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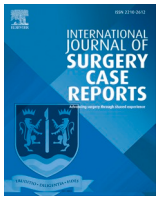

Case report

Recurrent schwannoma of digital nerve on both hands: A very rare case report

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ABSTRACT

Introduction and importance: Schwannoma is the most common benign tumor of peripheral nerves and usually occurs solitary with a very low risk of recurrence. Schwannoma of the hand, particularly involving the digital nerve, is sporadic and most commonly leads to a misdiagnosis due to its rarity. Histopathology remains the gold standard diagnostic, however, with a thorough physical examination and radiologic imaging, this benign tumor can be managed well with the preservation of nerve function.
Case presentation: We reported a rare case of digital nerve schwannomas on the bilateral hands of a 52-year-old white male with one recurrent mass on the right hand despite a previously complete debulking of the mass.
Clinical discussion: Intraoperatively, we removed lobulated whitish-yellow masses and left the nerve origin of the tumor intact (common palmar digital nerve and ulnar-side digital nerve). The histopathology supported the diagnosis of schwannoma. At the one-year follow-up, there is no recurrence and the patient remains asymptomatic.
Conclusion: Determining differential diagnosis by both clinical and preoperative imaging is essential, especially in the case of recurrence and multiple tumorous lesions. Malignancy or malignant degeneration should still be kept in mind. Complete removal is needed to prevent recurrence followed by a long-term follow-up.

1. Introduction

Schwannomas are benign tumors of the peripheral nerve sheath, arising from the Schwann cells. It is considered the most common form of peripheral nerve sheath tumor. However, its occurrence is rare (only accounts for less than 5 % of all soft tissue tumors) and mostly encountered during the third and sixth decades of life [1,2]. Clinical identification and treatment of schwannomas is challenging since these tumors can manifest as a painless growth for years before they are discovered. When tumors grow large enough to compress a nerve, pain, paresthesia, and other symptoms may develop [3]. Schwannomas share clinical symptoms and MRI results with other soft tissue tumors, a definitive diagnosis will only be made through the microscopic investigation with histopathologic sampling and immunohistochemistry (IHC) examination [4]. The presence of schwannoma in multiple areas is rare (only 10 % of cases). Furthermore, the recurrence or malignant differentiation of this benign lesion is extremely rare. We report a very rare case of digital nerve schwannoma on the bilateral hand with a recurrent mass that was treated in our center with complete tumor removal. The patient had been observed for 1-year follow-up without evidence of recurrence. Informed consent has been taken from the patient prior to the study and publication process. This case is reported according to the Surgical Case Report (SCARE) Guidelines [5].

2. Presentation of case

A 52-year-old male foreign photographer from the United States visited the outpatient clinic in our center with a slow-growing and painless swelling in the palmar area of the left hand and right middle fingers (Fig. 1). No numbness, tingling, or functional deficit was complained of by the patient. There was no history of any traumatic events or infection preceding the swelling. There was a history of mass on the same site on the right middle finger 22 years ago before the current hospital admission. It was removed 7 years after its first appearance and the diagnosis of schwannoma was confirmed. The mass reappeared in the same location 15 years ago. Another painless swelling appeared 7

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years ago on the left palm. No family history of similar symptoms.

On the physical examination, two palpable masses that shared the same characteristic were found on both hands. The masses were mobile, non-pulsating, and soft in consistency. The size of the mass were 20 mm × 10 mm and 15 mm × 10 mm on the left hand and the right middle finger respectively (Fig. 1). No tenderness was noted on palpation and the Tinel sign was negative. Sensory examination was normal. Revealed MRI of both hands (Fig. 2) showed a well-defined, strongly delineated, heterogeneously hyperintense lesion and other signs that were suggesting a peripheral nerve tumor (split fat sign, fascicular sign, bull's eye sign).

Excisional biopsy was performed under general anesthesia. Careful dissection under surgical loupe magnification was done to preserve the adjacent neurovascular bundle. A complete en bloc excision was achieved, leaving the nerve origin of the tumor intact (common palmar digital nerve of the left hand and ulnar digital nerve of the right middle finger). Lobulated whitish-yellow masses sized 20 mm × 10 mm (left) and 15 mm × 10 mm (right) were sent for histopathological examination (Fig. 3).

The histopathology examination showed multi-lobulated, plexiform, well-encapsulated masses that are characteristic of a benign lesion. The diagnosis of schwannoma was further suggested by the appearance of typical Verocay bodies (Fig. 4). After the surgery, the patient healed uneventfully. There was no complaint of tingling or numbness during the immediate post-surgical period and no evidence of recurrence at the 1-year follow-up. We still closely observe the patient because of the history of recurrence in this case.

3. Discussion

Schwannoma represents less than 5 % of all soft tissue tumors and its occurrence in the hand is rare. In the case of the upper extremity schwannoma, the ulnar nerve is the most common nerve involved, and the median nerve is the second most commonly involved. The occurrence of this lesion on the digital nerve is extremely rare and only reported in several literatures: Troy et al. [6], Pertea et al. [7], Han KJ et al. [8], Aleksić et al. [9], and Nicolescu et al. [10].

The etiology of schwannoma is poorly understood and appears to have a chromosomal pathology that causes mutations in SMARCBI and LZTR1 which are tumor suppressors genes for Schwan cell proliferation [7,10]. There is no gender preference and this lesion was reported within the age range of 30–60 years old. In our case, the patient is a 52-year-old man which is supported by the evidence of mean age reported by Pertea et al. [7] (58.5 years), Majumder et al. [11] (48 years), and Hirai et al. [12] (56 years). This finding suggested that the older age group might have a higher risk of schwannoma occurrence.

Schwannoma is typically asymptomatic. Thus, this slow-growing mass has a large differential diagnosis that makes a clinical diagnosis not always straightforward. At the early stage, it is often confused with ganglion cyst, lipoma, fibroma, neurofibroma, and xanthoma. It has an extremely slow rate of growth before it grows large enough and causes symptoms such as pain, paresthesia, and tingling. A Tinel sign along the course of the nerve may be positive in symptomatic patients (positive predictive value of 87.5 %). This sign is present in 4 % to 76 % of schwannoma. Moreover, the presence of major neurological signs such as pain, paresthesia, and tingling sensation suggests an unfavorable post-operative outcome. In our patient, despite the history of recurrence and a long onset of growth, there were no neurological problems and the Tinel sign was negative [1,7]. Thus, a good prognosis can be expected.

In patients that are presented with more than one lesion, differentiating schwannoma from neurofibromatosis is especially crucial. Schwannomas are often presented as an isolated lesion, non-intradermal, and appear on the flexor surface of the upper extremity. Multiple schwannoma is rare and only reported in 9–13 % of cases. Neurofibromatosis is generally seen in the subcutaneous tissue and on superficial layers of the body. However, its occurrence may also be associated with neurofibromatosis type 2 (NF2) in 3 % of cases [4,10]. In this presenting case, the masses were fairly mobile and the clinical diagnosis of schwannoma was made based on the physical examination and the histopathological result of the previously excised mass.

Diagnostic imaging, such as plain radiographs, ultrasounds, and especially MRIs, may be helpful in establishing a correct diagnosis. Although they do not have 100 % accuracy, they are potent non-invasive modalities for localizing the tumors, particularly to investigate the relationship between the tumor and the neurovascular bundle (Ultrasonography and MRI) which will be helpful in planning the surgery. Calcification (10 % of schwannomas), the split fat sign, and secondary osseous alterations such as bone scalloping can all be seen on plain radiographs. Ultrasonography (USG) may show a homogenous well-defined hypoechoic mass and its relationship with the associated nerve. This type of presentation is commonly misdiagnosed with ganglion cysts (38 % of cases) because they share a similar location, size, and pedunculated base. MRI remains the most useful diagnostic imaging that shows an isointense mass with the same intensity as the muscle in the T1-weighted image. Moreover, in digital nerve schwannoma, a subtle ring of fat can be seen at the inferior and superior margin of the mass (the split fat sign) in the T1-weighted image. In the T2-weighted image, schwannoma appears bright with a heterogeneous

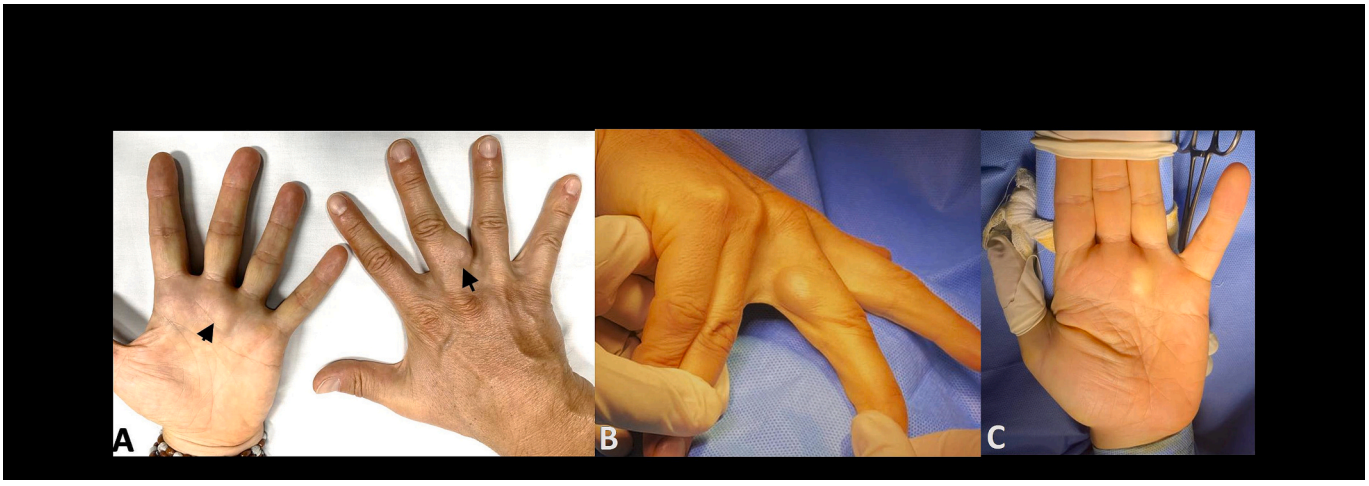

Fig. 1. (A) Swelling on both hands (black arrow). (B) Mass at the dorso-ulnar aspect of the right proximal phalanx. (C) Mass at the volar aspect of the left hand distal to the distal palmar crease between the 3rd and 4th digit.

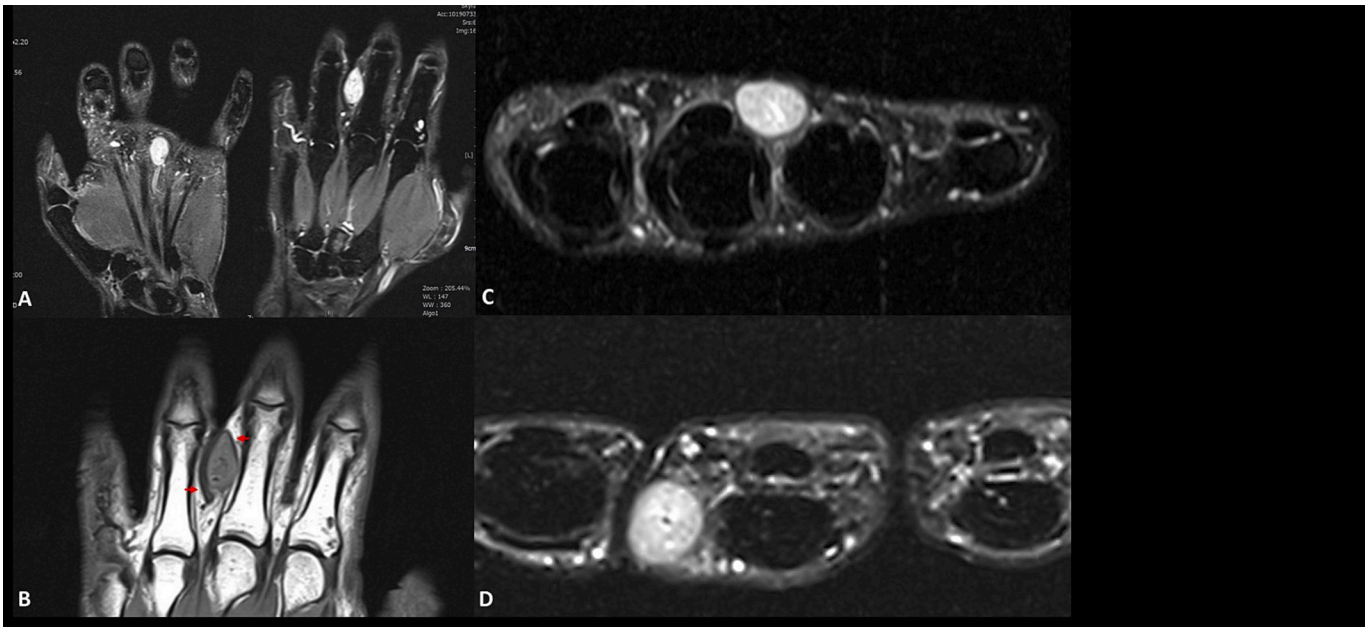

Fig. 2. (A) Magnetic resonance imaging (T2-weighted) of the right hand and left hand showed well-defined, sharply demarcated, heterogeneously hyperintense lesions. (B) A split fat sign (the thin peripheral rim of fat) was noted (red arrows) on the coronal view of T1-weighted image. (C & D) Multiple small ring-like structures with peripheral hyperintensity (fascicular sign) were noted within the mass on the axial view. A low signal intensity core surrounded by hyperintensity (fascicular sign) on figure d represents "bulls eye sign".

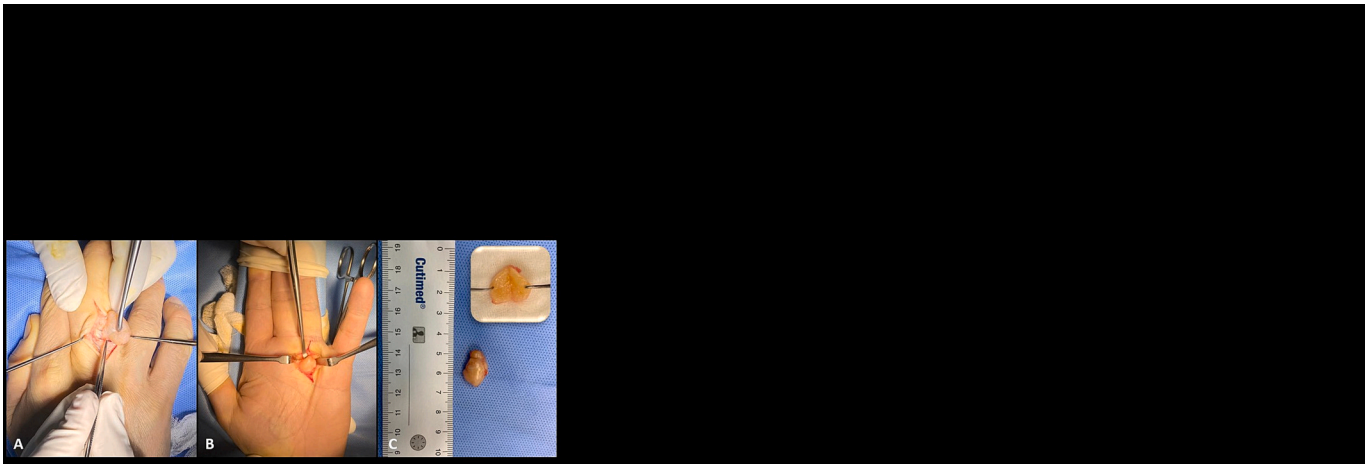

Fig. 3. Intraoperative findings of the masses on (A) right hand, and (B) left hand. (C) Whitish-yellow ovoid masses were sent for histopathology after successfully being enucleated.

hyperintensity. A lower intensity signal at the core of the mass (bull's eye or target sign) may also be observed in schwannoma and in another peripheral nerve sheath tumor, neurofibroma [2,7,10]. In our case, the MRI findings strongly supported a peripheral nerve sheath tumor diagnosis. Although there is a minimal risk of malignancy progression in these slow-growing, encapsulated tumors, the malignant peripheral nerve sheath tumor (MPNST) can even mimic this benign lesion on MRI [10]. Hence, the possibility of the malignant form should always be considered before it is proven otherwise.

The treatment options for schwannoma can include both surgical and nonoperative approaches. Non-operative management is an option for patients with no or minimal symptoms. However, for a larger and symptomatic lesion, surgery is indicated. Surgical excision and biopsy remain the most effective way of diagnosis and therapy while preserving nerve function. A meticulous dissection under magnification can achieve en-bloc tumor removal. The encapsulated nature of the mass made the dissection easier. A complete dissection also lowered the risk of recurrence. Enucleation is more easily accomplished with schwannoma than with neurofibroma. Neurofibromas are thought to arise in the endoneurium and may not be as successfully separated from the peripheral nerve without injuring nerve fascicles. However, if the nerve is damaged during excision, reconstruction with grafts or conduit is advisable to preserve the function. Adherent mass to the adjacent structure without clear evidence of a capsule should raise suspicion of malignancy. In that case, the nerve should be transected and reconstructed [6,8]. In our case, a complete en-bloc resection was done to both masses. The associated nerves seemed intact intraoperatively and no post-operative paresthesia was experienced.

The histopathological and immunohistochemical (IHC) examinations are the gold standard diagnosis for schwannoma. Microscopically, two distinct areas of proliferation can be seen which are the hypercellular area (Antoni A) and the hypocellular area (Antoni B). In the

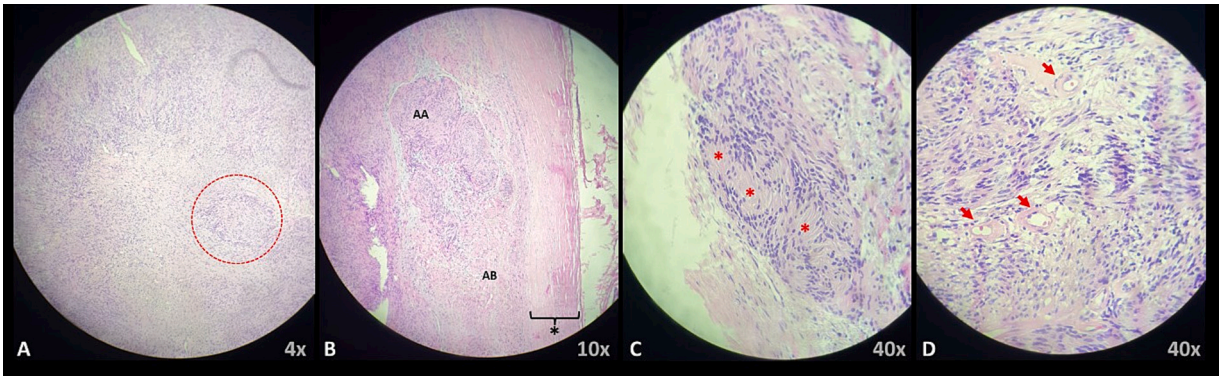

Fig. 4. Histopathologic findings of schwannoma of the digital nerve on the hematoxylin-eosin (HE) staining. (A) Fascicle (red dots). (B) Capsule (black asterisk), areas of hypercellularity/Antoni A (AA) and hypocellularity/Antoni B (AB). (C) Verocay bodies: Areas of nuclear palisading around fibrillary process (red asterisk). (D) Vessels with hyaline walls (red arrows).

hypercellular area, Verocay bodies that are formed from the palisading nucleus surrounding the clear fibrillary processes can be seen. IHC examination of schwannoma shows a positive for the S100 staining that is an important differentiation with the neurofibroma [7]. In our patient, histopathology examination with HE staining showed strong evidence of schwannoma.

After a definitive treatment, post-operative malignant transformation and recurrence of schwannoma are rare [3,7,9]. However, despite a meticulous resection under magnification, iatrogenic nerve injury may sometimes occur. Oberle et al. [13] reported immediate postoperative sensory deficits in six of 12 patients. Kim SL et al. [14] study also showed that an immediate neurological deficit was seen in 76.7 % of patients. Some known pre-operative risk factors for post-operative nerve complications are larger tumor size, more prolonged onset, and preoperative Tinel's sign. One or more fascicles that run through the tumor's body may be at risk during dissection. Postoperative neurological deficits were found to be closely associated with the failure of enucleation and intraoperative axonal injury. The transection of fascicles that run through the tumor is believed to be the major cause of postoperative neurological deficits [14].

After one year of post-operative observation, none of the above complications occurred. The sensitivity of the associated fingers was normal both pre-operative and post-operative. However, we still observed the patient annually to detect recurrence or the development of new lesions.

4. Conclusion

This case illustrates an unusual presentation of schwannoma (multiple, recurrence, and arising from the common and the collateral digital nerve). Although the diagnosis of schwannoma has been established in previous surgery, determining a preoperative differential diagnosis and valuing preoperative imaging is equally important, while malignancy should still be kept in consideration. A complete removal based on thorough preoperative planning should be done aiming for a painless and fully sensate hand with no recurrence.

Disclaimer

No patient or author details are included in the figures.

Consent for publication

The patient provided informed consent for the case details and accompanying images to be published.

Ethical approval

None. Because this paper is reporting a case which is consisted only one patient and not considered as human research. Thus, it does not typically require IRB review and approval.

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CRediT authorship contribution statement

All authors contributed to data analysis, drafting or revising the article, have agreed on the journal to which the article will be submitted, gave final approval of the version to be published, and agree to be accountable for all aspects of the work.

Conflict of interest

None declared.

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