NEURILEMMOMA: A Case Study

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INTRODUCTION

Neurilemmoma is the most common neurogenic tumor that can present in any nerve throughout the body. They are benign, encapsulated tumors of the nerve sheath. Their cells of origin are thought to be Schwann cells derived from the neural crest. Within the foot and ankle, the benign lesion essentially manifests itself as a palpable mass with symptoms similar to compressive neuropathy. Neurotologic symptoms tend to present during the late stages. Symptoms can range from being asymptomatic to chronic neuropathic pain.^{1,2} The cause of these neoplasms is unknown. Neurilemmoma can be associated with von Recklinghausen disease; when this is the case, multiple tumors often are present.^{3,4}

CLINICAL PRESENTATION

Neurilemmomas generally affect persons between the age of 20-50 years. There is no racial or sex predilection. Common locations for the tumors are the head and flexor surfaces of the upper and lower extremities, and the trunk. The mass is usually mobile within the center of the nerve.³

Tenderness to palpation is often present; secondary neurologic symptoms may occur if the tumor is large. Lesions in the sciatic nerve can mimic a herniated-disc within the lower-back. Lesions within the tibial nerve of the foot can mimic tarsal tunnel syndrome.⁵ The differential diagnoses include: fibroma, neurofibroma, neurosarcoma, ganglion cyst, giant cell tumor of tendon sheath, and lipoma.

Neurilemmomas can cause a functional deficit because of local pressure on the nerve. The mass can increase in size and can cause permanent nerve damage. Malignant degeneration, which is extremely rare, was described by Yousem and colleagues in 1985. Primary malignant tumors of this cell type do exist, but they are histologically distinct from neurilemmomas.

Because these tumors can present in many locations, the clinical presentation can be varied. Some may involve the spinal nerve roots and present with symptoms that mimic those of herniated disk disease of the spine. In the foot and ankle, neurilemmomas can present either as an asymptomatic mass or as localized pain and paresthesia resulting from pressure on the nerve.

The masses are slow growing and can exist for months to years without producing symptoms. The average time from onset of symptoms to diagnosis is 5.5 years. In an unusual case in which resection would lead to a significant functional deficit, these benign lesions can be observed.

DIAGNOSTIC PROCEDURES

Neurilemmomas are radiolucent, therefore plain radiographs will generally be nonspecific. The rare intraosseous lesion presents as a benign appearing, well-circumscribed lesion. Laboratory studies are generally not beneficial. Computer imaging, computed tomography (CT) and magnetic resonance imaging (MRI), are useful when evaluating neurilemmomas.⁶ When there is no bony destruction evident on plain radiographs, an MRI is more useful than CT when evaluating a neurilemmoma. It is generally recommended to perform a gadolinium contrast, which will help differentiate the neurilemmoma from fluid-filled cysts. On MRI, a neurilemmoma is usually round or oval with a moderately bright signal on T1-weighted images.⁷

STAGING

Neurilemmomas are generally graded using the Enneking system, which grades benign lesions ranging from 1 to 3. Grade 1 lesions are inactive, grade 2 lesions deform the surrounding tissues but are not destructive or locally aggressive, and grade 3 lesions are locally aggressive and may invade local tissues but do not have a metastatic potential. Generally, neurilemmomas are either grade 2 or 3.²

TREATMENT

As with most benign tumors, neurilemmomas respond well to local resection. On inspection, usually the nerve is splayed out over the lesion. The lesion is excised marginally, and the nerve fibers are spared. Interlesional resection is warranted when complete resection would result in permanent neurologic deficit. The most common complication is initial neuropraxia; however, this neurologic deficit can be permanent, depending on the resection of neurilemmoma. Generally, patients tolerate resection well, with complete and rapid relief of symptoms. Recurrence is unlikely following complete resection. Rare descriptions exist of malignant change in long-standing neurilemmomas, usually in patients with an underlying diagnosis of neurofibromatosis. Malignant change is extremely rare in isolated lesions.^{1,2}

CASE STUDY

A 54-year-old woman presented with pain on the plantar aspect of the left foot. She complained of some mild numbness on the bottom of her foot. She especially noticed pain during deep massage of her foot. She denied any trauma to her foot and her past medical history was unremarkable.

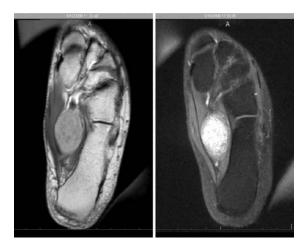


Figure 1. MRI axial view, T1-weighted (left) and T2-weighted (right).

Plain radiographs were negative and an MRI of her left foot demonstrated a large oval mass within the tibial nerve (Figures 1-3). Due to the significant size of the mass and intimate relationship to the tibial nerve, surgical excision was recommended.

A 12 cm curivilinear incision was made along the medial aspect of the foot (Figure 4). The plantar muscles were reflected plantarly (Figure 5), which exposed the soft tissue mass and tibial nerve. The soft tissue mass was encapsulated and mass was intertwined within the tibial nerve (Figure 6). Blunt dissection was performed to remove the soft tissue mass in total (Figure 7). A fresh frozen study was performed and the soft tissue mass was identified as a neurilemmoma (Figure 8). Standard closure was performed (Figure 9). A nonweight-bearing cast was applied for 4 weeks, followed by 2 week partial weight bearing with a fracture boot. At 8 weeks, full activity was resumed. The patient did not develop any permanent nerve damage.



Figure 2. MRI coronal view, T1-weighted (left) and T2-weighted (right).

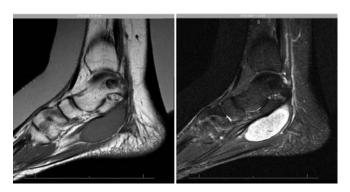


Figure 3. MRI sagittal view, T1-weighted (left) and T2-weighted (right).



Figure 4. Curvilinear medial incision.



Figure. 5. Plantar reflection of muscles.



Figure 6. Neurilemmoma intertwined within tibial nerve.



Figure 7. Excision of neurilemmoma.



Figure 8. Neurilemmoma approximately 5 cm x 2.5 cm.



Figure 9. Layered closure.

REFERENCES

- Downey MS. Gredlein CM. Soft Tissue Masses. In Banks AS, Downey ME, Martin DE, Miller SJ. McGlamry's Comprehensive Textbook of Foot and Ankle surgery. Philadelphia: Lippincot Williams & Wilkins; 2001. p. 1358.
- Campanacci M. Bone and Soft Tissue Tumors: Clinical Features, Imaging, Pathology and Treatment. 2nd ed. New York (NY): Springer-Verlag; 1999.
- 3. Stull MA, Moser RP, Kransdorf MJ, et al. Magnetic resonance appearance of peripheral nerve sheath tumors. *Skeletal Radiol* 1991;20:9-14.
- Woodruff JM, Selig AM, Crowley K, et al. Schwannoma (neurilemoma) with malignant transformation. A rare, distinctive peripheral nerve tumor. *Am J Surg Pathol* 1994;18:882-95.
- 5. Belding RH. Neurilemoma of the lateral plantar nerve producing tarsal tunnel syndrome: a case report. *Foot Ankle* 1993;14:289-91.
- 6. Cerofolini E, Landi A, DeSantis G, et al. MR of benign peripheral nerve sheath tumors. *J Comput Assist Tomogr* 1991;15:593-7.
- Mikami Y, Hidaka T, Akisada T, et al. Malignant peripheral nerve sheath tumor arising in benign ancient schwannoma: a case report with an immunohistochemical study. *Pathol Int* 2000;50:156-61.