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Case Report

Giant Schwannoma of ulnar nerve: case report☆

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ABSTRACT

Schwannomas are the most common benign neoplasms of the peripheral nerves in the upper limbs.

Although many are asymptomatic, they can produce a mass effect, thus impinging against soft tissues or interfering with joint function.

The authors present a case report and a review of a giant Schwannoma in the ulnar nerve.

Schwannoma gigante de nervo ulnar: relato de caso

Palavras-chave:
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Punho
Mão
Neurilemoma
Nervo ulnar

RESUMO

Os schwannomas são os tumores benignos mais comuns dos nervos periféricos nos membros superiores.

Embora muitos sejam assintomáticos, podem produzir um efeito de massa e assim comprimir os tecidos moles adjacentes ou interferir com a função articular.

Os autores apresentam um relato de caso e uma revisão de um caso de schwannoma gigante no nervo cubital.

Introduction

Primary neural tumors of the upper extremity are rare and represent less than 5% of soft-tissue neoplasms of the upper extremity.[1–4] Among peripheral nerve tumors, neurilemmomas are the most common[5,6] and are communally known as Schwannomas once they originate from the cells of the Schwan.[7]

☆ Work developed in the Centro Hospitalar São João, Porto, Portugal.

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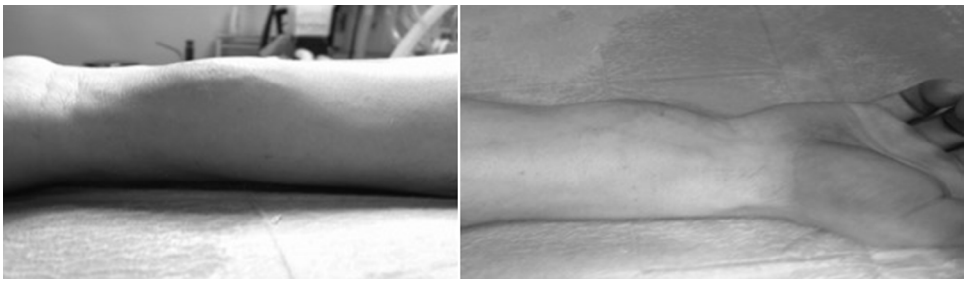

Fig. 1 – Volar tumefaction before surgery.

They are non-invasive tumors arising from peripheral nerve sheaths and are encapsulated by epineurium.[6–8] Usually, this tends to occur in patients aged 30–60 years and has no race or sex predisposition.[9,10] As they usually grow slowly and appear as painless swellings it may take many years before being correctly diagnosed. The onset of symptoms is usually associated with location rather than the size of the tumor as symptoms of neural compression arise with the growth of the mass.[11]

Although painless Schwannomas have been reported, spontaneous pain or pain after working, paraesthesia and motor weakness are the main complaints. Tinel's sign is usually present around the mass.[8–10]

The simple removal of the tumor after careful dissection is generally enough since the recurrence and malignant transformation rates are low.[1–4]

Nevertheless, the diagnosis and further treatment are not always prompt and accurate, which may lead to irreversible damage to the affected nerve with all the ensuing consequences.

The objective of this paper is to present a rare clinical case of a giant ulnar Schwannoma of the distal forearm.

Methods

All procedures performed in studies involving human subjects were approved by the Research Committee and the Declaration of Helsinki 1964 and its subsequent amendments or comparable ethical standards.

The author's present the case of a 50-year-old right-handed man, that works as a mason, who came to our office complaining of a 5-year-old growing mass of the ulnar side of the distal forearm (Fig. 1). There was no history of any significant trauma and he had not any relevant past medical history.

During our observation, the patient complained of pain and numbness in the 4th and 5th fingers that increased during flexion of wrist and fingers. He also complained of a progressive loss of strength to his left hand. The level of discomfort increasingly hindered his ability to perform his daily living activities.

Physical examination showed hypoaesthesia in the ulnar nerve territory and positive Tinel's sign precutting the ulnar mass. Despite his complains of hand weakness, the strength of the intrinsic muscles of the left hand was 5/5 without evidence of weakness or loss of range of motion

Ultrasound tests showed a hypoechoic nodule suggesting the possibility of a giant nervous sheath tumor measuring 57 mm × 27 mm × 36 mm (volume 29, 4 cc).

An MRI was performed and showed the area of the ulnar nerve with a fusiform mass with 73 mm × 35 mm × 27 mm. It presented a heterogeneous signal hyperintensity on T2 with heterogeneous contrast enhancement, outlining "split-fat sign" and "string sign". Well-defined limits were seen, with no evidence of invasion to adjacent structures. These characteristics were compatible with clinical suspicion of Schwannoma and for this reason he was proposed to surgery.

Results

A sterile tourniquet dissection assisted with loupe magnification was used in the approach to the tumor. A longitudinal incision centered in the mass was made and the first step was to identify the nerve proximal and distal to the tumor reducing injury and traction neuropraxia. After that, the dissection began with the help of the operating microscope (Fig. 2).

A longitudinal incision was created between the nerve and the tumor sheath. Once the outer layer of the tumor was identified, a plane was developed between the nerve and the tumor wall. Slow, deliberate, circumferential dissection was made to the delivery the tumor (Fig. 3).

Once the tumor was removed, the nerve was inspected for injury, the tourniquet released and precise hemostasis completed. A drain was used and the team opted for an immobilization of the forearm.

The macroscopic examination showed a nodular formation with 35 g and 7.5 cm × 3.5 cm × 2.7 cm dimensions, smooth outer surface and yellowish. Microscopic examination showed that the neoplasm as composed of spindle cells with slight atypia sometimes arranged in bundles. In the periphery of the tumor fragments of myelin fibers were observed. The neoplasm was encapsulated and the margins of the excision were clean. Necrosis was observed with very low mitotic index (<1/10CGA). Immunohistochemical study (I13/804) showed diffuse labeling in cells described for vimentin and S100 protein.

All these findings indicate a benign tumor of the nerve sheaths, most probably a Schwannoma.

The patient stayed in the hospital for 2 days after surgery. The outcome of operation was good with no complications registered. Furthermore, the patient did not require any physiotherapy.

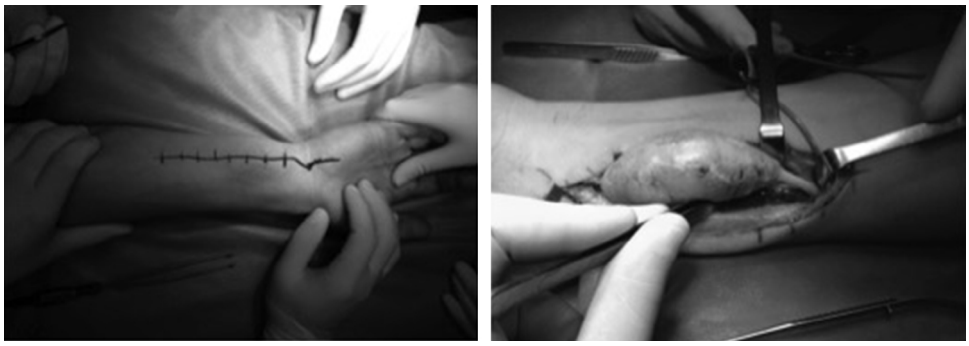

Fig. 2 – Surgical incision and approach to tumor.

Six months after the surgery the patient was very satisfied with the results with full ulnar motor and sensory function and did not have any pain or paresthesias. The subjective feeling of weakness of his hand was also lost.

Discussion

Despite rare, tumors should be taken into account in the differential diagnosis of masses in the upper limbs. In this context, it is important to remember that Schwannoma, in these cases, is the most common tumor.

Although most of these tumors show a positive Tinel sign, paresthesias, and transverse mobility their clinical identification is somehow subjective and for this reason, they are frequently misdiagnosed due to similarities with other soft tissue tumors as lipoma, fibroma, ganglion or xanthoma.[9,12]

Neurofibromas, in particular, cannot be distinguished from Schwannomas despite their differences. Solitary neurofibromas grow intraneurally and infiltrate the nerves, potentially requiring resection of all or part of nerves producing a nerve deficit.[11]

It is important to have an accurate diagnostic of the mass once it will not just interfere with the surgical procedure to take but also in the information and expectations that we can give to the patient.

Taking this into account the use of ultrasound or MRI can help to differentiate some pathologies. Nevertheless, neither MRI nor ultrasonography is 100% accurate in differentiating neurofibroma from Schwannoma.[13]

In this case, we treated an ulnar Schwannoma in the wrist, which is quite rare. We incised the capsule and removed the mass intracapsularly, as in other series.[10,14]

As described in previous papers, it is our understanding that the risk of neural damage is lower with intracapsular

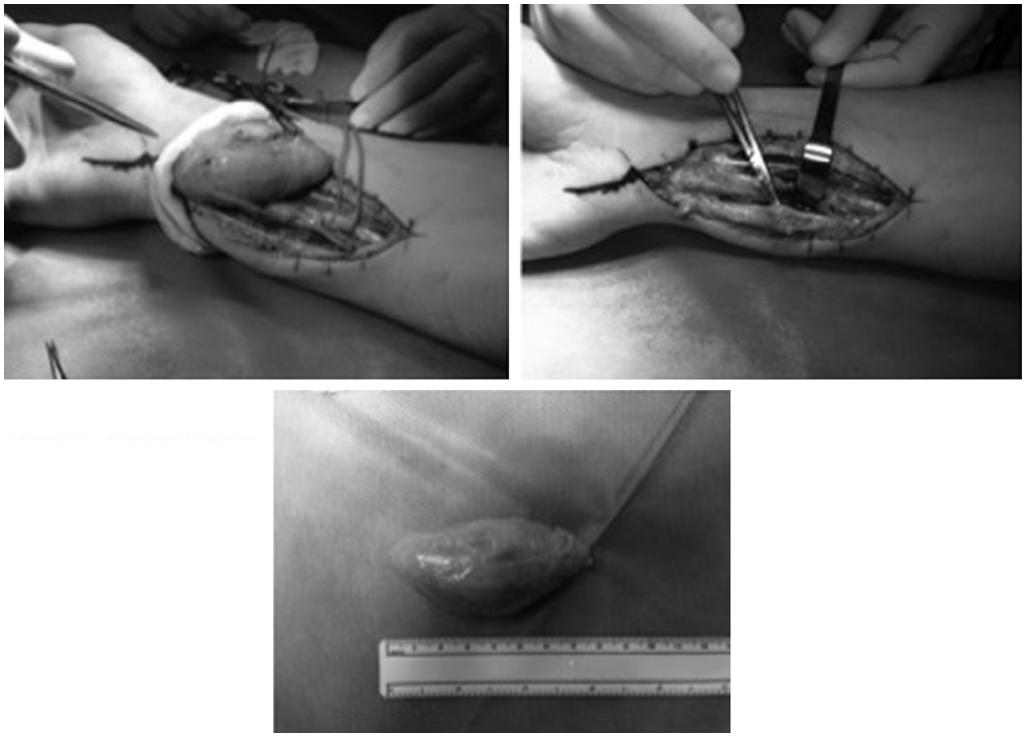

Fig. 3 – Surgical excision of the tumor.

enucleation.[11] In this case no evidence of any nerve fascicles involvement with the tumor was found, which is rarer.

As described in the literature, the presented results were excellent, as confirmed by post-op visits, with a complete improvement of the previous symptoms.

Conflicts of interest

The authors declare no conflicts of interest.

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