CASE REPORTS

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Large schwannoma of the median nerve at the distal forearm

Veliki švanom medijalnog nerva u distalnom delu podlaktice

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Abstract

Introduction. Schwannoma, also known as neurilemmoma, is a rare but one of the most common tumors of the peripheral nerves. It originates from Schwann cells of the peripheral nerve sheaths. Schwannoma mostly occurs in adults between 20 to 70 years of age. The regions where the tumor most commonly occurs are the head and neck, but it can occur almost anywhere in the body or the organs. Schwannomas are usually up to 2.5 cm in size but may grow to 4–5 cm. In this study, the rare case of large schwannoma of the median nerve in the distal part of the forearm is presented. **Case report.** A 46-year-old male patient was referred to a plastic surgeon with a diagnosis of lipoma on the anterior side of the distal third of the left forearm. Ultrasound and magnetic resonance imaging were done, and the surgery was performed after that. An encapsulated tumor of the median nerve was found, and the tumor was completely removed without nerve damage. Histological analysis showed a benign schwannoma of cellular type and biphasic shape. In the postoperative course, there was transient paresthesia. One year after surgery, no tumor recurrence nor neurological deficit was recorded. **Conclusion.** Schwannoma is the most common benign tumor of peripheral nerves. Schwannomas over 5 cm in size are extremely rare. Appropriate physical examination, preoperative imaging studies, and histological verification are required for the final diagnosis. The method of choice in treating large schwannomas is complete surgical excision.

Key words:
diagnosis; median nerve; neurilemmoma; peripheral nervous system neoplasms; schwann cells.

Apstrakt

Uvod. Švanom, poznat i kao neurilemom, je redak ali jedan od najčešćih tumora perifernih nerava. Potiče od Švanovih ćelija nervnog omotača. Švanom se najčešće javlja kod odraslih osoba, između 20 i 70 godina starosti. Uglavnom se javlja na glavi i vratu, ali se može pojaviti na skoro svim delovima tela ili u organima. Švanomi su najčešće veličine do 2,5 cm, ali mogu dostići i dimenzije do 4–5 cm. U ovom radu prikazan je bolesnik sa švanomom medijalnog nerva u distalnom delu podlaktice. **Prikaz bolesnika.** Osoba muškog pola, starosti 46 godina, upućena je specijalisti platične hirurgije, sa dijagnozom lipoma na prednjoj strani distalne trećine leve podlaktice. Urađeni su ultrasonografija i magnetska rezonanca, a nakon toga je urađena operacija. Utvrđeno je da se radi o inkapsuliranom tumoru medijalnog nerva i tumor je uklonjen u celini, bez oštećenja nerva. Histološka analiza je pokazala da se radilo o benignom švanomu celularnog tipa i bifaznog oblika. U postoperativnom toku postojala je prolazna parestezija. Godinu dana nakon operacije nije zabeležen recidiv tumora, niti neurološki deficit. **Zaključak.** Švanom je najčešći benigni tumor perifernih nerava. Švanomi dimenzija preko 5 cm su izuzetno retki. Adekvatan klinički pregled, preoperativna dijagnostika i histološka verifikacija neophodni su za postavljanje konačne dijagnoze. Kompletna hirurška ekscizija velikih švanoma je metoda izbora u lečenju ovih tumora.

Ključne reči:
dijagnoza; n. medianus; švanom; nervni sistem, periferni, neoplazme; švanove ćelije.

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Introduction

Schwannoma (neurinoma, neurilemmoma) is a tumor of nervous tissue that arises from Schwann cells of the nerve sheath. It is almost always a benign tumor (in more than 90% of cases), and with neurofibroma, it is the most common tumor of nervous tissue [1]. Schwannoma accounts for less than 8% of soft tissue neoplasms. It usually occurs in middle age, equally regarding race and gender [2]. It can develop almost anywhere in the body, but the most common localizations are the head and neck (more than 40% of cases). The vestibular nerve is the most common location, with a prevalence of 1 in 2,000 adults. Incidentally diagnosed schwannoma on magnetic resonance imaging (MRI) study or autopsy are more common. The occurrence of schwannoma in parenchymatous organs is rare.

Schwannoma is oval, soft, light in color, encapsulated and clearly demarcated, mobile, and usually up to 3 cm in diameter. Its growth is mostly slow and asymptomatic because it dislocates nerve fibers. In most cases, schwannoma is a solitary tumor, and multiple forms are rare and hereditary [1, 3]. Schwannoma is thought to be caused by dysfunction of the *Nf2* tumor suppressor gene. The histological appearance of schwannoma is biphasic because it consists of the Antoni A and Antoni B areas. Antoni A areas are composed of compact spindle cells with elongated nuclei arranged in fascicles and bands. Antoni B areas consist of hypocellular zones and a loose myxoid matrix. There are several forms of schwannoma: classical (most common), cellular, plexiform, and melanotic [4]. The diagnosis of schwannoma is based on the clinical picture, ultrasound examination, MRI, and histological findings. The differential diagnosis includes various subcutaneous soft tissue tumors. The treatment is surgical, and it involves complete surgical excision of the entire tumor, with generally good results. Tumors of the forearm soft tissues are numerous because of the different tissues in that region. Many of them, like schwannoma, are oval, soft, mobile, and painless. Besides, forearm nerve tumors are rare, and they can be misdiagnosed as fibroma, lipoma, fibrolipoma, ganglion, giant cell tumors of tendon sheaths, subcutaneous hemangiomas, malignant subcutaneous sarcomas, etc. There are not many reports of cases of larger median nerve schwannomas in the forearm [5].

Therefore, the aim of this study was to present a rare case of the large schwannoma of the median nerve in the distal part of the forearm, the diagnostic procedures, and the treatment result.

Case report

A 46-year-old man was referred to a plastic surgeon with a diagnosis of a lipoma of the left forearm. Six months earlier, the patient noticed a subcutaneous growth that gradually increased. Pain occurred occasionally, especially during flexion of the fingers and after pressure on the tumor. The tumor was on the anterior side of the distal third of the left forearm, as a solitary subcutaneous soft and mobile oval mass, with well-defined borders (Figure 1). The dimensions of the tumor were about 6 × 4 cm, and there were no changes on the skin above the tumor. The tumor was mobile from medial to lateral but not in the longitudinal direction. There were no pulsations nor fluctuations, and Tinel's sign was positive. The neurocirculatory finding on the hand was normal, and the axillary lymph glands were not enlarged. Additional tests, such as ultrasound examination and MRI were performed. Ultrasonography showed a round, homogeneous, and hypoechoic tumor with regular margins and no internal blood flow (Figure 2). MRI showed a well-circumscribed mass in the distal part of the anterior forearm with the displacement of surrounding structures and without direct invasion. Typical MRI features for schwannoma were present – T1-weighted iso-to hypointensity (Figure 3) and T2-weighted heterogeneous hyperintensity (Figure 4).

Surgery was performed under general anesthesia and under tourniquet control. After a longitudinal lazy-S incision, an encapsulated tumor of the median nerve was identified

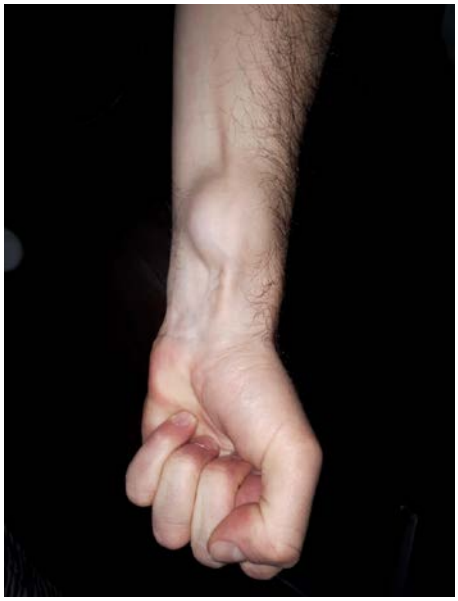

Fig. 1 – Soft tissue mass of the distal forearm.

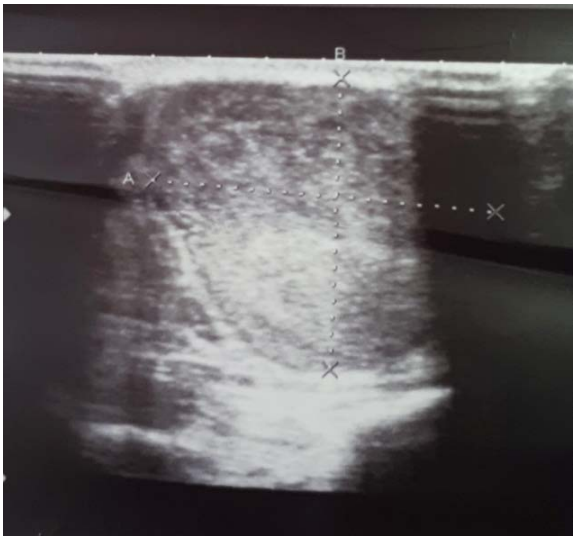

Fig. 2 – Ultrasonography finding of a round, homogeneous, and hypoechoic tumor, with regular margins and no internal blood flow.

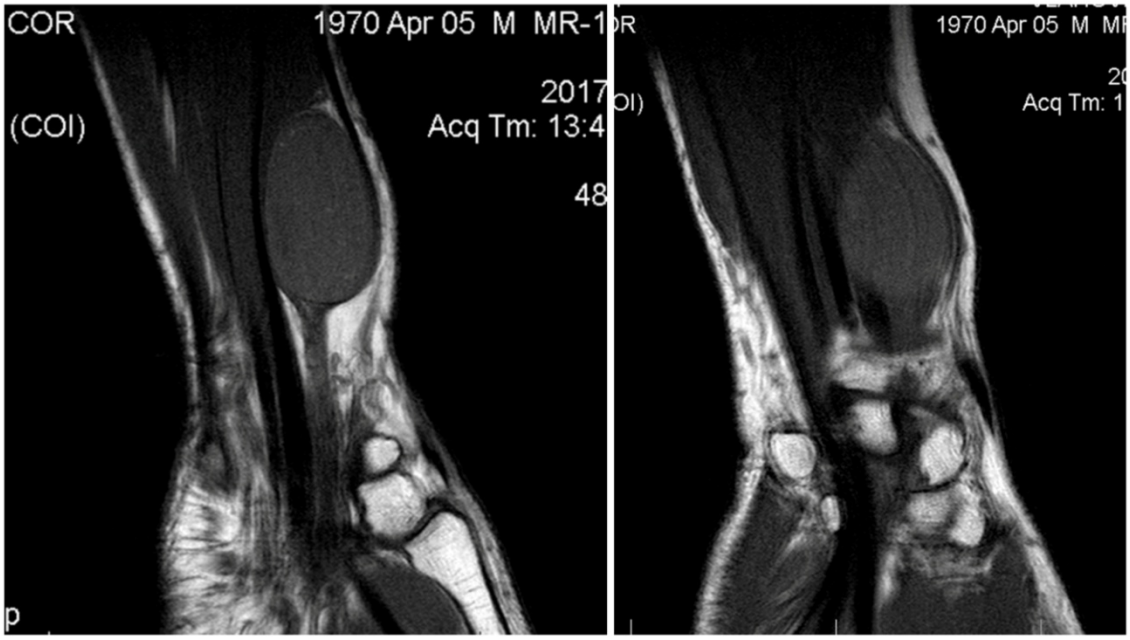

Fig. 3 – T1-weighted sagittal magnetic resonance images of a well-defined large oval mass at the flexor aspect of the forearm along the course of the median nerve, proximal to the carpal tunnel.

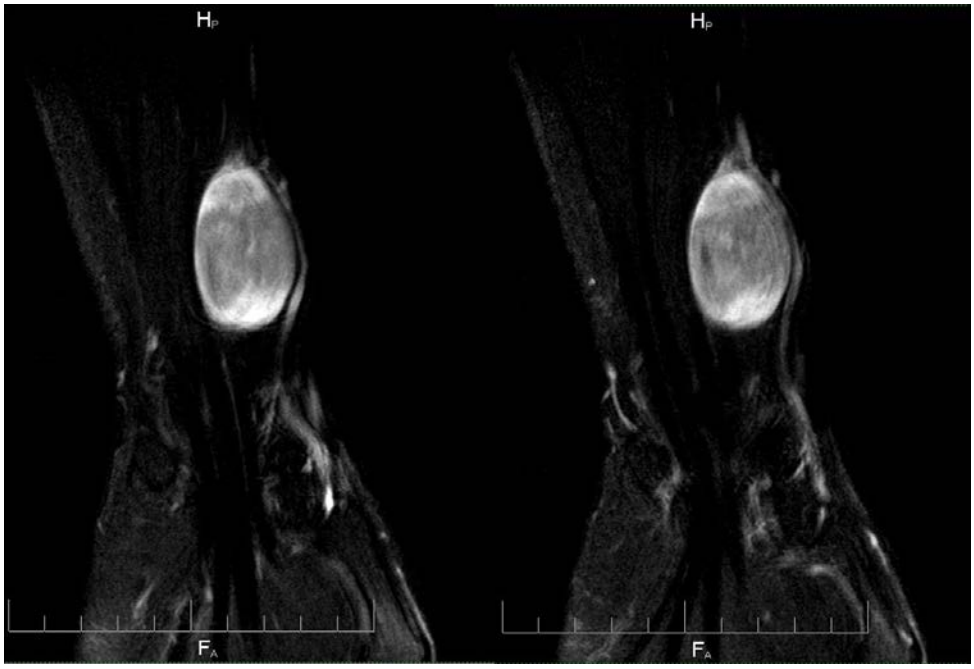

Fig. 4 – T2-weighted sagittal magnetic resonance images that show heterogeneous hyperintensity of well-defined large oval mass at the flexor aspect of the forearm.

(Figure 5). The color of the tumor was yellow, and it was very similar to lipoma. The tumor was exposed using binocular loupes, and after identifying the site where there was a splaying of fascicles, an epineurotomy was done longitudinally. Normal nerve fibers were identified proximally, and the tumor was dissected from fascicles and shelled out from the median nerve. The nerve was inspected to confirm continuity. The dimensions of the tumor were 6.2 × 4.3 cm.

Histological examination showed a well-encapsulated cellular proliferation composed of a biphasic tumor with highly cellular areas, called Antoni A area (Figure 6a), nuclear palisades area (Verocay bodies, Figure 6b), in addition to hypocellular areas (Antoni B, Figure 6c). On immunohistochemistry, the tumor cells were strongly positive for S-100 (Figure 6d).

In the postoperative period, there was transient paresthesia in the innervation area of the median nerve, and the wound healed regularly with good motor function (Figure 7). One year after surgery, there were no signs of tumor recurrence or neurological deficit. The ninhydrin test confirmed a good sensory function.

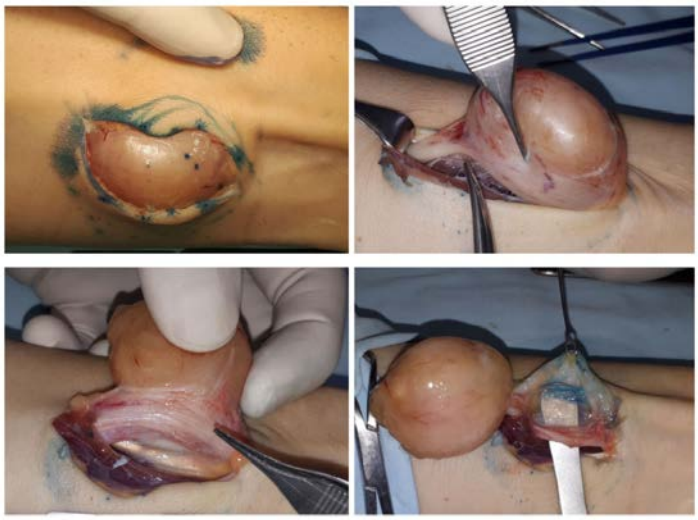

Fig. 5 – A large tumor of the median nerve, during surgery.

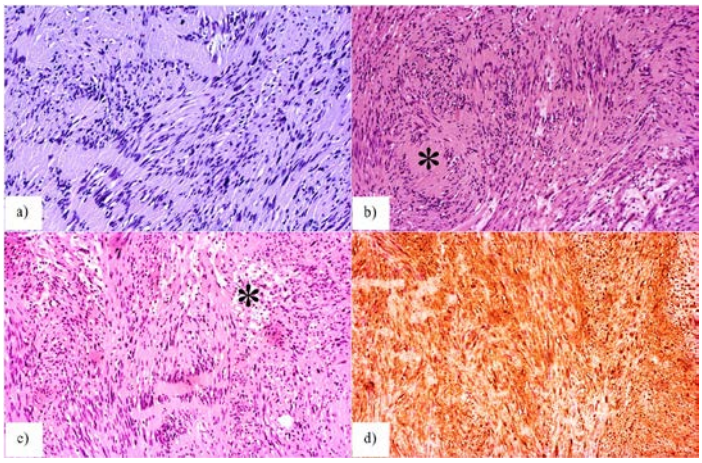

Fig. 6 – Histological examination of the tumor, hematoxylin and eosin staining, × 400 magnification: a) Tumor cells arranged in Antoni A areas; b) Nuclear palisading around the fibrillary process (Verocay body, marked by an asterisk); c) Tumor composed of loose hypocellular areas (Antoni B area, marked by an asterisk); d) Immunohistochemical staining of the tumor cells with S-100 showing strong immunoreactivity of the tumor.

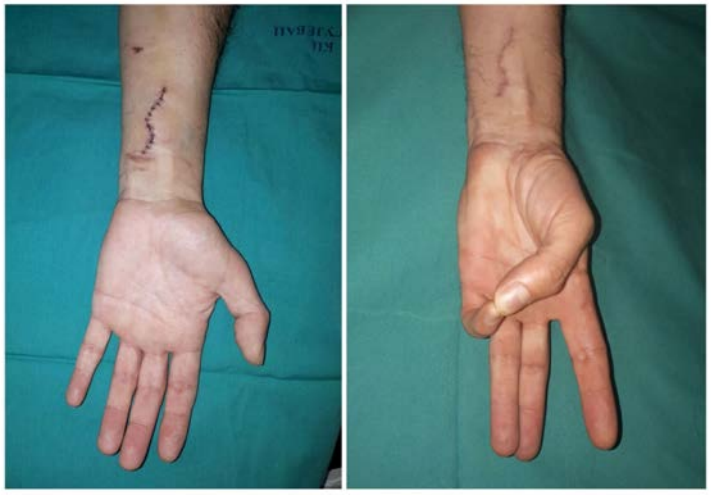

Fig. 7 – Seven days (left side image) and three months (right side image) after the surgery with good wound healing and good hand function.

Discussion

Schwannoma is a rare and the most common nerve tumor that can affect central and peripheral nerves of all types (motor, sensitive, and autonomic). It belongs to the group of peripheral nerve sheath tumors (PNST) [4, 6, 7]. Schwannoma was reported first by Verocay in 1910 and was named "schwannoma" by Masson in 1932. Schwannoma is usually benign, solitary, and asymptomatic [3]. Schwannomas almost always appear in adults. They are extremely rare in children, and only a few cases of congenital schwannoma have been described. Schwannoma most commonly occurs in adults between 20 and 50 years of age, with no difference regarding gender [8]. The presented patient was 46 years old when schwannoma was diagnosed. Almost half of the schwannomas are located on the head, mostly the vestibular nerve, but they can be found in any part of the body, even in the bones [9]. Most schwannomas are solitary, with diameters from 1.5 to 3 cm [10]. The schwannoma of the presented patient was unusually large, with dimensions of 6 × 4 cm. A noticeable subcutaneous mass, which is oval and soft, may be seen on the extremities. The tumor may present in different colors, such as yellow, white, brown, gray, or pink.

Symptomatology depends on the nerve type, tumor size, and anatomical region. Most schwannomas are asymptomatic. Numbness and pain occur in larger schwannomas and in those located in specific areas [11]. Schwannomas are usually painful to palpation on and around the mass when located superficially on extremities. The main difference between schwannoma and neurofibroma is that neurofibroma infiltrates the nerve. Lipoma is often found on the forearm as a soft, painless mass, mobile in all directions, even vertically, and it is usually not fixed in depth. They are similar to fibrolipomas but are of a slightly harder consistency. Ganglions are mostly painless and soft in consistency but often fixed to deeper structures, such as tendon sheaths, and are uncommon in the forearm. Giant cell tumors of tendon sheaths are lobular, are not soft, are fixed to tendons, and are not common tumors in the forearm. Subcutaneous hemangiomas can be found as soft and hard tumors, usually have a different color from the surrounding skin, can be pulsating, and sometimes have a lobular surface. Malignant subcutaneous sarcomas are less common, usually have a harder consistency, are fixed, and often cause stronger motor and sensory deficits.

Schwannoma can be associated with genetic disorders. The occurrence of multiple schwannomas is referred to as schwannomatosis, which is an autosomal dominant inherited disease. It is also called neurofibromatosis (NF) type 3, and there is a mutation in *SMARCB1* or *INI1* tumor suppressor gene [11].

Malignant schwannoma is rare and belongs to the group of sarcomas referred to as malignant PNST. It is most common in type I NF (gene defect in chromosome 17), with a risk of 10%. Malignant alteration of schwannoma to epithelioid sarcoma has also been described [4].

Subcutaneous tumors of the forearm are relatively common, but tumors of the forearm nerves are rare. There are some statistics on the frequency of subcutaneous soft tissue tumors. Still, in the hand region, the most common are ganglia (more than 50% of cases), followed by giant cell tumors of tendon sheaths, mucous cysts, lipomas, glomus tumors, vascular tumors, and, finally, tumors of the nerves (schwannoma, neurofibroma) [12, 13]. Concerning hands, schwannomas of the median nerve make up 0.1 to 0.3% of all hand tumors. In the presented patient, schwannoma of the median nerve in the distal part of the forearm had an extremely rare localization for this tumor type.

The diagnosis of schwannoma is made by physical examination, additional imaging, and biopsy [13]. On extremities, there is a soft and oval subcutaneous mobile mass throughout the nerve, typically without motor deficit. Differential diagnosis includes many soft tissue tumors. It is difficult to distinguish schwannoma from neurofibroma in a physical examination because, due to the slow growth of a benign nerve tumor, nerve function adapts to this compression effect [14]. Schwannomas can be distinguished from lipomas and ganglions by painful palpation and the Hoffman-Tinel sign. If there is a motor deficit, malignancy should be suspected. The significance of this case presentation is the fact that the patient was referred to a plastic surgeon by a primary care doctor with an initial diagnosis of a lipoma. That can be misleading for some less experienced surgeons who might believe that it really is a lipoma, and as a consequence, iatrogenic nerve injury may occur during the surgery.

Ultrasonography of schwannoma shows round or ovoid, hypoechogenic homogenous mass with clear margins [15]. On the extremities, this finding is similar to ganglia when there is a small lesion or to lipomas and fibrolipomas when the tumor is larger. In smaller schwannomas, there is a cystic appearance, but in large and long-lasting tumors, heterogeneous lesions may be seen, with signs of hemorrhage, calcifications, and fibrosis [16]. In that case, it is not easy to distinguish it from vascular lesions and even malignancies.

MRI is essential in diagnosing schwannoma. It is sometimes the diagnostic method of choice [16], especially for schwannomas of smaller nerves. MRI finding is typical, with T1-weighted iso-to hypointensity, hyperintensity on T2-weighted images, and postcontrast enhancement [12]. The borders of the tumor are smooth and well-defined, with a split fat sign. Otherwise, malignancy can be suspected, particularly if the tumor is over 5 cm in size. Entering and exiting nerve sign is usually seen on extremities. MRI is good for evaluating the anatomical site of the tumor, the size and extent of the tumor, and the relationships between the nerve and other tissues [17]. It is believed that MRI angiography has up to 91% accurate diagnosis of schwannoma.

The main treatment for schwannoma is surgical removal. Complete tumor excision is usually possible since the tumor does not involve nerve fibers. On the extremities, it is important to perform the surgery under tourniquet control, using the meticulous surgical technique to remove the tumor completely without damaging the nerve fibers. After dissection from proximal to distal and tumor identification, epineurotomy is performed, followed by circumferential dissection. In this way, the whole tumor is usually removed without any

difficulty [18]. In contrast, excision of neurofibroma and malignant schwannoma is often accompanied by nerve damage, and nerve grafting may be required. Complications after the excision of schwannoma are rare and transient paresthesia is the most common as a result of neuropraxia. Motor deficits are rare, and the recurrence rate is low. Early physical therapy is of great importance [9]. The patient we present in the paper had no complications after the surgery, and the hand function was preserved.

The limitation of this article was the fact that the MRI was done without contrast; therefore, we were unable to present the images with post-contrast T1-weighted sequences, which would have been better since they show more clear characteristics of schwannoma.

Conclusion

Median nerve schwannomas in the forearm are rare, especially those of larger dimensions. It is essential to distinguish them from other tumors of soft tissues, especially lipomas. The significance of this case study is that it presents a rare large-sized tumor of the median nerve with a rare localization. Physical examination is mandatory, but ultrasonography and MRI are very useful in choosing an appropriate surgical technique. Histopathological examination is also mandatory for differential and final diagnosis. Complete excision of large schwannomas in extremities is possible with adequate diagnosis and microsurgical technique under tourniquet control, without major complications or recurrence after surgery.

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