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RESEARCH ARTICLE

SOFT TISSUE MASS AND ELECTRICAL DISCHARGE: WHAT'S INVOLVED?

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Abstract

An 11-year-old girl presented with a gradually evolving, painful arm mass over 6 months. Clinical examination revealed a 4cm soft mass causing electric discharge pain on percussion, without signs of inflammation. Ultrasound depicted a well-defined, heterogeneous mass connected to the median nerve with moderate vascularization. Subsequent MRI showed an eccentric fusiform mass within the median nerve, exhibiting a muscle-like signal on T1-weighted images, a heterogeneous signal on STIR, and enhancement after contrast. No bony destruction or muscle invasion was observed. Peripheral nerve sheath tumors, including neurofibromas and schwannomas, were suspected. MRI played a crucial role, revealing characteristic signs like split fat, target, and fascicular signs, aiding in distinguishing between benign and malignant lesions. Intra-tumoral changes, like cystic degeneration seen in schwannomas, were noted. Malignant tumors, such as MPNSTs, showed distinct features like increased size, heterogeneous appearance, perilesional edema, and bony destruction. In summary, the patient's arm mass, initially detected through ultrasound and further characterized by MRI, raised concern for peripheral nerve sheath tumors, highlighting the significance of imaging in diagnosing and distinguishing these lesions.

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Introduction:-

Peripheral nerve sheath tumors account for nearly 12% of the benign and 7-8% of the malignant soft tissue neoplasms. Neurofibromas and schwannomas constitute the benign category, while the malignant peripheral nerve sheath tumor (MPNST) comes under the malignant category. Magnetic resonance imaging (MRI) plays a pivotal role in the identification, characterization, and differentiation of benign versus malignant nerve sheath tumors. This article reports a case of a soft tissue mass with electrical discharge revealing a median nerve schwannoma.

Clinical presentation:

An 11-year-old girl presented with a painful arm mass that had been progressively evolving for 6 months. Clinical examination revealed a 4cm mass that was soft to palpation, causing electric discharge pain on percussion, with no inflammatory signs.

Imaging findings:

Ultrasound of the upper limb reveals a rounded, well-limited soft tissue mass of heterogeneous echogenicity with evidence of continuity with the median nerve and presenting a posterior acoustic enhancement (Figure 1a), measuring

approximately 4cm with moderate vascularization on color Doppler(Figure1b). The findings raised concern for a neurogenic tumor.

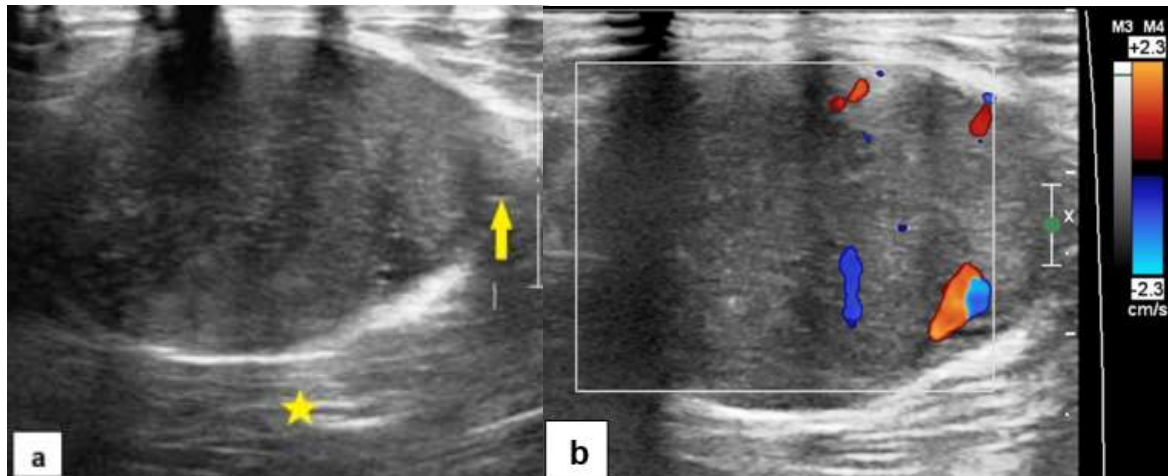


Figure 1:Ultrasound scan of the medial border of the arm.B-mode (figure 1a) and color Doppler mode (figure 1b) showing a roughly rounded, well-limited mass of heterogeneous echostructure appearing to connect with the median nerve (yellow-arrow) and showing posterior enhancement (star).This mass is vascularized by color Doppler.

Further evaluation with MRI revealed an eccentric fusiform mass within the course of the median nerve. On T1-weighted images, the mass exhibited isosignal to muscle (figure 2a) with a split fat sign(Figure 2b) and a heterogeneous signal on STIR(Figure 3a) with a T2 bright rim sign (figure 3b).It also presented a heterogeneous contrast uptake (figure 4).

No bony destruction or muscular infiltration were noted.

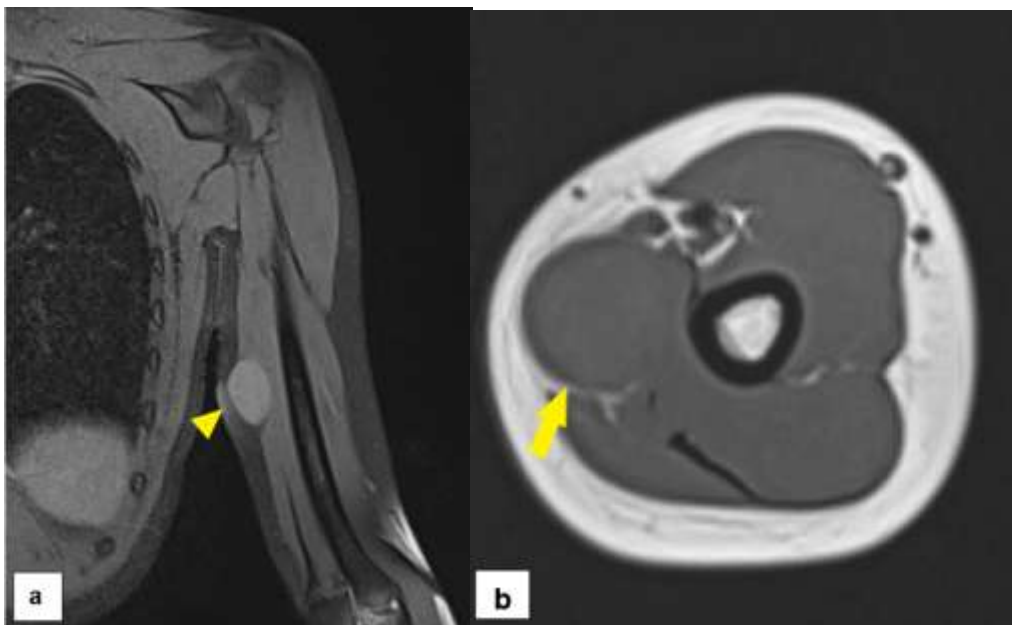


Figure 2:- Upper limb MRI: Coronal T1 fat-saturated(figure 2a) and axial T1 images (figure 2b) showing a well-defined round lesion (arrowhead), which is isointense to muscle with a fatty interface between the lesion and the arm muscular tissue (arrow).

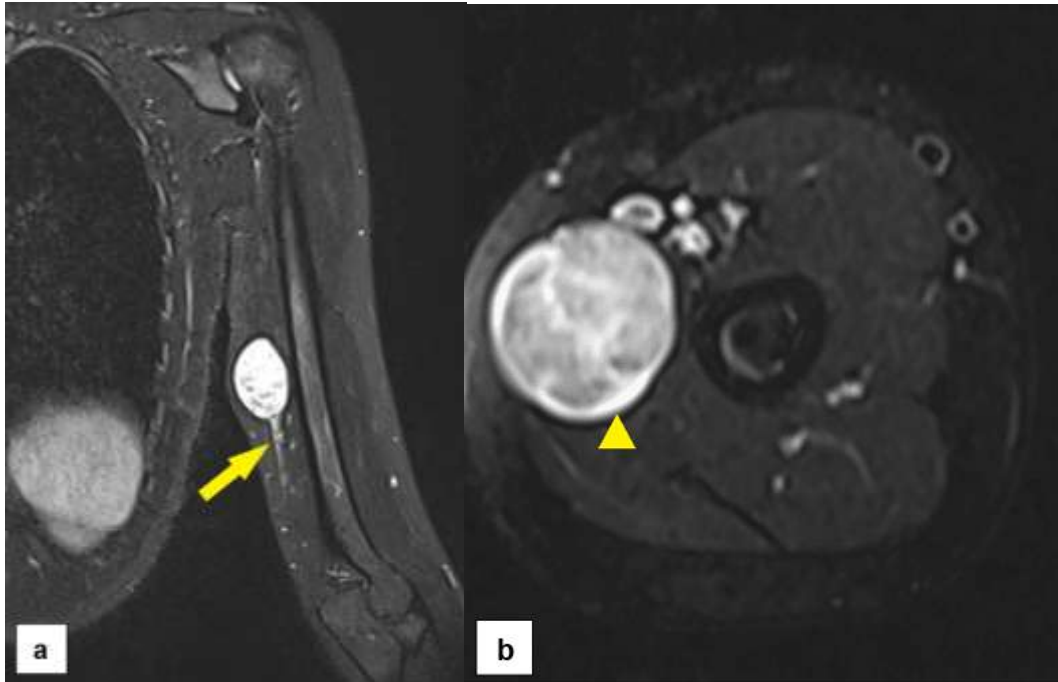


Figure 3:- Upper limb MRI;coronal STIR (figure 1a) and Axial T2 fat-saturated images (figure 1b) showing a well-defined round lesion heterogeneously hyperintense on STIR along the course of the medial nerve (arrow) with an entering and exiting nerve sign and T2 hypersignal ring (arrowhead) corresponding to the bright rim sign.

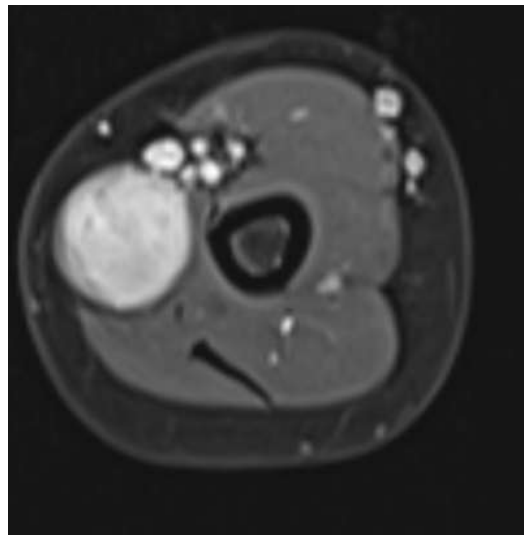


Figure 4:- Upper limb MRI: Axial T1 fat-saturated post-contrast image showing a heterogeneous contrast uptake of the mass.

Discussion:-

Peripheral nerve sheath tumors encompass a diverse group, predominantly benign, and are relatively uncommon in the general population. Specific types, such as neurofibromas and schwannomas, may arise sporadically or be associated with neurofibromatosis (NF).

Patients with these tumors typically present with a soft tissue mass, pain, or focal neurologic symptoms, with benign tumors often showing a slower, prolonged progression compared to the rapid growth of malignant tumors [1].

Imaging, especially MRI, plays a crucial role in characterizing these tumors. Ultrasound can reveal homogenous, hypoechoic features with posterior acoustic enhancement and continuity with peripheral nerves. While ultrasound can't always differentiate between schwannomas, neurofibromas, and malignant nerve sheath tumors, it helps assess their connection to peripheral nerves, guiding the diagnosis[2]. MRI is often recommended for comprehensive characterization.

MRI is invaluable in managing peripheral nerve sheath tumors. Imaging signs such as the entering or exiting nerve sign, split fat sign, fascicular sign, and target sign aid in distinguishing between benign and malignant neurogenic lesions. For instance, the entering or exiting nerve sign is associated with the tapered ends of neurofibromas, while schwannomas are eccentrically located in relation to the nerve. The target sign, typically seen with neurofibromas, represents a central fibrocollagenous core surrounded by myxomatous tissue. The fascicular sign, common in benign tumors, appears as ring-like structures on T2-weighted images. Lastly, the split fat sign, indicating fat at the upper and lower poles of a lesion on T1-weighted images, suggests the intermuscular location of the tumor and benignity, as malignant tumors tend to invade surrounding tissues. Additionally, a thin rim of T2 hyperintensity is more commonly observed in schwannomas.

Intra-tumoral cystic degeneration, more prevalent in schwannomas than neurofibromas, can lead to "ancient schwannomas" characterized by calcification and hemorrhage[3].

Malignant peripheral nerve sheath tumors (MPNST) exhibit distinct features. Benign tumors usually remain under 5 cm; significant growth raises suspicion of malignancy requiring histopathological confirmation. MPNSTs appear more heterogeneous on both T1-weighted and T2-weighted images due to necrosis and hemorrhage, a feature shared with degenerated schwannomas. Perilesional edema, seen as increased signal intensity on T2-weighted images adjacent to the primary lesion, indicates malignancy, typically absent in benign lesions. Malignant tumors may display solid or peripheral enhancement, distinguishing them from neurofibromas with central enhancement (the "Target sign"). Similar enhancement patterns can occur in degenerated schwannomas. Bony destruction supports malignancy, rare in benign tumors[4,5].

Final diagnosis:

Median nerve Schwannoma.

Differential diagnosis list:

Neurofibroma of the medial nerve.

Malignant peripheral nerve sheath tumors MPNST.

Ganglion cyst.

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