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A rare form of schwannomatosis on the volar surface of the hand and wrist: A case report☆

Muge Kirac [a,*,1], Abdullah Burak Kara [b,2], Halil Ibrahim Bulut [c,2], Gulcin Basdemir [d,2], Danyal Gumus [e,f,2,3], Korhan Ozkan [e,f,2]

[a] Acibadem Mehmet Ali Aydinlar University, School of Medicine, Department of Orthopedics and Traumatology, Istanbul, Turkey
[b] Istanbul Medeniyet University Prof. Dr. Suleyman Yalcin City Hospital, Orthopedics and Traumatology, Istanbul, Turkey
[c] Istanbul University-Cerrahpasa, School of Medicine, Istanbul, Turkey
[d] Istanbul Galata University, Faculty of Dentistry, Pathology Department, Istanbul, Turkey
[e] Acibadem Atasehir Hospital, Department of Orthopedics and Traumatology, Istanbul, Turkey
[f] Acibadem Atasehir Hospital, Department of Orthopedic Surgery, Istanbul, Turkey

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ABSTRACT

Background: Schwannomas, which are also known as neurilemomas, constitute the most common type of peripheral nerve tumors. These tumors can grow slowly and occur as painless swellings over the course of several years before their diagnosis. The aim of this report is to present a case of a very rare plexiform schwannoma originating from second digital nerve and multiple schwannomas from third and fourth digital nerves and palmar cutaneous branch of median nerve with emphasis given on differential diagnosis and treatment strategy.
Case Presentation: A 15-year-old patient presented with soft tissue masses on the volar surface of the hand, initially diagnosed as benign nerve sheath tumors. Surgical excision was performed twice, preserving nerves and tendons. Recurrence occurred, and subsequent biopsy confirmed benign nerve sheath tumors. Microscopic examination revealed multinodular/plexiform schwannoma in the larger lesion and simple schwannomas in others, leading to a diagnosis of multiple schwannomatosis. All surgeries resulted in intact sensory and motor function.
Conclusion: Surgical treatment can be curative and effectively employed for concomitant schwannoma tumors. Nevertheless, with careful planning and execution, surgery remains a promising option for patients with concomitant schwannomas. Further research and long-term follow-up studies are needed to fully understand the outcomes and refine the techniques used in these surgical treatments.

Introduction

Schwannomas, which are also known as neurilemomas, constitute the most common type of peripheral nerve tumors. It was first described by Verocay in 1908, while the name "Schwannoma" was subsequently established by Masson in 1923. They are originated from the Schwann cells of the nerve sheath and characterized by their benign nature and well-defined encapsulation [1]. These tumors grow slowly and occur as painless swellings over the course of several years before their diagnosis. They can reach a size of 4 cm prior to the onset of symptoms caused by nerve compression. Occasionally, malignant transformations have been documented. They reach their highest incidence between the third and sixth decades and are distributed equally among men and women. Typically, they are solitary; however, there have been instances of

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* Correspondence to: Kayışdağı Cad. No:32 Ataşehir, İstanbul, Turkey.
E-mail address: mugekirac@gmail.com (M. Kirac).
[1] https://orcid.org/0009–0005-5615–7087

[2] The author, their immediate family, and any research foundation with which they are affiliated have not received any financial payments or other benefits from any commercial entity related to the subject of this article.

[3] https:o//orcid.org/0000–0002-8985–8219

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multiple forms and have demonstrated a possibility of recurrence [2]. In this report we aimed to present a case of a very rare plexiform schwannoma originating from second digital nerve and multiple schwannomas from third and fourth digital nerves and palmar cutaneous branch of median nerve with emphasis given on differential diagnosis and treatment strategy.

Ethical Considerations

This study adhered to the ethical principles outlined in the Declaration of Helsinki. All participants provided informed consent prior to their inclusion in the study, ensuring they were fully aware of the nature, benefits, and potential risks of the procedures. Institutional Review Board (IRB) approval was waived, and patient confidentiality was maintained throughout the study.

Case

A 15-year-old patient was admitted to our hospital in 2018 due to a mass that had been growing on the volar surface of the hand for the last one year. The MRI examination revealed soft tissue lesions measuring 8 $\times$ 10 mm in diameter located near the flexor tendons at the carpal tunnel level of the patient. Additionally, there were solid soft tissue lesions measuring approximately 5 cm in diameter with a multiloculated

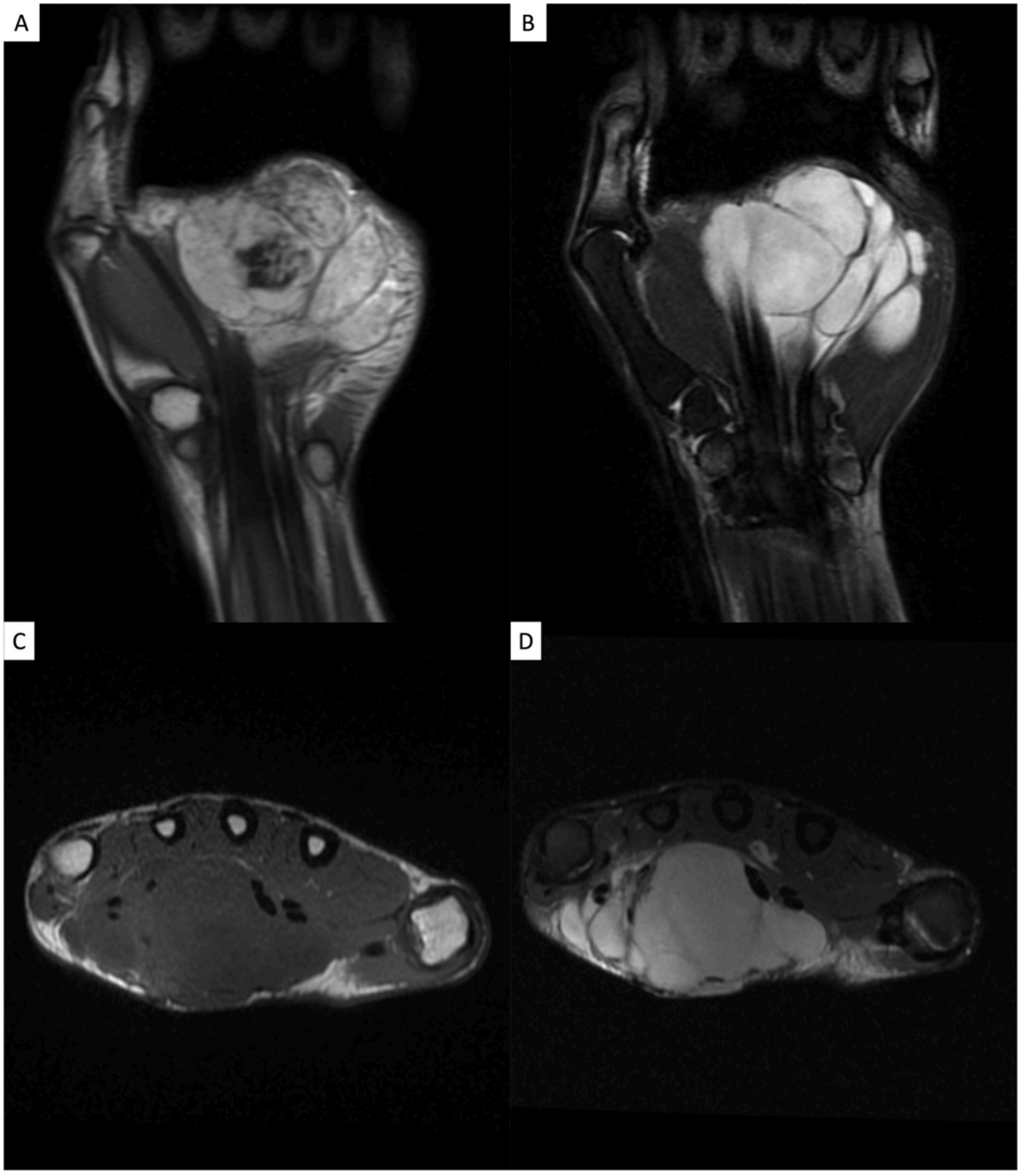

Fig. 1. (A) Coronal T1-weighted MRI of the left hand showing a heterogeneous tumor in the plam with mixed singnal intensity. (B) Coronal T2-weighted MRI of the left hand demonstrating a well-circumscribed tumor with high signal intensity. (C) Axial T1-weighted MRI of the left hand illustrating the tumor with intermediate signal intensity relative to surrounding tissues. (D) Axial T2-weighted MRI of the left hand highlighting the tumor with high signal intersity,consistent with fluid content or a cystic nature.

septate appearance in the palmar region. Thus, lesions originating from the neural sheath were initially considered in the differential diagnosis. The identified lesions were situated in the interstitial space between the flexor tendons and were observed to cause displacement of the tendons in certain areas. A biopsy was performed through a mini open incision and a diagnosis was made as a benign nerve sheath tumor. Afterwards a volar incision was made to resect the soft tissue masses that had originated from the digital branches of the median nerve on the volar site of hand, at the 3rd and 4th metacarpal levels. The nerves were carefully preserved during the resection procedure, and the mass underwent pathological examination, resulting in a diagnosis of multiple schwannomas.

During the second year following the surgical procedure, a new recurrent lesion is identified on the volar surface of the hand, and surgery is recommended to the patient again. However, the patient had refused from attending any further medical examinations until the year 2023. Prior to the most recent surgical procedure, the presence of soft tissue masses located on the volar surface of the hand and wrist was identified. An MRI examination displayed a large, solid soft tissue lesion measuring 72 × 70 mm between the craniocaudal extension surrounding the flexor tendons in the hand, palm, and palmar face. The lesion causes pressure on the tendons, leading them to disperse and expand. A second lesion displaying similar features was identified immediately proximal to the wrist, in close proximity to the flexor tendon groups, with a size of 16.5 mm in longitudinal direction. (Fig. 1).

Subsequently, an open biopsy was conducted, yielding a result that correlates with the findings of the previous biopsy, indicating the presence of a benign nerve sheath tumor. Surgical tumor excision under general anesthesia was done with preserving of the second, third and fourth digital nerves with all tendons and lumbrical muscles. Second incision was done to remove the mass lesion just proximal to wrist. Ultimately, no nerve was compromised with intact postoperative sensory and motor function in the wrist and all fingers. (Fig. 2).

In gross examination, encapsulated mass with shiny tan color in cut surface, composed of neighboring connecting nodules of variable sizes was seen (45 ×50 mm) with many small juxtaposed but connected lesions. Microscopically, the big lesion consisted of spindle cells forming alternating hyper- and hypocellular areas, in a collagenized and focally myxoid background. There was no significant cellular atypia, aberrant mitotic activity, or necrosis. Immunohistochemical study revealed S100 (+), CD34(+), EMA(-), claudin4(-) expression profile of the tumor. Given these findings, multinodular/plexiform schwannoma was diagnosed. However, all other lesions including the one located in the wrist were consistent with simple schwannomas and the diagnosis of multiple schwannomatosis was reached [3].

Discussion

Schwannoma, perineurioma and neurofibroma are some of the major categories of benign nerve sheath tumor classification. Although the variable anatomical distribution tendencies affect the success of their surgical removal, these tumors have similar prognostic outcomes with their benign clinical behavior and low recurrence rates. According to Rockwell et al.'s research, 7.5 % of all cases of schwannoma occur in the hand and wrist [4]. Among all schwannomas, plexiform schwannomas account for up to 5 % of all schwannomas. Plexiform schwannomas are a rare benign proliferation of Schwann cells that presents as a multinodular growth pattern (plexiform) and only 0.7 % of all cases of schwannoma are characterized by the presence of multiple schwannomas [5].

Schwannomas exhibit slow growth and lack infiltrative properties, resulting in infrequent occurrence of nerve dysfunction. Thus, they have the ability to remain without symptoms for an extended duration as in our case. Schwannomatosis, also known as congenital neurilemmomatosis, is a rare syndrome defined by the development of multiple schwannomas of the peripheral nervous system and with no involvement of the vestibular nerves [6,7].

Ultrasonography (USG) and MRI are valuable for distinguishing between different types of soft tissue masses. Schwannoma can be mistakenly identified as neuroma, lipoma, or ganglion [4]. USG differentiates between solid or cystic tumors and determines the size of the tumors. Schwannoma exhibits a uniform, low-echoic, and pseudocystic appearance on USG, similar to that of a ganglion cyts. However, MRI is the optimal imaging technique for accurately identifying the characteristics, position, and proximity of lesions to anatomical structures, facilitating diagnosis and surgical planning. Schwannomas are characterized by their appearance on MRI, where they exhibit high signal intensity on T2 scans and low signal intensity on T1 scans, as in our case. Malignant peripheral nerve sheath tumors (MPNST), such as malignant schwannoma and neurofibrosarcoma, and benign peripheral nerve sheath tumors (BPNST) exhibit similar characteristics when observed using MRI. The preoperative diagnosis can pose challenges and may result in misdiagnosis so preoperative biopsy is utmost importance in suspected cases with respect to size and growth pattern [4]. Due to its fast-growing characteristics and size an open biopsy was performed in our case.

The main significance in the differential diagnosis of these tumors, which may overlap clinically and histomorphologically, is that they can occur in the setting of tumor predisposition syndromes, of which, neurofibromatosis and schwannomatosis make the classical examples. Hybrid benign peripheral nerve sheath tumors, which were introduced

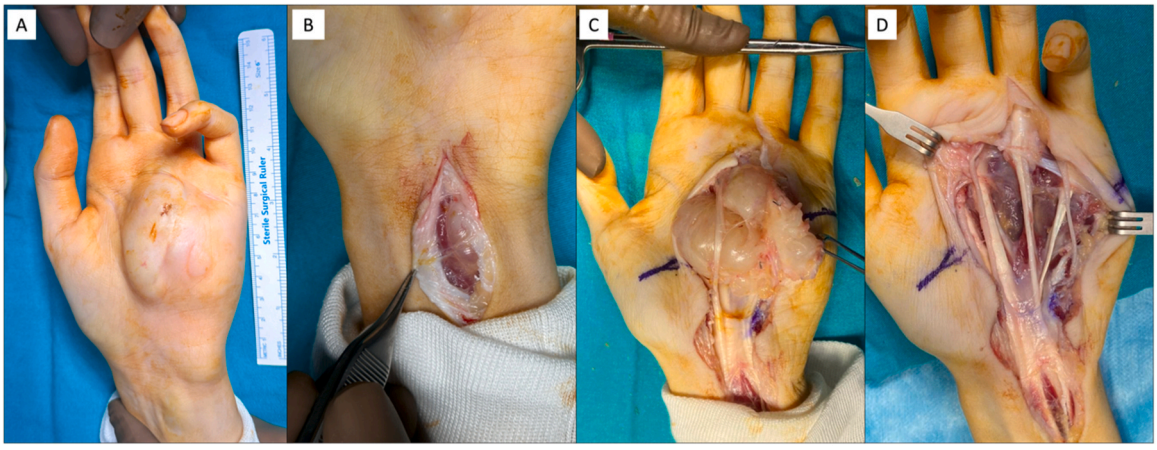

Fig. 2. (A) Preoperative view showing the patient's left hand with a palpable mass in the plam.(B) Initial incision over the palmar mass, exposing the underlying tissue. (C) Dissection and exposure of the mass, revealing its encapsulated nature. (D) Post-excision view showing the preservation of digital nerves, tendons, and lumbrical muscles.

to the literature through the use of immunohistochemical and ultrastructural studies, are another increasingly recognized category that consists of components that differentiate into more than one of these formerly mentioned tumors. Our case, in addition to typical schwannomatous areas, also showed areas of parallel arranged tumor cells rather than haphazard, necessitating the evaluation of a hybrid tumor with some degree of perineurioma component. However, immunohistochemical findings, with diffuse and strong staining of S-100 and complete absence of expression for the perineural differentiation markers EMA and claudin-4, were diagnostic of a pure schwannoma, and a hybrid tumor was excluded [3].

According to our assessment, intact expression of INI1/SMARCB1 (which is partially or completely lost in certain types of schwannomatoses) and because the nodules do not show discontinuity was suggestive of multinodular/plexiform schwannoma in the big lesion. However, different lesions juxtaposed to the gross lesion and the one in the wrist were also consistent with schwannomatosis. So, our case was unique due to the diagnosis of multinodular plexiform and schwannomatosis which is extremely rare in the English-speaking literature [8].

Radiosurgery is an advanced technological intervention increasingly utilized in the treatment of various solid tumors and metastases, including vestibular schwannomas [9–11]. Despite its growing application, peripheral nerve schwannomas present unique challenges that typically preclude the use of radiosurgery [10]. These challenges include their anatomical location, the frequent occurrence of multiple tumors, and the complexity of their localization which can hinder the effectiveness of radiosurgery [11].

In our case, the decision to employ radiosurgery was influenced by the distinctive location and multiple nature of the tumor. Thence, we successfully managed the case using conventional surgery. This approach enabled us to preserve both nerve functions and tendons, demonstrating the potential for conventional surgery to effectively treat complex schwannomas while maintaining critical physiological functions.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Data availability

No data was used for the research described in the article.

References

[1] Milenković S, Mitković M. Common peroneal nerve schwannoma. Hippokratia 2018;22(2):91.
[2] Kang HJ, Shin SJ, Kang ES. Schwannomas of the upper extremity. J Hand Surg Edinb Scotl 2000;25(6):604–7.
[3] Meyer A, Billings SD. What's new in nerve sheath tumors. Virchows Arch 2020;476 (1):65–80.
[4] Rockwell GM, Thoma A, Salama S. Schwannoma of the hand and wrist. Plast Reconstr Surg 2003;111(3):1227–32.
[5] Ha C, Kang YK, Ha JW, Han DH, Lee JK, Han SH. Segmental schwannomatosis in the upper extremity: a case report and review of the literature. Arch Hand Microsurg 2021;27(1):93–8.
[6] Chen S lin, Liu C, Liu B, Yi C jun, Wang Z xin, Rong Y bo, et al. Schwannomatosis: a new member of neurofibromatosis family. Chin Med J (Engl) 2013;126(14): 2656–60.
[7] Lee SH, Kim SH, Kim BJ, Lim DJ. Multiple schwannomas of the spine: review of the schwannomatosis or congenital neurilemmomatosis: a case report. Korean J Spine 2015;12(2):91–4.
[8] Bhaumik S, Johnson T, Gali V. A case of multiple plexiform schwannomas. Cureus 2022;14(8):e28140.
[9] Lim M, Adler JR. CyberKnife radiosurgery for extremity schwannomas: technical note and case report. Stereotact Funct Neurosurg 2006;84:60–3.
[10] Yao C, Zhou H, Dong Y, et al. Malignant peripheral nerve sheath tumors: latest concepts in disease pathogenesis and clinical management. Cancers 2023;15:1077.
[11] Pellerino A, Verdijk RM, Nichelli L, et al. Diagnosis and treatment of peripheral and cranial nerve tumors with expert recommendations: an european network for rare cancers (EURACAN) initiative. Cancers 2023;15:1930.