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An Unusual Schwannoma in the Proximal Part of the Left Arm – A Case Report

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Abstract **Case Report**

Background: Schwannomas are the most common benign tumors of peripheral nerves, originating from Schwann cells and accounting for 90% of all peripheral neural tumors. However, their occurrence along the musculocutaneous nerve in the upper extremity is rare. Due to their slow growth and nonspecific symptoms, diagnosis can often be delayed or challenging. Imaging modalities such as ultrasonography (USG) and magnetic resonance imaging (MRI) play a crucial role in preoperative diagnosis, though definitive confirmation requires histopathological analysis. ***Case Presentation:*** We present the case of a 22-year-old female who presented with swelling, numbness, weakness, and paresthesia in the left arm. Clinical examination revealed a mobile, round, firm mass on the volar aspect of the proximal part of the left arm with a positive Tinel's sign but no neurological deficits. Laboratory findings showed mild anemia, but other parameters were within normal limits. Imaging studies suggested an intramuscular peripheral nerve sheath tumor. USG revealed a fusiform hypoechoic mass with characteristic "split fat sign" and increased vascularity, while MRI demonstrated an encapsulated lesion with hyperintense T2 signal, fascicular sign, and intense post-contrast enhancement, consistent with schwannoma. Histopathological examination confirmed the diagnosis, revealing spindle-shaped cells with wavy nuclei and Verocay bodies, without evidence of malignancy. ***Conclusion:*** This case report highlights the importance of integrating clinical, radiological, and histopathological findings to achieve an accurate diagnosis of schwannomas, particularly in atypical locations such as the upper extremities. Early identification and appropriate management are crucial in preventing complications and preserving nerve function.

Keywords: Schwannoma, Unusual, Proximal, Left Arm.

INTRODUCTION

Schwannomas, which develop from Schwann cells, are the most common benign tumors of the peripheral nerves, accounting for 90% of all peripheral neural tumors and less than 8% of all benign soft tissue tumors [1, 2]. Schwannomas typically involve the ulnar nerve; less than 7% of these tumors are located along the median nerve sheath in the upper extremities [3]. Schwannomas are typically solitary tumors ranging from 1.5 to 3 cm in diameter, and large tumors are rare [4].

Due to the tumor's rarity, slow growth, and absence of specific symptoms, diagnosis can occasionally be difficult, delayed, or wrong [3-7]. This type of benign tumor of the peripheral nerve is most often located in the large nerve trunks, and in the upper limb and is more often located on the anterior aspect, frequently involving the ulnar nerve, in over 75% of cases occurring distal to the elbow [5-8]. In 90% of cases, the schwannoma is a tumor with a single location.[9] The presence of multiple schwannomas is related to neurofibromatosis type 2 (NF2) [5-8]. While clinical diagnosis lacks specific signs, a schwannoma may be suggested by a slow-growing mass along a nerve pathway. Such masses are often mobile transversely, relatively fixed longitudinally, and may cause pain upon palpation [8-10]. Imaging investigations like ultrasonography (USG) and magnetic resonance imaging (MRI) description have a major role in preoperative diagnosis, being considered helpful for establishing a correct diagnosis in 90% of cases [8-12]. None of these

imaging investigations is 100% accurate in diagnosing schwannoma [13].

Schwannomas' clinical symptoms and radiological images can be similar to the other soft tissue tumors. Intraoperative findings and microscopic analyses with immunohistochemistry features make differentiating schwannomas from other tumors easy [14].

In this study, we report a rare schwannoma in a 22-year-old female patient and present the clinical, radiological, and histological findings.

Case History

A 22-year-old female initially presented with swelling, numbness, weakness, and paresthesia in the left arm. On physical examination, a round, hard, mobile mass was detected at the volar aspect of her left proximal arm. Tinel's sign was positive. No neurological deficits were detected. The patient had no comorbidities and no remarkable medical or surgical history. Her general condition was optimal. Physical examination also revealed that she had a BMI of 27.4 kg/M^2. The pulse rate was 94 beats per minute and blood pressure was 110/80 mmHg. The respiratory rate was 17 cycles per minute.

Laboratory Findings:

The patient had a hemoglobin of 8.6 g/dl, and ESR was found 35 mm/1st hour. The red blood cell count was 4.5x 10^{12} /L, the platelet count was 200x 10^9 /L and the WBC count was 7x 10^9 /L indicating normal findings. The patient had a random plasma glucose of 5.6 mmol/ml. Prothrombin time (PT) was 15.00 seconds with an index of 86.67%, and APTT (Activated Partial Thromboplastin Time) was 30.30 seconds which was normal.

Histopathology findings revealed a benign neoplasm composed of spindle-shaped cells containing wavy nuclei. Verocay bodies and thick-walled blood vessels were also seen. No malignancy was seen and the diagnosis was made as Schwannoma.

Ultrasonogram Findings:

Ultrasonography (USG) of the left arm revealed a fairly large, fusiform-shaped hypoechoic mass, measuring approximately 4.2 x 1.7 cm, located in the proximal region, likely within the left biceps muscle. The mass is positioned about 0.5 cm beneath the skin surface. Triangular hyperechoic areas were observed at both the proximal and distal ends of the lesion, suggesting the presence of a "split fat" sign. Color Doppler imaging showed significantly increased blood flow within the lesion. Findings were suggestive of an intramuscular peripheral nerve sheath tumor in the left arm, with the possibility of a Schwannoma or, less likely, a Neurofibroma. [Figure 1]

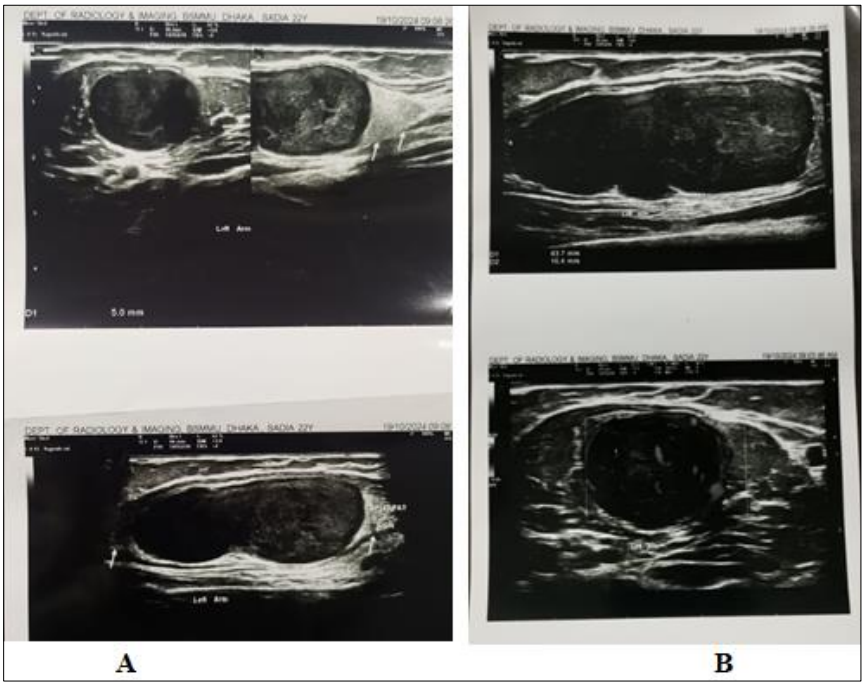

Figure 1 (A &B): Ultrasound of the patient

MRI (Magnetic Resonance Imaging) Findings:

MRI of the left mid-arm shows an oblongated encapsulated T1WI isointense & T2WI heterogeneously hyperintense lesion (measuring about CC-4.47cm x AP-1.9 cm x TD-2.0 cm) was noted in the anteromedial aspect of the left mid arm, intramuscular in a location

within biceps muscle. The lesion possibly arises from the biceps brachi branch of the musculocutaneous nerve. Following IV contrast (Gd-DTPA) administration, the lesion showed an intense enhancement. A thin, hyperintense rim is observed around the lesion on T2-weighted images. Triangular areas of T2 hyperintense fat signal were present at both the upper and lower ends of the lesion, indicating preserved fat and forming the characteristic "split fat sign." Additionally, multiple small circular areas of low signal intensity, surrounded by mild hyperintensity on T2-weighted images, create a "fascicular sign," suggestive of nerve involvement. MRI findings were consistent with an intramuscular peripheral nerve sheath tumor, most likely a schwannoma. A neurofibroma was considered a differential diagnosis. [Figure 2]

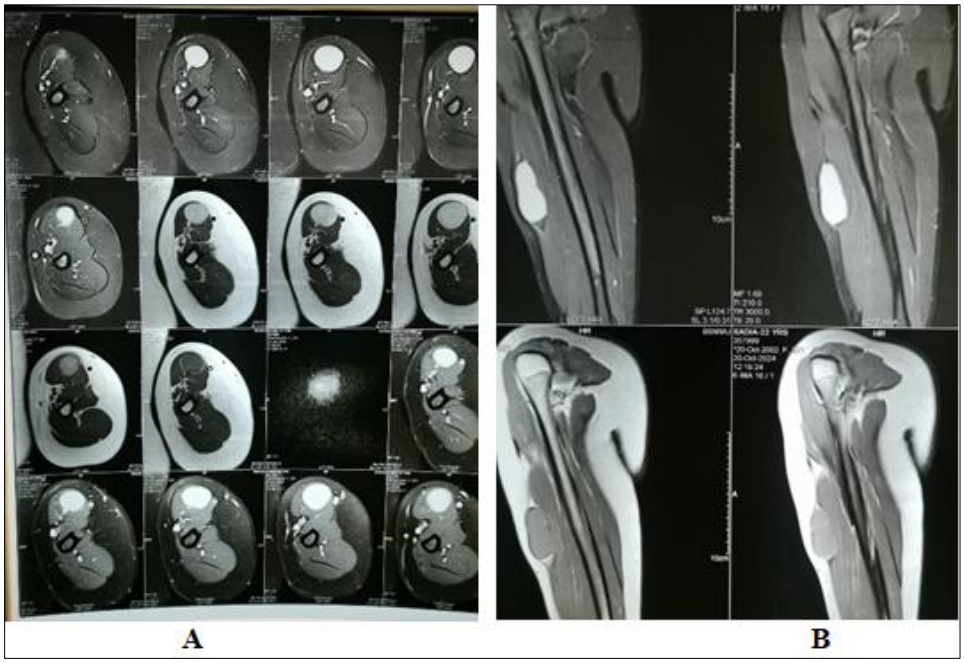

Figure 2 (A &B): MRI of the patient

DISCUSSION

Schwannomas are benign tumors that also known as neurilemmoma originate from Schwann's cells and comprise 5% of all soft tissue sarcomas [15]. Schwannosmas are common soft tissue tumors of the extremities that can be categorized as proximal i.e., those on the brachial plexus and upper arm, or distal i.e., those on the forearm and hand [16].

Most Schwannomas are covered by a neural capsule and rarely transform into malignant tumors [15]. There is no gender bias in the contraction of Schwannomas and prevalence across ages 20 to 60 is fairly even, except during middle age, during which you are more likely to contract Schwannomas [17]. Schwannomas become larger over time but rarely exceed a diameter of 3 cm [17], which is why in most cases Schwannomas gradually put pressure on neural bundles, delaying visible symptoms of Schwannomas and thus diagnosis until the late stage of tumorigenesis [17]. Fortunately, in most cases, Schwannomas remain as a distinct mass from neural tissues allowing relatively easy and complete surgical removal of these masses without causing neural deficits [18].

As aforementioned, neural deficits caused by Schwannoma are rare, but neurological symptoms arising from mass-induced pressure on nerves have been reported, the symptoms of which are a positive Tinel's sign, pain around the area of pressurization upon gentle tapping [17]. Approximately 30−70% of Schwannoma patients experience such pain and over 20% complain of hypoesthesia [17]. A recent report on a group of Schwannoma patients found all of them had to some degree hypoesthesia and/ or Tinel's sign [19]. The non-specific symptoms of Schwannomas mean that suspecting someone of a soft tissue sarcoma or neuroma is difficult just by physical examination or even by plain radiography and differentiating it from tenosynovitis or Cubital tunnel syndrome is challenging [20, 21]. Even for such symptoms, a physician should consider a slowly growing independent tumor that may originate deep inside the nerve where palpation induces a transverse but not a vertical movement of an otherwise fixed mass [18]. In this case report, the patient came with the symptoms of swelling, numbness, weakness, and paresthesia in the left arm, and no neurological deficits were detected in our patient.

Accurate diagnosis is crucial for the effective treatment of schwannomas. Ultrasound (USG) is useful in determining the tumor's location, its origin from the nerve, and its relationship with surrounding unaffected nerve fibers [1]. MRI also plays a key role in

preoperative diagnosis, as schwannomas typically appear isointense to muscle on T1-weighted images and hyperintense on T2-weighted images [22].

Schwannomas usually show uniform enhancement after contrast administration, but larger tumors such as the one, in this case, may exhibit more heterogeneous enhancement. A post-contrast T1-weighted MRI can help differentiate the tumor from the surrounding nerve fascicles. If there is suspicion of a malignant peripheral nerve sheath tumor, a preoperative biopsy is recommended to guide treatment planning [23]. When performed by an experienced surgeon, a preoperative biopsy using high-resolution ultrasound guidance in combination with MRI can be done safely and effectively.[1]

Preoperative identification is difficult and the risk of misdiagnosis is high because their symptoms are not always clear. As reported by other authors the Tinel sign, which was positive in our patient, is the most useful alert sign in the diagnosis [21-24]. A suspected schwannoma should be considered When it is possible to observe an isolated, palpable slow-growing mass, usually located on the volar surface of the limb with a positive Tinel sign [5]. Microsurgical resection is recommended for the treatment of all symptomatic schwannomas at first diagnosis and for asymptomatic cases with MRI evidence of increasing tumor size [5-25]. Neurological complications, such as sensory or motor deficits or neuropathic pain, may arise following schwannoma enucleation [5-27].

Patients with schwannomas often experience worsening neurogenic pain and may develop neurological deficits over time. In this case, an ultrasound-guided biopsy was successfully performed without any complications.

CONCLUSION AND RECOMMENDATIONS

In conclusion, schwannomas arising from the musculocutaneous nerve in the proximal part of the left arm are uncommon, making their diagnosis challenging. However, imaging modalities such as ultrasound (USG) and MRI play a crucial role in identifying these tumors preoperatively. Histopathological evaluations are essential to confirm the diagnosis and distinguish schwannomas from other peripheral nerve sheath tumors. With advanced microsurgical techniques, complete enucleation of large schwannomas can be achieved safely, minimizing the risk of complications and preserving nerve function.

Further studies and case reports with long-term follow-up need to be done to monitor for any recurrence or complications and improve treatment strategies for this rare condition.

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