Intracapsular Micro-Enucleation of a Painful Superficial Peroneal Nerve Schwannoma in a 60-Year Old Man: A Rare Encounter

Abbas A. Ramadan
Yousef A.I. Abousedu
Omar M. Ghanem
Salem A.M. Alsubiee
Aisha Y. AlAbkal
Hussain H. Jarkhi
Gazi A. Matar

Patient: Male, 60-year-old
Final Diagnosis: Right superficial peroneal nerve schwannoma
Symptoms: Lump on the lateral aspect of the right upper leg • pain
Medication: —
Clinical Procedure: Intracapsular micro-enucleation of the lesion
Specialty: Neurosurgery

Objective: Rare disease
Background: Schwannomas are the most common benign peripheral nerve sheath tumors, localized mainly to the cranial and upper extremity nerves. Their occurrence in the lower limbs is uncommon, and specific involvement of the superficial peroneal nerve is exceedingly rare. We report a case of a painful right superficial peroneal nerve schwannoma that was excised via the intracapsular micro-enucleation technique.

Case Report: A 60-year-old South Asian man presented with a 2-year history of a painful lump on the lateral aspect of the right upper leg. Clinical examination revealed a firm mass located at the proximal lateral aspect of the right leg, measuring approximately 3×2.5 cm. Severe tenderness over the mass was present. The Tinel test was positive. There were no sensory or motor deficits or history of neurofibromatosis. Imaging showed features suggestive of a schwannoma. Surgery was indicated; intracapsular micro-enucleation was performed. Histopathological assessment of the tumor demonstrated Antoni A and B patterns with nuclear palisading and Verocay bodies, hallmarks of a schwannoma. The postoperative period was uneventful; no neurological deficits were noted.

Conclusions: The case described is considered rare, with no data on disease epidemiology in the literature. We provide a brief review and add pivotal data to the literature. Despite its rarity, one should remain cognizant of the condition and consider it in the differential diagnosis of nontraumatic leg pain. Based on our experience, corroborated from previous case reports, and the satisfactory outcome of our case, we advocate the intracapsular micro-enucleation technique when possible for schwannomas.

Keywords: Middle East • Neurilemmoma • Peripheral Nervous System Neoplasms • Peroneal Nerve • Schwannomatosis

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/936056
Schwannomas, also known as neurilemmomas, neuromas, or Schwann cell tumors, are benign, well-capsulated tumors that arise from Schwann cells of the peripheral nerve sheath [1]. It is the most common benign peripheral nerve sheath tumor (PNST), localized mainly to the cranial and upper extremity nerves [1,2]. Its manifestation on lower limb nerves is uncommon and accounts for 1% of all cases, with specific involvement of the superficial peroneal nerve being exceedingly rare [3-6]. In fact, after an extensive search of the literature, there were no available data on the occurrence of the superficial peroneal nerve, further emphasizing its rarity worldwide.

Several Middle Eastern countries have reported lower limb schwannomas in the common peroneal, deep peroneal, and tibial nerves, which are the most common locations for this tumor to arise from [7-10]. To date, there have been scarce cases of superficial peroneal nerve schwannomas in the literature, with none reported from the Middle East [3,11-15]. In contrast to other published case reports on the same condition, we elaborated and provided comprehensive illustrations on the radiological, surgical, and pathological findings of the case, which is a major distinguishing feature. Here, the authors present a case in Kuwait of a 60-year-old man with a painful right superficial peroneal nerve schwannoma that was excised via the intracapsular micro-enucleation technique.

Case Report

A 60-year-old South Asian male farmer with no medical history presented to the neurosurgical clinic with a chronic painful lump on the lateral aspect of the right upper leg of a 2-year duration. The lump preceded the pain. There was a 2-year evolution in symptomatology (progressive growth and pain). Over the last 3 months, the lump had become excruciatingly painful. The patient stated that the pain was located at the site of the lump with distal radiation to the right big toe. The pain was described as “stabbing” in nature and was aggravated by walking, crouching, leg-crossing, and touching the lump. There were no pain-relieving factors. He denied any history of trauma and lumps elsewhere in the body. The family history was absent for neurofibromatosis.

Clinical examination revealed a firm, minimally mobile mass located at the proximal lateral aspect of the right leg, measuring approximately 3×2.5 cm. The mass was non-fluctuant and non-pulsatile and did not transilluminate. There were no overlying skin changes. Severe tenderness over the mass was present. There were no sensory or motor deficits. The pain was reproduced with passive and active (with and without resistance) dorsiflexion, plantarflexion, and eversion. The Tinel test was positive, eliciting severe pain radiating to the right big toe. No other masses were detected.

Results of investigations including routine blood tests, D-dimer, and Doppler ultrasound were unremarkable. Motor and sensory nerve conduction studies revealed right superficial peroneal sensory neuropathy with no electrophysiological evidence of motor fascicle involvement and no generalized large fiber polyneuropathy. Further evaluation was performed by magnetic resonance imaging (MRI) of the right leg with and without intravenous contrast (Figure 1). MRI showed a well-defined, oval-shaped, heterogeneously enhancing mass measuring 3.2 cm in length and 2 cm in width, located just lateral to the fibula at the proximal third of the right leg, and exhibiting T1 isohypointensity and T2 hyperintensity, respectively. The findings were suggestive of superficial peroneal nerve schwannoma.

Surgery was indicated, and we performed intracapsular enucleation of the lesion under microscopy, with images taken during surgery (Figure 2). During surgery, the well-defined true capsule that envelopes the schwannoma, which consists of the perineurium of the nerve bundle of origin surrounded by the deep layers of the epineurium, was incised far from any visible nerve stems under microscopy. The tumor was then exposed, dissected from the adjacent fascicles, and enucleated while safeguarding adjacent structures, without apparent fascicular injury noted after lesion excision. The epineurium of the superficial peroneal nerve was then sutured and layer to layer closure was achieved.

The excised lesion measured 3.5×2.0×1.0 cm and was sent for histopathological assessment, which included the use of special stains. A conclusive diagnosis was made on the basis of histopathological examination, which demonstrated features that were pathognomonic of a schwannoma as illustrated (Figure 3).

The postoperative period was uneventful, and the patient reported significant improvement in his symptoms. He followed up in the neurosurgical clinic 2 weeks after surgery, where he was ambulating comfortably with no pain and no motor or sensory deficits.

Discussion

There have been very few cases of superficial peroneal nerve schwannomas reported worldwide across different continents, with no reports yet on disease epidemiology in the literature. This case raises awareness of that fact that even though lower limb schwannomas are uncommon, PNST should always be considered as part of the differentials of any patient presenting with nontraumatic leg pain. It also reinforces the use of...
the intracapsular micro-enucleation technique for schwannomas, which has been supported by several studies discussed throughout this report.

Schwannomas can be present in various locations of the peripheral nervous system, but up to 45% of cases occur in the head and neck region, followed by the upper limb nerves [16]. The occurrence in the leg or in the foot and ankle region is rare [11]. Involvement of the sciatic nerve has been reported to be the most common location for lower limb schwannomas, accounting for approximately 50% of cases [17]. They are solitary in 90% of cases; multiple tumors should raise the suspicion of syndromic associations (neurofibromatosis type 2, schwannomatosis, and carney complex) [1]. Schwannomas usually affect individuals between the ages of 50 to 60 years, with no sex or racial dominance [1].

Growth of these tumors is slow, and they never traverse through the nerve; remaining in the sheath overlying it [3,18]. This explains why symptomatology onset, such as severe pain, can appear years later, which occurred in our case, or can present as a clinically silent incidental finding. In fact, there can be an approximate 5-year delay between symptoms and diagnosis [1]. In contrast, other documented cases of superficial

Figure 1. Magnetic resonance imaging (MRI) of the patient’s right leg. A well-defined, oval shaped lesion visible just lateral to the fibula at the proximal third of the right leg. (A) Iso-hypointense signal on axial T1-weighted image without contrast (asterisk). (B) Heterogeneous enhancement on T1-weighted image with intravenous contrast. (C) Sagittal view on T1-weighted image with contrast showing lesion (arrow) arising from the superficial peroneal nerve. (D) Clear visualization of the lesion measuring 2 cm in width on proton-density sequence.
peroneal nerve schwannomas reported symptom durations of 6, 12, and 36 months [3,11-14]. Further analysis of all the scarce reports on superficial peroneal nerve schwannomas revealed that the ages of the patients ranged between 40 and 65 years, and the most common presenting symptoms were pain and a lump [3,11-15]. Sex distribution was also equal in these cases, with 3 female and 3 male patients. Diagnosis was achieved via MRI in most of the cases except 2, which interestingly used only ultrasonography to establish the diagnosis [13,15]. Our current case was not dissimilar in terms of demographics, presenting symptoms, and choice of diagnostic modality to most of the aforementioned reported cases.

A systematic-based approach to evaluating patients with non-traumatic leg pain is recommended and involves a detailed history and clinical examination, laboratory tests (especially D-dimer), followed by ancillary studies, including several
imaging modalities and nerve conduction studies. MRI has been reported to be the preferred imaging technique in the diagnosis of tumors of the peripheral nervous system [19-21]. Electrophysiological studies before surgery may be of limited use, and their application during surgery is more critical as it helps to identify and preserve the fascicles in relation to the tumor, thereby significantly reducing risk of neurological injury during tumor resection [17].

The criteria for surgery of benign PNST include the following: progressive increase in size of the swelling, increase in pain, and worsening neurological deficit [17]. Tumor size alone is never a criteria for surgical intervention [17]. In the present case, owing to the unbearable nature of the pain, surgery was advised. Surgical techniques used for schwannomas can range from subtotal excision (50% or more) to gross total excision (>90% of the tumor), leaving behind the tumor capsule adherent to the fascicles (ie, intracapsular excision) to complete

Figure 2. Sequential perioperative findings. Surgical excision of patient’s right upper leg tumor under microscopy. (A) Antero-lateral approach. Tumor borders defined via clinical examination and a horizontal skin incision marked. (B) Gross visualization of the lesion. (C) Microscopic visualization of the lesion. (D) Superficial peroneal nerve fascicles identified (asterisks) and was attached to the tumor (arrowhead). (E, F) Tumor exposition, dissection and enucleation performed. (G) Superficial peroneal nerve (asterisk) after excision of the tumor, with capsule left behind and epineurium sutured. (H) Gross appearance of the tumor measuring 3.5×2.0×1.0 cm.

Figure 3. Histopathological assessment of the lesion under microscopy. (A) Biphasic pattern composed of hypercellular (Antoni A, arrow) and hypocellular (Antoni B, arrowhead) components that are arranged in intersecting fascicles. Blood vessels have thickened hyalinized walls (asterisks) (hematoxylin and eosin [H&E] stain, ×10) (B) Nuclear palisading is prominent (asterisks). Formation of Verocay bodies which consists of 2 layers of palisading spindle cells with an anuclear zone in between (arrows). There was no evidence of mitosis, necrosis, nuclear atypia, or pleomorphism (H&E stain, ×20). (C) Immunohistochemical staining for S100 was strongly and diffusely positive in the lesional cells [S100 stain, ×40].
Excision of the proximal and distal nerve stems of the tumor, including its capsule (ie, extracapsular excision) [17].

Extracapsular enucleation is one of the operative techniques used in schwannoma surgery [22,23]. However, by removing the capsule that is adherent to the fascicles, the risk of fascicular injury and postoperative neurological deficit is higher [23-25]. For that reason, in our case, we used the intracapsular microscopic enucleation technique with en bloc resection, as this has been shown to be superior to the extracapsular excision technique in terms of providing satisfactory results with a low risk of nerve injury and postoperative neurological deficits [18,23,24,26]. This technique has also been performed on a similar reported case of superficial peroneal nerve schwannoma with satisfying outcomes [3]. It was also proposed that en bloc resection is not advised and instead intracapsular piecemeal micro-enucleation (“tumor resected piece by piece”) is considered safe and reliable since the main purpose of schwannoma surgery is for symptomatic relief rather than tumor excision [24]. This technique has also been supported by another large study involving 422 schwannoma cases, which preferred to excise large tumors by piecemeal debulking [17].

Schwannomas can be completely excised without major loss of function and without recurrence. However, meticulous dissection using microscopy is required in large-sized schwannomas, as was done in the present case, since the risk of fascicular injury and neurological deficit after surgery seems to be higher in these cases [23,27]. Intraoperative electrophysiological monitoring may also be crucial in preventing neurological deficits [28]. Surgical removal of peripheral nerve tumors should be performed by a surgeon with microsurgical and peripheral nerve experience to minimize serious iatrogenic nerve injury and postoperative morbidity.

Classic schwannoma, which was seen in the current case, is the most common type, but several variants exist and include ancient, cellular, melanotic, and plexiform schwannomas [1,29]. The histopathological appearance of classical schwannomas is described as having a “biphasic pattern” with 2 distinct histological regions: Antoni A tissue is characterized by pericellular spindle cells, that can form palisading rows with an acellular zone in-between (Verocay bodies); and Antoni B tissue shows a hypocellular pattern with a background of loose connective tissue [1,29]. A well-formed collagenous capsule and hyalinized vessels are also consistent findings [29]. Immunohistochemical staining for S100 protein is a useful marker to identify neoplasms derived from Schwann cells and was used in the present case to further support the diagnosis [30]. Malignant transformation of benign schwannoma is uncommon, but they can occur and account for 5% to 10% of all soft-tissue sarcoma [31].

The prognosis is excellent after adapted surgical management with sparing of nerve fascicles [1]. Recurrence of schwannomas after total excision is rare, but most importantly, patients should be thoroughly counseled before surgery on the potential occurrence of neurological deficits [32].

Conclusions

Schwannomas are benign PNSTs, which rarely ever occur on the superficial peroneal nerve. With this rare case reported from the Middle East region, we aimed to share our experience, contribute further data to the literature, and consolidate the paucity of literature available worldwide. Lower limb schwannomas are overlooked, but despite their rarity, one should remain aware of the condition and consider it as part of the differential diagnosis in patients presenting with nontraumatic leg pain. Based on our experience with this case and corroboration from previous case reports, we highly advocate the intracapsular micro-enucleation technique. Further studies are necessary to investigate the epidemiology of the disease in different cohorts.

Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

1. Sheikh MM, De Jesus O. Schwannoma. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2021.
2. Knight DM, Birch R, Pringle J. Benign solitary schwannomas: A review of 234 cases. J Bone Joint Surg Br. 2007;89(3):382-87
3. Nascimento G, Nomi T, Marques R, et al. Ancient schwannoma of superficial peroneal nerve presenting as intermittent leg pain: A case report. Int J Surg Case Rep. 2015;6C:19-22
4. Sharig O, Radha S, Konan S. Common peroneal nerve schwannoma: An unusual differential for a symptomatic knee lump. BMJ Case Rep. 2012;2012:br2012007346
5. Patil S, Dinesh Babu K, Reddy S, et al. Schwannoma of the superficial peroneal nerve in 12-year-old female child: A case report. Open J Orthop. 2014;4(7):189-93
6. Tantia H, Ashok D, Mariappan K, et al. A rare case of schwannoma of the superficial peroneal nerve with split fat sign. East African Scholars J Med Sci. 2020;31(1):399-401
7. Sharma RR, Pawar SP, Dey P. An occult schwannoma of the deep peroneal nerve presenting with neuralgia mimicking sciatica: Case report and review of the literature. Ann Saudi Med. 2000;20(1):57-59
8. Öz TT, Aktaş B, Özkan K, et al. A case of schwannoma of the common peroneal nerve in the knee. Orthop Rev (Pavia). 2017;9(1):6825
9. Abdulnasser AW, Khalef M. Schwannoma – a cause of common peroneal nerve neuropathy. A case report. J Trop Nephrol Urol. 2004;22(10):10-13
10. Komurcu E, Golge UH, Kaymaz B, Erdogan N. Polleitale schwannoma mimicking baker cyst: An unusual case. J Surg Case Rep. 2013;2013(8):r0066
11. Laurencin CT, Bain M, Yue JI, Glick H. Schwannoma of the superficial peroneal nerve presenting as web space pain. J Foot Ankle Surg. 1995;34(6):532-33
12. Maselli F, Testa M. Superficial peroneal nerve schwannoma presenting as lumbar radicular syndrome. J Back Musculoskelet Rehabil. 2019;32(2):361-65
13. Trăistaru R, Enăchescu V, Manuc D, et al. Multiple right schwannoma. Rom J Morphol Embryol. 2008;49(2):235-39
14. Nkaoui M, Sasbou Y. [Entrapment neuropathy in the foot revealing schwannoma of the superficial peroneal nerve: outcome of conservative surgical treatment.] Pan Afr Med J. 2017;28:161 [in French]
15. Pajazetovic A, Dahukey A. A case of schwannoma of the lateral aspect of the superficial peroneal nerve. J Am Podiatr Med Assoc. 2021;111(2):Article_17
16. Abreu I, Roriz D, Rodrigues P, et al. Schwannoma of the tongue – a common tumour in a rare location: A case report. Eur J Radiol Open. 2017;4:1-3
17. Desai KI. The surgical management of symptomatic benign peripheral nerve sheath tumors of the neck and extremities: An experience of 442 cases. Neurosurgery. 2017;81(4):568-80
18. Ozdemir O, Ozsoy MH, Kurt C, et al. Schwannomas of the hand and wrist: Long-term results and review of the literature [published correction appears in J Orthop Surg (Hong Kong). 2006 Aug;14(2): following 229]. J Orthop Surg (Hong Kong). 2005;13(3):267-72
19. Murphey MD, Smith WS, Smith SE, et al. From the archives of the AFIP. Imaging of musculoskeletal neurogenic tumors: Radiologic-pathologic correlation. Radiographics. 1999;19(5):1253-80
20. Cerofolini E, Landi A, DeSantis G, et al. MR of benign peripheral nerve sheath tumors. J Comput Assist Tomogr. 1991;15(4):593-97
21. Ansari I, Ansari A, Graison AA, et al. Head and neck schwannomas: A surgical challenge – a series of 5 cases. Case Rep Otolaryngol. 2018;2018:4074905
22. Li X, Zhong G, Xu K, et al. Surgical strategies for peripheral nerve schwannoma based on the intraoperative neurophysiological monitoring. Laparoscopic and Robotic Surgery. 2019;2(3):65-69
23. Date R, Muramatsu K, Ihara K, Taguchi T. Advantages of intra-capsular micro-enucleation of schwannoma arising from extremities. Acta Neurochir (Wien). 2012;154(1):173-78
24. Takase K, Yamamoto K, Imakiire A. Clinical pathology and therapeutic results of neurilemmoma in the upper extremity. J Orthop Surg (Hong Kong). 2004;12(2):222-25
25. Sooraj T, Shanmugasundram S, Ambujam G, et al. Rare sites of benign nerve sheath tumours: (Median nerve, radial nerve and peroneal nerve schwannoma). Int J of Surg. 2019;31(1):259-61
26. Kim SM, Seo SW, Lee JY, Sung KS. Surgical outcome of schwannomas arising from major peripheral nerves in the lower limb. Int Orthop. 2012;36(8):1721-25
27. Raj C, Chantelot C, Saab M. Predictive factors of postoperative deficit and functional outcome after surgery for upper limb schwannomas: Retrospective study of 21 patients. Hand Surg Rehabil. 2020;39(3):229-34
28. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: Diagnostic overview and update on selected diagnostic problems. Acta Neuropathol. 2012;123(3):295-319
29. Stefansson K, Wollmann R, Jerkovic M. S-100 protein in soft-tissue tumors derived from Schwann cells and melanocytes. Am J Pathol. 1982;106(2):261-68
30. Albert P, Patel J, Badawy K, et al. Peripheral nerve schwannoma: A review of varying clinical presentations and imaging findings. J Foot Ankle Surg. 2017;56(5):632-37
31. Furniss D, Swan MC, Morriss DG, et al. A 10-year review of benign and malignant peripheral nerve sheath tumors in a single center: Clinical and radiographic features can help to differentiate benign from malignant lesions. Plast Reconstr Surg. 2008;121(2):529-33