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GIANT SCHWANNOMA OF THE LEG – A RARE CASE REPORT

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ABSTRACT
Schwannoma is a benign tumour of the peripheral nerve sheath originating from the Schwann cells that grows slowly eccentric to the nerve axis and is a rare occurrence in the lower limb. Peripheral nerve sheath tumours are common neoplasm with different biological features ranging from benign to malignant. They are mostly misdiagnosed as benign soft tissue skin condition, such as lipoma, myxoma, or ganglion cyst. Most of these tumours are less than 5 cm in size, whereas those larger are termed giant schwannomas. Malignant transformation of a schwannoma is rare. The usual symptoms of schwannoma of the lower limb are tingling, burning sensations signifying peripheral neuropathy and weakness, paralysis of the affected limb denoting motor impairment. Magnetic resonance imaging and biopsy are the most useful diagnostic methods followed by surgical excision, which is the treatment of choice. Postoperative complications are minimal and rare. Here, we report a case of a giant schwannoma and its management by surgical excision and the resultant defect resurfaced with a pedicled flap.

KEYWORDS: Benign, Nerve Sheath, Tumour, Giant Schwannoma, Surgical Excision

INTRODUCTION

Peripheral nerve sheath tumours originate from nerve sheath cells and are classified as benign or malignant according to World Health Organization (WHO) criteria. The benign tumours of peripheral nerves include schwannoma, neurofibroma and perineuroma, the first two being the most common. (1-3) Schwannomas are rare benign tumours arising from peripheral nerve sheaths and are encapsulated by the epineurium arising from Schwann cells of the peripheral nerve sheath. (4) They are predominantly localized to the head and neck region, the sacral plexus, the brachial plexus, and the sciatic nerve. Schwannoma occurring in the lower limbs is extremely rare, comprising only 1% of schwannoma cases. (5,6) They usually often resemble a ganglion cyst, Morton's neuroma, lipoma, or vascular malformation. (7) The tumours are generally asymptomatic due to slow growth and adaptation of the nerve fibre to the space occupied by the tumour within nerve sheath. Total resection of tumour after careful intraneural dissection results in good outcomes as the recurrence is low. Malignant transformation of schwannomas is extremely rare. (8)

Case Report

64 year old male swelling in left lower leg for about 20 years. He was apparently normal 20 years back when he noticed a small swelling in the left lower leg, insidious in onset and gradually progressive to attain the current size. He gives history of pain for 1 month which was pricking type and non-radiating. There is no history of trauma, ulceration or bleeding from the swelling. There was no history of comorbid illnesses. On examination, a swelling of size 6 x 4cm was present in the left lower limb 5cm above the left lateral mallelolus. The surface was smooth, skin was pinchable with restricted mobility and firm in consistency. (Fig. 1) There was no significant regional lymphadenopathy. MRI showed a large well defined lobulated T1 iso and T2/ STIR heterogenously hyperintense solid lesion showing mild restricted diffusion and thin internal septations, deep to superficial fascia in anterolateral aspect of left lower leg between anterior and lateral compartment muscles causing mass effect on anterior compartment muscles of leg displacing medially, with the lesion is abutting lateral aspect of fibula and anterior aspect of interosseous membrane suggestive of a sarcoma. (Fig. 2) Ultrasound-guided tru-cut biopsy revealed fragments of neoplasm showing hypercellular and hypocellular areas and the neoplastic cells being spindle-shaped with minimal pleiomorphism, elongated nuclei and scanty cytoplasm. The neoplastic cells are focally arranged in fascicles and forming Verocay bodies, with no evidence of necrosis or increased mitotic activity suggestive of a benign spindle cell neoplasm. The patient was planned for wide local excision with flap cover. Under combined spinal epidural anaesthesia, an incision made around the swelling with a 1cm margin and deepened in layers. The tumour was seen infiltrating into periosteum of fibula, the periosteum was removed using periosteum elevator and the lesion was excised in toto. (Fig. 3) Haemostasis was achieved and the patient was handed over to plastic surgery team.

The defect size was 11 x 10 cm on the distal third of left leg exposing the underlying tendons and fibula. Peroneal artery perforator propeller flap was planned. The peroneal perforator was marked with hand held Doppler, planning in reverse done, and the flap was marked. Tourniquet was applied and flap elevated. The perforator was identified and found to be in good quality, flap elevated, propelled into defect and inset done with 2-0 nylon. Donor site raw area was reduced and covered with SSG and dressing applied. (Fig. 4) Histopathological examination revealed a well-circumscribed neoplasm composed of hypercellular (Antoni A) and myxoid hypocellular (Antoni B) areas. The hypercellular area comprises spindle cells having elongated wavy nuclei with tapered ends arranged in fascicles.

Nuclear palisading around the fibrillary process (Verocay bodies), hyalinization, and areas of haemorrhage with hemosiderin deposition were also noted. (Fig. 5) The margins are free of neoplasm. Immunohistochemistry studies diffuse strong cytoplasmic and nuclear positivity in 100% of tumour cells (S-100), focally membranous positivity in 30% of cells with moderate intensity (SMA) and Ki67 labelling index of 2% which are in favour of spindle cell neoplasm of neural origin.

Fig. 1 – Clinical photograph of the lesion

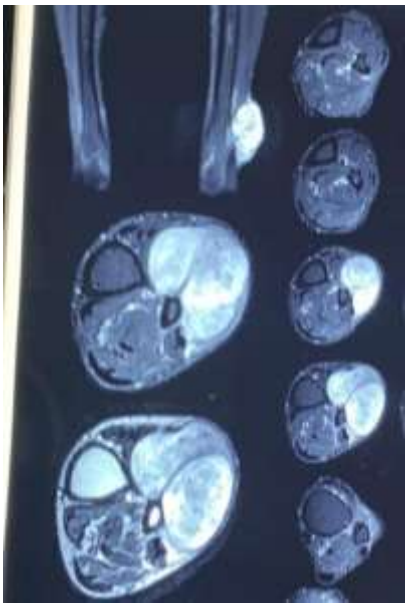

Fig. 2 – MRI of the lesion

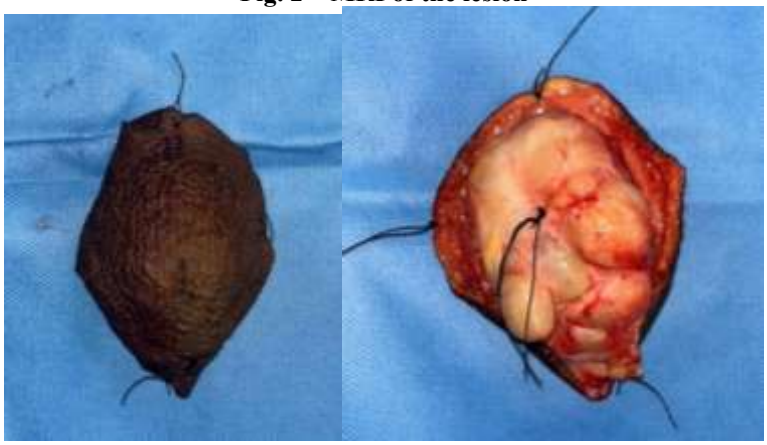

Fig. 3 Photograph of the specimen

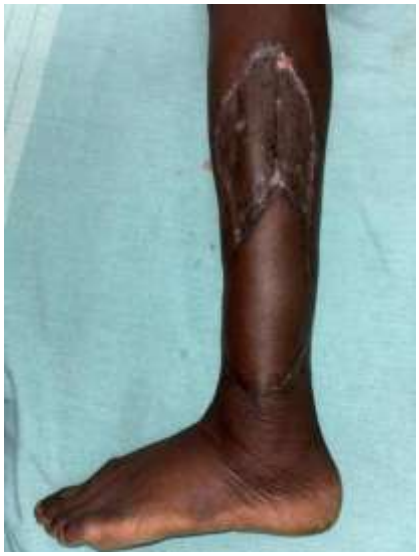

Fig. 4 – Flap & SSG well settled

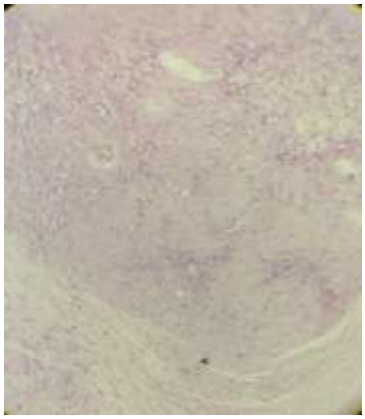

Fig. 5 – Histopathology of the lesion

DISCUSSION

Schwannomas, also known as neurilemmomas, are the most common peripheral nerve sheath tumours that derived almost entirely from Schwann cells, responsible for myelination of the peripheral nerve fibres. (9) They represent approximately 5% of all benign soft tissue tumours and are the most common peripheral nerve tumours. (8,10) Schwannomas most commonly present between the second and fifth decade of life. There is no gender or racial predilection. (11) Schwannomas are solitary, isolated, well-capsulated and sporadic in 90 % of all cases with 10 % of schwannomas occur at multiple sites. These cases are considered systemic due to genetic abnormalities in conditions such as neurofibromatosis type 2, Schwannomatosis, and Carney complex. (12) Those larger than 5 cm are referred to as giant schwannomas. (13) They are typically encapsulated, well-defined tumours with biphasic patterns. The capsule of schwannomas is comprised of three layers: the epineurium, the perineurium from the nerve bundle of origin, and the pericapsular tissue that may contain nerve fibres. The tumour capsule is often covered by tortuous blood vessels. Axons are not usually present within the tumour, which allows schwannomas to be removed without damages to the underlying nerve fascicles. (14)

They most commonly arise in the distribution of the eighth cranial nerve in the head and neck region, but their presence on the lower limb is exceptionally rare. Peripheral schwannomas are mostly asymptomatic due to their slow-growing nature, but symptoms arise due to compression of adjacent nervous structures. (15,16) From nerve origin, schwannomas are classified into three types. The first type originates in the major nerve located intermuscularly which can be symptomatic. The second is from the minor superficial sensory branch located subcutaneously. The third originates from bundles of the intramuscular motor nerve. The latter two types are usually asymptomatic. This classification system is useful for preoperative diagnoses and could inform the choice of operative procedures as surgical excision of the major nerve type may cause neurological deficit. (17,18) Under the microscope and consist of two components: Antoni A areas, which are densely packed with spindle-shaped cells forming palisades or Verocay bodies, and Antoni B areas, which are more loosely arranged with hypocellular myxoid stroma. (19) Immunohistochemistry shows expression of S-100 protein. Degenerative changes that characterize an ancient schwannoma include interstitial hyalinization, cyst formation, calcification on and haemorrhage, along with degenerative nuclear atypia, but without any mitotic activity. (6,8,20)

MRI is important in assessing the tumour's size, edema and invasion of surrounding structures. (15) Schwannomas typically appear as a well-circumscribed, encapsulated mass, most commonly up to 5cm, and enhanced with contrast administration. (21) Tumors often demonstrate an eccentric mass near the affected nerve in a well-defined capsule with homogenous isointense or hypointense on T1-weighted images and high intensity on T2-weighted images. Intramuscular mass may be surrounded by fat that can create the “split fat sign” on T1-weighted images on the long axis of the affected limb. The “target signs” is noticed in 50 % of cases which has central low-intensity signal and peripheral high-intensity signal due to zonal effect from centrally localized fibrous tissue and peripherally localized myxoid material. (8)

The preferred choice of treatment for schwannomas arising in the extremities is complete surgical excision, even if the tumour has not spread out of the capsule. (22) There are two main approaches in resecting schwannomas: extra-capsular and intra-capsular enucleation. However, recent studies reported neurological deficit in nearly 70 % of cases. (23) Some researchers have suggested a modified microsurgical technique with palliative, intra-capsular enucleation, which is safer for nerve fascicles and minimizes complications. (24) Intra-capsular excision has been reported to reduce the neurological complication compared to extra-capsular excision with major deficit observed in only 3 % of cases. Postoperative outcome and prognosis of the tumour are overall good, aside from minor residual weakness and neurologic symptoms, which disappear over time. The rate of recurrence or malignant transformation of the tumour is low. (25)

CONCLUSION

Schwannomas are the most common peripheral nerve sheath tumours that derived from Schwann cells, and constitute about 1% of all schwannoma cases, therefore very rare. Ultrasound and MRI are valuable for initial diagnosis and evaluation but only a histopathological analysis can provide a definitive diagnosis. The treatment of choice for schwannoma is surgical enucleation or resection without damaging the involved nerve. The prognosis of peripheral schwannoma is favourable after surgical excision since it is a benign, localized tumour, with minimal post-operative complications.

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