the right diagnosis which includes thorough history and physical examination. Information regarding onset, duration, and progression of the swelling; presence of associated symptoms; and any history of trauma should be specially sought. Physical examination findings like size, location, consistency, mobility and any signs of inflammation or trauma can help characterize the mass and narrow the differential diagnosis.

Owing to diverse clinical presentation, and absence of hematological findings, isolated or primary MS remains a diagnostic challenge. There is often a delay in diagnosis as MS is initially misdiagnosed as lymphoma, undifferentiated cancer, malignant melanoma, extramedullary hematopoiesis and inflammation [2]. A presumptive diagnosis of MS may be made via the presence of leukemic blasts in a peripheral blood smear. Definitive diagnosis requires immunohistochemistry and immunophenotyping. Diagnostic work-up of MS should include tissue biopsy, immunohistochemical staining and flow cytometry, which will reveal elevated levels of myeloid cell markers [5]. Cytogenetics of the tissue biopsy must be performed, as MS has been associated with multiple chromosomal abnormalities [6], Imaging is an important diagnostic and prognostic modality. It is essential for the evaluation of tumor size and localization, which can be either solitary or multifocal. Computerized Tomography (CT) and MRI can help differentiate MS from other lesions (e.g. hematoma, hamartoma or abscess, etc.). PET scan should be considered to determine if there are multiple lesions. PET-CT is also beneficial, especially for monitoring response and planning radiation therapy (3).

Prognosis and Outcomes

Due to the rarity of MS, the prognosis and outcome remain unclear. Extramedullary MS has a diverse anatomic clinical presentation and the outcomes may vary significantly according to age, sex, race and sites of presentation [7]. The patients with isolated MS may have a better prognosis when compared with AML patients without MS. Systemic chemotherapy is associated with improved survival outcomes. Relapses with isolated MS are very rare and systemic relapse almost always follows after a median time of approximately 7 months [8].

Treatment

Due to an elevated rate of progression from isolated MS to AML in patients receiving localized treatment, systemic chemotherapy based on conventional AML protocols is the recommended treatment regimen. Local surgery or radiotherapy has an adjunctive role so as to help delay the transformation to AML and improve the outcome. In our case, chemotherapy alone was successful in complete resolution of the mass. At 1 year follow up our patient was completely asymptomatic. Post therapy MRI and PET scans have remained normal with no evidence of residual or recurrent tumor.

Learning Points

- Pediatric soft tissue masses are mostly benign or reactive but rarely can be a manifestation of a hematological malignancy like MS.
- MS, a form of extramedullary AML, is a rare entity that can develop at any site in the body and have myriad clinical presentations.
- Timely initiation of systemic chemotherapy is essential for an optimal outcome in patients with MS.

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