

The First Reported Case of Schwannoma of Superficial Radial Nerve in the Forearm in Saudi Arabia.

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Received: 03-Aug-2022, Manuscript No. SCR-22-18680; **Editor assigned:** 05-Aug-2022, Pre QC No. SCR-22-18680 (PQ); **Reviewed:** 17-Aug-2022, QC No. SCR-22-18680 (Q); **Revised:** 19-Aug-2022, Manuscript No. SCR-22-18680 (R); **Published:** 27-Aug-2022, doi: 10.35248/2161-1076.22.12.8.401

Abstract

This report documents the involvement of Schwannoma in the superficial radial nerve in the forearm. An 18-year-old male patient presented with a soft swelling in his radial left forearm for 3 years. Neurological examination shows pain and electrical sensation on palpation over the mass otherwise no motor dysfunction. Subsequently, surgical exploration revealed that the superficial radial nerve was infiltrated by Schwannoma. The encircling tumor underwent nerve-sparing enucleation. The lesion was a pale-yellow solid mass encapsulated in a smooth membrane. Histological examination yielded the diagnosis of Schwannoma. The result of surgical enucleation was satisfactory. Postoperatively, there was no neurologic deficit and the patient had normal sensation and motor function.

Keywords: Schwannoma • Superficial radial nerve • Peripheral nerve • Sensory nerve • Neurofibromatosis type • Nerve sheath tumor

Introduction

Schwannomas, also known as neurilemmomas, are generally benign peripheral nerve sheath tumors developing from Schwann cells. Schwannoma is the most common tumor of the peripheral nerve (approximately 90% of all nerve tumors) of ectodermal origin. Frequently occurring in the mixed nerves, it accounts for approximately 5% of all soft tissue tumors [1-3]. They usually occur in the 3rd to 6th decade of life with no predilection for either sex. Schwannomas of the upper extremity have a rate of 19% of the cases [4]. It appears as a well-circumscribed oval lesion with an extravascular growth, they are usually asymptomatic painless swelling and may present with pain and paresthesia over the involved nerve. The clinical signs and symptoms are often misunderstood and somehow associated with other soft tissue tumors such as ganglions, or tenosynovitis [2, 5]. We present in this report, an 18-year-old male patient with Schwannoma involving the superficial radial nerve of the left forearm and its surgical management.

Case Presentation

History

An 18-year-old male patient, who is a known case of neurofibromatosis type 1, right-handed, presented with left forearm swelling (in the radial middle third) that had been present for three years. The swelling was increasing gradually and slowly in size with time, associated with mild discomfort or pain whenever he used his left upper limb frequently. There was no reported history of trauma, discharge, or bleeding from the mass. There was no weakness in his grip power. There were no other similar swellings in his body. There is no family history of neurofibromatosis. No history of smoking and no drug or food allergies.

Physical examination

The patient was conscious, alert, and oriented. His vital signs were within normal. Regarding his left forearm, there was a swelling in the radial middle third forearm that was oriented longitudinally over the distribution of the superficial radial nerve (Figure 1). The size of the swelling was 3.5 cm × 6 cm. It had a smooth surface, hard but elastic, immovable tumor with well-defined edges. There was no bruit or thrills. The swelling was not compressible. The overlying skin had normal color. He had positive Tinel's sign and pain over the swelling, otherwise no motor deficit.

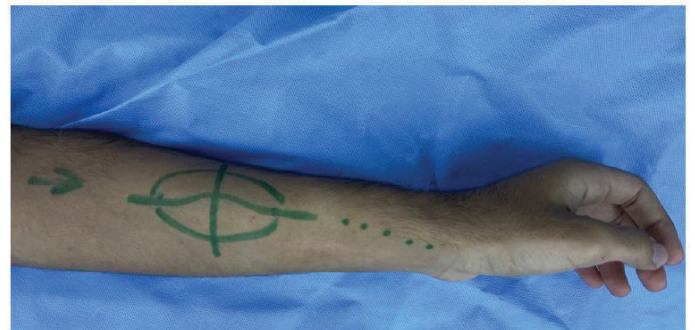


Figure 1. Showing swelling in the radial aspect of the middle third of the left forearm.

Investigations

His preoperative lab results were within normal limits.

Ultrasound

A well-defined oval-shaped structure that is heterogeneously isoechoic and measures 5.0 cm × 1.7 cm is seen at the anterior aspect of the left mid forearm (at the area of concern) showing central flow on colour doppler imaging. This lesion can be a localised neurofibroma with a known history of neurofibromatosis 1. However, further evaluation by Magnetic Resonance Imaging (MRI) is advised (Figure 2).

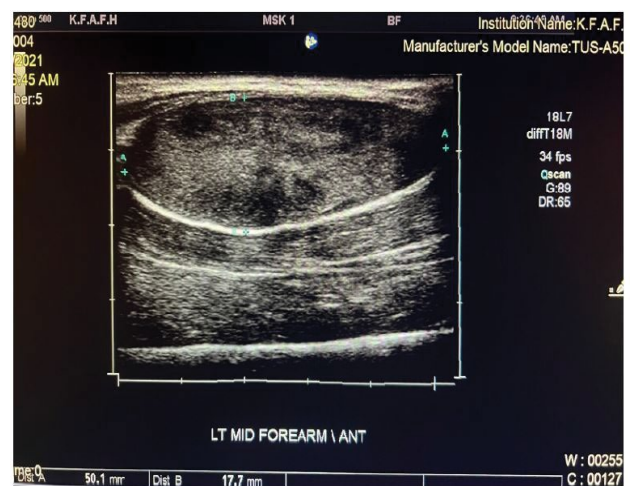


Figure 2. Showing left mid forearm mass.

Magnetic resonance imaging (MRI)

MRI of the left forearm before and post 4 contrast.

Findings: known history of neurofibromatosis 1. The examination was correlated with the prior ultrasound performed on 27-01-2022. The previously seen well-defined oval-shaped intramuscular lesion seen at the anterior lateral aspect of the left mid forearm measures 5.7 cm × 2.4 cm × 2 cm demonstrates iso-signal intensity in T1 and marked heterogeneous high signal on T2 weighted images showing central hypo intense heterogeneous signal and diffuse irregular heterogeneous intense enhancement on T1 post-contrast images. This lesion is seated between and causes the mass effect on the adjacent extensor carpi radialis tendons as well as brachial radialis as well as flexor pollicis and flexor digitorum superficialis muscles along the expected location of the radial neurovascular bundles. No obvious abnormal enhancement could be seen in the imaged muscles. No significant joint effusion. The bone marrow signal intensity of the visualized bones appears relatively maintained.

Conclusion: With the given history of neurofibromatosis, above-described well-circumscribed lesion with plates heterogeneous enhancement could be related to peripheral nerve sheath tumour (neurofibroma/schwannoma) in a proper clinical setting without excluding other aetiology for clinical and histopathological correlation (Figures 3 and Figure 4).



Figure 3. Showing superficial soft tissue swelling in the left mid forearm.

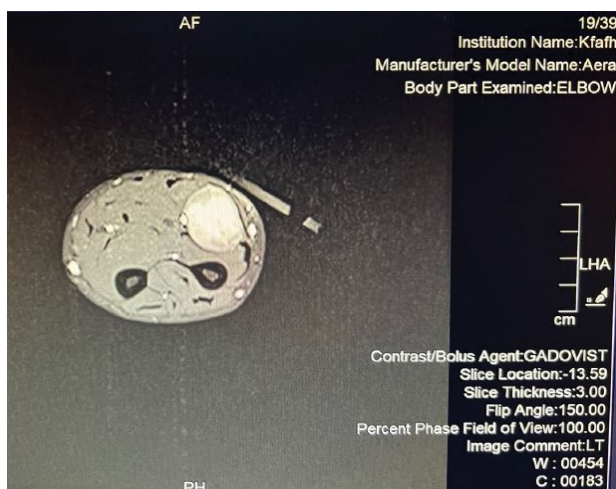


Figure 4. Showing superficial soft tissue swelling in the left mid forearm above the radius.

Procedure

After a thorough discussion with the patient and his parents, counselling regarding the possible nature of the swelling, and the procedure's risks versus benefits, he was elected to undergo an excisional biopsy of that swelling in his left mid forearm under general anaesthesia as a day surgery procedure.

We decided to approach the swelling through a lazy S-type longitudinal incision, after inflation of a tourniquet 210 mmHg, using a 15- blade (Figure 1). Following the elevation of skin flaps, subcutaneous tissue dissection was carried out using a tenotomy scissor until we reached the site of the swelling. It was evident intraoperative, that this swelling had a fusiform character with fibro fatty texture, pale yellow in colour, and encapsulated in a smooth membrane. It was found along the distribution of a superficial radial nerve, covered proximally by brachial radialis muscle (Figures 5a and 5b).



Figure 5 (a, b). Intraoperative view of the tumour encircling the superficial radial nerve.

This tumour was encircling the superficial radial nerve only. It was clear that this nerve had been compressed and flattened throughout its course. We proceeded toward precise micro vascular tumour dissection and complete enucleation using 3.5x loupe magnification and micro instruments (Figure 6).

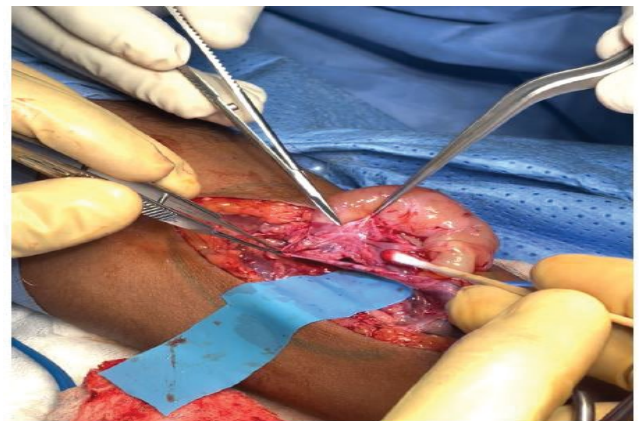


Figure 6. Enucleation of the tumour was possible with precise microvascular tumour dissection.

After achieving satisfactory tumour enucleation. However, there was no iatrogenic injury to the superficial radial nerve. The specimen was sent to the histopathology lab for definitive diagnosis. The procedure was uneventful and the patient was discharged home on the same day in stable condition (Figure 7).



Figure 7. Schwannoma of the superficial radial nerve -intraoperative aspect: a) tumour dissection and isolation b) superficial radial nerve after schwannoma enucleation c) complete tumour excision.

Histopathology

Left forearm mass: Consistent with Schwannoma. Sections reveal a well-defined lesion showing proliferation of ovoid to spindle and serpentine

nuclei, with no nuclear atypia, pleomorphism, or increased mitotic figures, with scattered lymphocytes and small to medium-sized blood vessels. The panel of immunohistochemical stains performed on the block (B) with all controls showing appropriate reactivity: The tumor cells are positive for S-100. Negative for CD34, Beta-catenin, Desmin, and Muscle-Specific Actin (MSA).

Follow-up

His first postop visit to the clinic was one-week postop. The patient did not report any sensory deficits. The Patient was satisfied with the enucleation done. Sutures were removed in the 2nd visit which was two weeks postop. The patient was referred to our occupational therapist for a range of motion exercises and scar management.

Discussion

Schwannomas, also known as neurilemmomas, are generally benign peripheral nerve sheath tumors developing from Schwann cells, it accounts for approximately 90% of all nerve tumours of ectodermal origin and it accounts for approximately 5% of all soft tissue tumors [1-3]. Approximately 90% of the schwannomas are sporadic [6], they usually occur in the 3rd to 6th decade of life with no predilection for either sex [4]. The clinical signs and symptoms are often misunderstood and somehow associated with other soft tissue tumors such as ganglions, or tenosynovitis [2, 5]. Due to the rarity of the tumor, its slow growth, and the absence of specific symptoms, diagnosis is often difficult, late, or incorrect, the diagnosis of Schwannoma is made by clinical examination and imaging tests [7]. Open biopsy of soft tissue tumors is not recommended when a peripheral nerve sheath tumor is suspected due to the risk of iatrogenic nerve injury [8]. Surgical treatment is based on symptoms or aesthetic concerns [9]. Its distribution in the body is very different, being frequently found in the ear, nose, and throat regions, followed by the trunk and then by the upper limbs (19%) and lower limbs (17.5%) [10]. Although a frequency of (7%) has been reported in the literature for radial nerve schwannomas [11], another very rare location like Schwannoma of the thumb has been reported in the literature [12]. Of 24 patients reported by Adani et al., there were no patients younger than 22 years of age. The ulnar nerve is most often affected, but involvement of the median, muscle-cutaneous and digital nerves has been described in the literature. Schwannoma may cause neurological symptoms and pain, which is not constant at clinical evaluation [13]. Perteau et al. reported that upper extremity cases would typically range between 31 years to 85 years, 17 patients were studied, and the ulnar nerve is most often affected, followed by the median, collateral digital, and lastly radial nerve [9]. Senol et al. reported a rare type of Schwannoma involving the radial nerve of a 21-year-old male patient [14]. Gosk et al. reported 34 patients, the youngest patient was 18 and the eldest was 81 years of age, in most cases the ulnar and median nerves were involved [15]. Tang et al. conducted a retrospective review of 8 adult patients ranging from 20 years to 88 years of age, 75%. 6 patients of the tumour occurred at the level of the elbow or distal to it [16]. Of 34 patients reported by Adani R et al., the youngest patient was 20 and the eldest was 78 years of age, the radial nerve was involved only in two patients [2]. Although the symptoms are nonspecific or even absent, sometimes the association of a positive Tinel sign and imaging tests (US and MRI) may suggest a diagnosis of schwannoma. There was not a reliable method to predict the presence of fascicular involvement by the tumor, including a preoperative MRI. Immunohistochemical staining was positive in all cases for S100 protein; confirming the diagnosis and differentiating it from neurofibroma [3, 9, 16]. Surgical excision or enucleation of the mass using microsurgical techniques and a meticulous microsurgical approach was possible without fascicular involvement and in some cases; resection of the involved fascicles was performed. Depending on the type of nerve involvement, a functional deficit can result [16].

Conclusion

Schwannoma, a rare benign nerve sheath tumor, must be suspected to avoid misdiagnosis or late diagnosis that may result in significant neurological damage. Although reported to be most common during the 3rd to the 6th decades of life, it may also occur as reported earlier. Symptoms are nonspecific, sometimes the association of a positive Tinel sign and imaging tests (US and MRI) may suggest a diagnosis of schwannoma. Surgical enucleation is a satisfactory treatment option when it comes to dealing with Schwannoma that is encircling a major nerve. Enucleation using precise microsurgical techniques and instruments to avoid additional nerve trauma and preserve unaffected nerve fibers. The aim of surgical enucleation is to avoid any sensory deficit from an iatrogenic injury that could ensue during the enucleation process and relieve the symptoms caused by the compressing tumor. Immunohistochemical staining was performed: S100 was positive, confirming the diagnosis of schwannoma and excluding the malignant tumors. This tumor is rare and should be thought of in patients with soft tissue tumors of the limb. A clean technique with complete tumor excision is the solution for excellent postoperative outcomes and to avoid recurrence.

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