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Case Report

A Rare Type of Peripheral Nerve Sheath Tumor: Radial Nerve Schwannoma

Nadir Görülen Bir Periferik Sinir Kılıfı Tümörü Tipi: Radial Sinir Şıvanomu

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ABSTRACT

Schwannomas, also known as neurilemmomas, are generally benign peripheral nerve sheath tumors developing from Schwann cells. Peripheral nerve sheath tumors account for less than 8% of soft tissue neoplasms. Schwannomas are characterized by a slow-growing and non-infiltrating pattern. We report a 21-year-old, right-handed male, with a mass at his right elbow anterolateral region, that was slowly enlarging and became more painful over time. Magnetic resonance imaging of the right upper extremity revealed a 2.5x2 cm mass with heterogeneous contrast enhancement. The patient underwent complete removal of the lesion. The histopathological diagnosis was schwannoma. The postoperative course was uneventful. Clinically, these tumors may be misdiagnosed as other benign tumors, such as lipomas, synovial cysts or hemangiomas. During surgery, care should be taken to protect the nerve. Schwannomas in the upper extremities can be excised completely with preservation of nerve function and total removal lowers the risk of recurrence.

KEYWORDS: Nerve sheath tumor, Radial nerve, Schwannoma

ÖZ

Nörilemmoma olarak da bilinen şıvanomlar, "Schwann" hücrelerinden köken alan, genellikle iyi huylu periferik sinir kılıfı tümörleridir. Periferik sinir kılıfı tümörleri, yumuşak doku tümörlerinin %8'inden azını oluşturmaktadır. Şıvanomların özelliği, yavaş büyümeleri ve yayılım göstermemeleridir. Burada, sağ dirsek anterolateral bölgede, yavaş büyüyen ve zaman içerisinde ağrılı hale gelen kitlesi olan 21 yaşında, sağ elini kullanan bir erkek hasta sunuldu. Sağ üst ekstremite manyetik rezonans görüntülemesinde, heterojen kontrast tutulumu gösteren 2,5x2 cm'lik bir kitle olduğu görüldü. Kitle tamamen çıkarıldı. Histopatolojik tanı şıvanom olarak geldi. Operasyon sonrası dönemde herhangi bir sorunla karşılaşılmadı. Klinik olarak bu tümörler lipom, sinovyal kist, hemanjiom gibi diğer iyi huylu tümörlerle karışabilir. Cerrahi sırasında sinirin korunması için özen gösterilmelidir. Üst ekstremitelerdeki şıvanomlar, sinir fonksiyonu korunarak tamamen çıkarılabilmektedir ve tamamen çıkarılma, rekürrens riskini azaltmaktadır.

ANAHTAR SÖZCÜKLER: Sinir kılıfı tümörü, Radial sinir, Şıvanom

INTRODUCTION

Peripheral nerve sheath tumors are uncommon. Schwannoma, the most common type of peripheral neural sheath tumors, accounts for nearly 5% of soft tissue tumors of the upper extremities (6). Generally, schwannomas located at the upper extremities are seen in 19% of cases (3), and the most common site is immediately distal to the elbow (12). These tumors have a tendency to occur at the major nerves and at the flexor surfaces of the upper and lower extremities (7). Schwannomas arising in the radial nerves, located at the extensor compartment, are very rare.

CASE REPORT

A 21-year-old right-handed male presented with a slowly-enlarging mass in his right elbow anterolateral region with progressive pain over time.

The neurological examination was normal. The mass was mobile in a transverse plane and immobile along the parallel axis. Magnetic resonance imaging (MRI) of the right upper extremity revealed a mass with a size of 22 x 20 x 18 mm. The mass was located on the radial nerve course, medial to the midline and near biceps brachii muscle tendon and brachial artery, anterior to the supinator and brachial muscles, and posterior to brachioradial muscle. The mass was hypointense on T1-weighted MRI (Figure 1), hyperintense-heterogeneous on T2-weighted MRI (Figure 2), and hyperintense on fat sat images (Figures 3A, B). Peripheral contrast enhancement was seen with gadolinium injection (Figure 4).

The operation was performed with microsurgical dissection. The tumor had a capsule, and not all the nerve fibers were englobed. It was removed without damage to the surrounding nerve. The histopathological diagnosis was schwannoma. The postoperative course was uneventful without any complications.

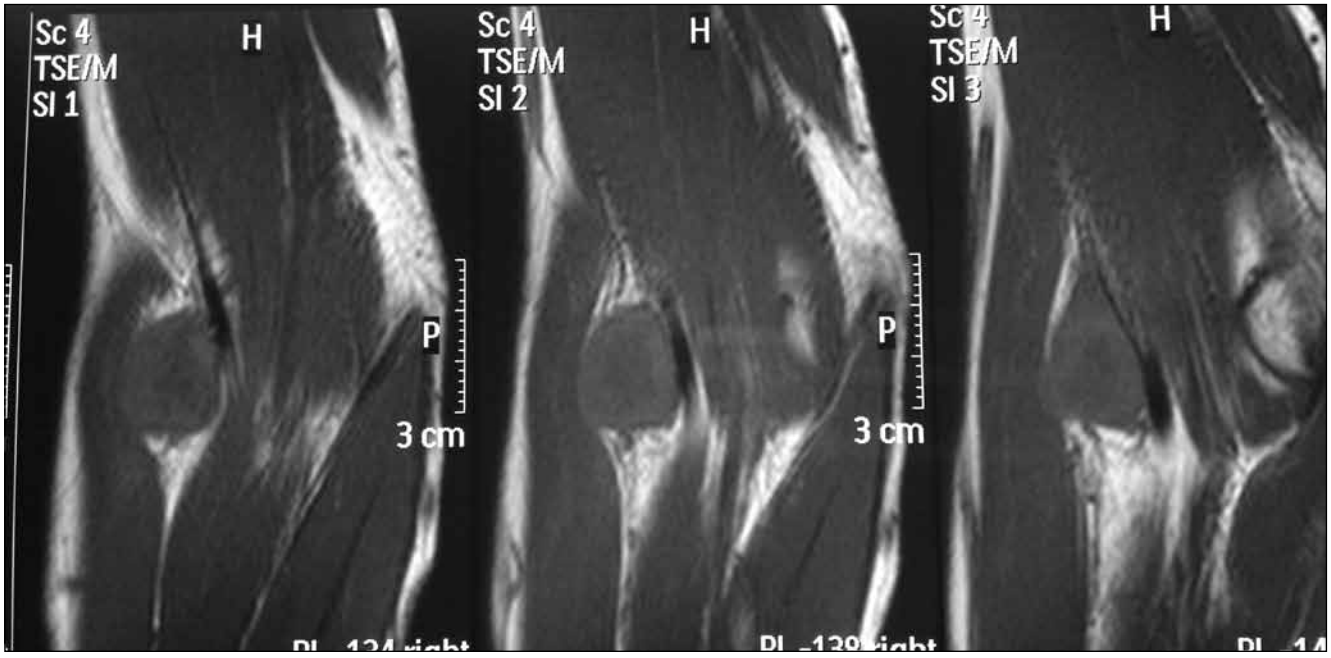

Figure 1: T1-weighted sagittal magnetic resonance imaging revealed an isointense mass that was measured as 22x20x18 mm.

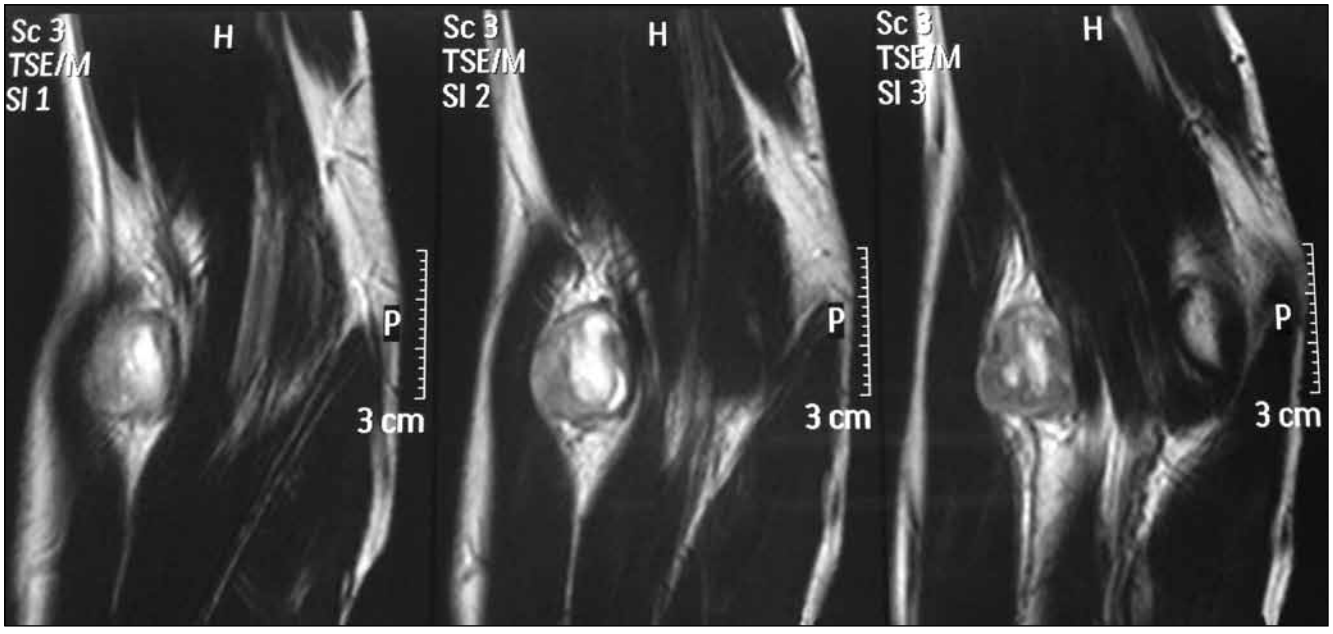

Figure 2: T2-weighted sagittal magnetic resonance imaging revealed a hyperintense-heterogeneous lesion.

DISCUSSION

Schwannomas are noninvasive tumors of the peripheral nerve sheath. According to the epidemiological study by Sandberg et al., schwannomas are the most common peripheral nerve sheath tumors of the upper extremities, with a frequency of 64% (13). The ratio of occurrence in the upper limbs to lower limbs is 2:1 and the tumor is usually seen in 30 to 60 years old patients. There is no sex or race predominance (11).

Although a frequency of 7% has been reported in the literature for radial nerve schwannomas, we have found only a few case reports (4). Kim et al. (9) and Lee et al. (10) have reported cases with radial nerve schwannoma. Ozdemir et al. have reported long-term results of 14 cases of schwannoma of the hand and wrist, of which 10 cases were localized in the median nerve, and 4 cases in the ulnar nerve (11). In addition, Adani et al. have analysed 24 patients with schwannoma of the upper limbs (1). They have reported ulnar nerve involvement in 14 patients, median nerve involvement in 4 patients, musculocutaneous nerve involvement in 3 patients and digital nerve involvement in 3 patients. In an epidemiological study performed by Sandberg et al. covering 22 years, the median nerve was

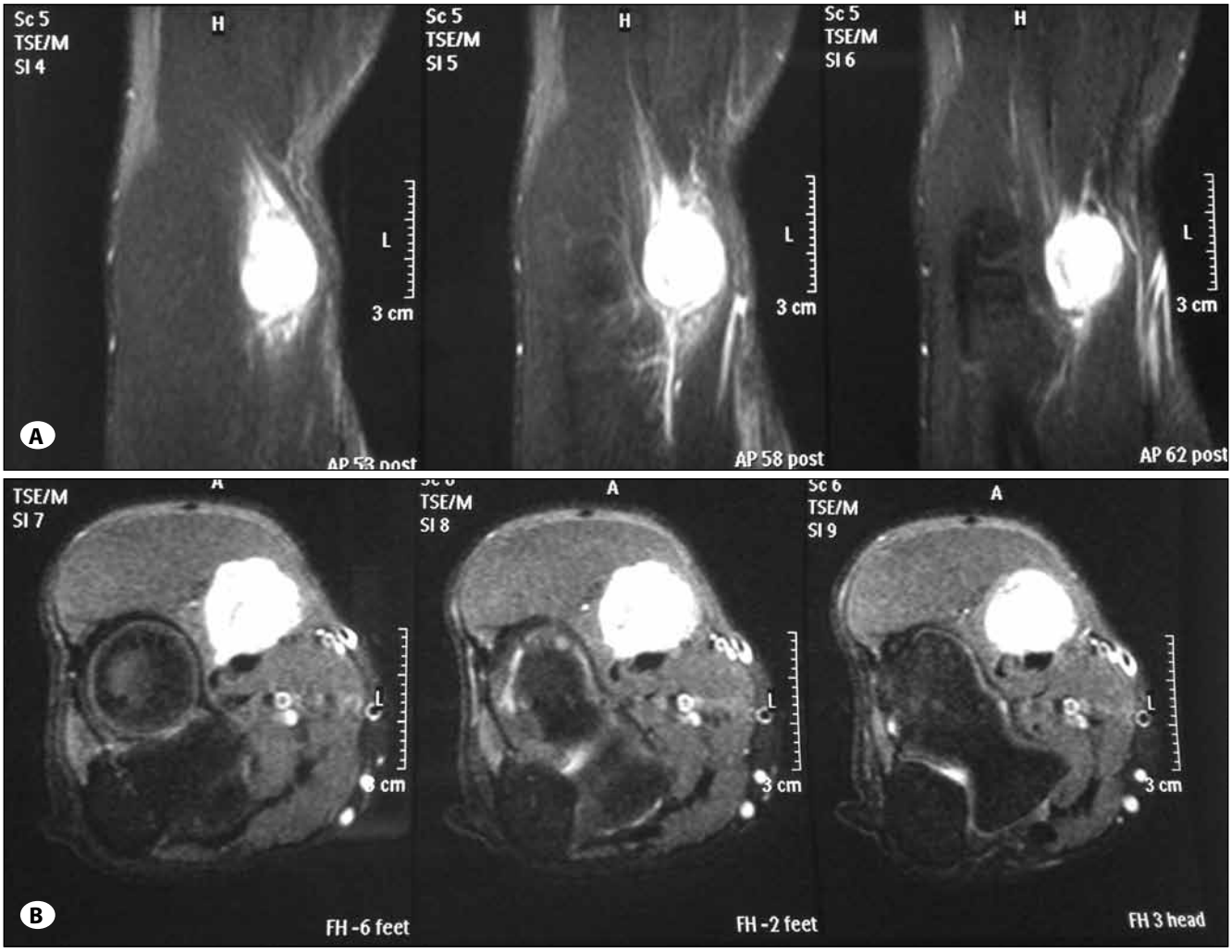

Figure 3A, B: Fat sat sagittal and axial images with gadolinium injection.

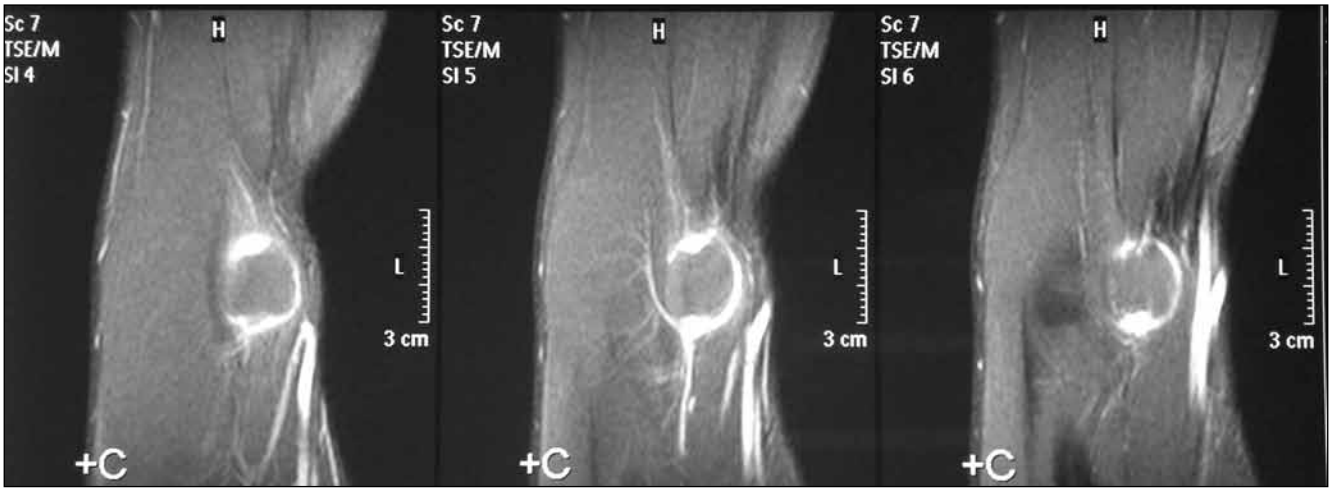

Figure 4: Peripheral contrast enhancement can be seen with gadolinium injection in sagittal imaging.

reported to be the most commonly affected nerve, followed closely by ulnar and digital nerves in 42 schwannomas of 53 patients with 68 tumors (13).

Clinical presentations of schwannomas vary. Although tumors without pain have been reported, symptoms such as pain, paresthesia and Tinel's sign are usually associated with the location of the tumor. A literature review of patients with schwannoma of the hand and wrist reveals that the interval between onset of symptoms and surgery varies between a few months and 37 years (11). Tumors located in the digital area present symptoms earlier than tumors located in the wrist and palm regions.

Benign tumors, such as neurofibroma, ganglion cyst, lipoma, xanthoma, and rarely malignant tumors should be considered in the differential diagnosis. There are no distinct criteria in differentiating schwannoma and neurofibroma clinically. Neurofibromas tend to occur at 20-30 years of age and schwannomas at 30-50 years of age. Our patient was 21 years old (11,13).

Ultrasonography can be used in the preoperative diagnosis of soft tissue tumors of the extremities. However, Hoglund et al. (5) have reported that ultrasonography had a 59% rate of accuracy as the first diagnostic alternative. Differentiating schwannoma from other tumors may not also be possible, because all reveal similar hypoechoic masses with distal sound enhancement. Although MRI is more accurate in locating and making the differential diagnosis of schwannomas, it does not have 100% accuracy either. Electrodiagnostic tests (i.e., electromyography) may have normal results in the absence of a neurological deficit, since only a small part of the nerve fibers are affected. Needle or open biopsy is not appropriate for the diagnosis either, as they can cause scarring, and fascicular damage may occur during subsequent resection (12).

Some sensory nerves can be encountered in or near the operative field and can be damaged by transection, retraction or cautery that can be the cause of neurological morbidity during surgery (14). Nerve sheath tumors are usually removed en bloc, but minimizing retraction of the surrounding fascicles is important.

Different surgical techniques, involving removing the tumor either with or without its capsule, have been described. We preferred to remove the tumor with its capsule according to the technique performed by Kececi et al (8). Donner et al. (4) also recommends extracapsular excision. On the other hand, Ozdemir et al. (11) have nucleated the tumor and removed the lesion intracapsularly in their study. They have suggested that there is less neural damage with this technique.

Although recurrence rates after surgical removal are low, incomplete tumor resection or misdiagnosis of multiple tumors may cause recurrences. Artico et al. (2) have reported recurrences in different areas of the nerves of the same extremity, not in the operated region, in their study of 119 cases with benign nerve sheath tumors.

CONCLUSION

In this case report, we have highlighted a rare location of schwannoma, the radial nerve, and have emphasized that the differential diagnosis should be done carefully in order to preserve the nerve during the surgical approach.

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