**Final Diagnosis:**

Schwanomma of the posterior tibial nerve
**Definition:**
Tumor of the nerves sheath arising from schwann cells

CN schwannomas are usually isolated lesions, except when they are associated with neurofibromatosis.

Malignant schwannomas almost never seen.
**Frequency:**

**In the US:** Schwannomas account for 6-8% of intracranial neoplasms. Autopsy studies have shown that the incidence rates of occult vestibular schwannomas are as high as 2.7%. A study of patients undergoing MRI for indications other than the evaluation of schwannoma revealed an estimated prevalence of 0.07%.

**Vestibular schwannomas** are the most common CN schwannomas, followed by trigeminal and facial schwannomas and then glossopharyngeal, vagus, and spinal accessory nerve. Schwannomas involving other cranial and peripheral nerves are rare.
Mortality/Morbidity:

Morbidity resulting from schwannomas includes nerve dysfunction and brainstem compression. Mortality can result from mass effect with brainstem compression.
**Race:**
No racial predilection has been described in schwannomas.

**Sex:**
No sex predilection has been described in schwannomas.
**Clinical Presentation:**

Mostly **asymptomatic solitary mass**

Typically, presenting symptoms of schwannomas are based on the affected nerve.
Investigations:

MRIs is the golden standard for initial diagnosis or initial diagnosis is the golden standard for initial diagnosis.

USIs another method but rarely used.

Immuno-histopathology: schwannomas are highly reactive for S-100 protein.
Management

Surgical excision

Stereotactic radiosurgery (ie, gamma knife radiosurgery) largely has replaced surgical resection for the treatment of vestibular schwannomas.

Other CN schwannomas also can be treated with radiosurgery.
<table>
<thead>
<tr>
<th>TUMOR site</th>
<th>Number of cases</th>
<th>Distribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Post. Tibial N.</td>
<td>4</td>
<td>USA (2001)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Spain (1990)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>France (1989)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Italy (1982)</td>
</tr>
<tr>
<td>Ant. Tibial N.</td>
<td>1</td>
<td>France (1993)</td>
</tr>
<tr>
<td>Plantar branches of post. tibial N.</td>
<td>4</td>
<td>Japan 2 cases (2004)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>USA (2001)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Italy (1981)</td>
</tr>
<tr>
<td>Tibial N.</td>
<td>3</td>
<td>Turkey (2001)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>USA 2 cases (1992, 1991)</td>
</tr>
<tr>
<td>Nerve Type</td>
<td>Count</td>
<td>Country</td>
</tr>
<tr>
<td>-------------------------------</td>
<td>-------</td>
<td>------------------</td>
</tr>
<tr>
<td>Deep peroneal N</td>
<td>1</td>
<td>Oman (1999)</td>
</tr>
<tr>
<td>Sciatic N.</td>
<td>2</td>
<td>Greece (2005)</td>
</tr>
<tr>
<td>Brachial plexus</td>
<td>1</td>
<td>USA (1991)</td>
</tr>
<tr>
<td>Spinal accessory N.</td>
<td>2</td>
<td>Italy (1992), China (1990)</td>
</tr>
<tr>
<td>Suprascapular N.</td>
<td>1</td>
<td>Oman (2001)</td>
</tr>
<tr>
<td>C1 &amp; C2</td>
<td>42</td>
<td>(1982-1992)</td>
</tr>
<tr>
<td>Sacral</td>
<td>25</td>
<td></td>
</tr>
<tr>
<td>T12 to L2</td>
<td>1</td>
<td>Canada (1997)</td>
</tr>
<tr>
<td>TUMOR site</td>
<td>Number of cases</td>
<td>Distribution</td>
</tr>
<tr>
<td>----------------------------</td>
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<td>----------------</td>
</tr>
<tr>
<td>L5</td>
<td>1</td>
<td>Japan (1996)</td>
</tr>
<tr>
<td>Intraosseous (L4)</td>
<td>1</td>
<td>China (1998)</td>
</tr>
<tr>
<td>Cauda Equina</td>
<td>1</td>
<td>Japan (1991)</td>
</tr>
</tbody>
</table>
## Malignant schwannomas

<table>
<thead>
<tr>
<th>TUMOR site</th>
<th>Number of cases</th>
<th>Distribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median N. with mets.</td>
<td>1</td>
<td>Germany (1988)</td>
</tr>
<tr>
<td>Medial plantar branch of post.tibial N.</td>
<td>1</td>
<td>USA (1990)</td>
</tr>
<tr>
<td>Intraosseous epitheloid schwannoma</td>
<td>1</td>
<td>2007</td>
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<tr>
<td>Endobronchial mets from malignant schwannoma</td>
<td>1</td>
<td>(UK) 1984</td>
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OBJECTIVE AND IMPORTANCE: Neoplasms of peripheral nerves can be obscured, especially during the early phase. The author reports a patient with a posterior tibial nerve neurilemoma (schwannoma). For a decade, the tumor was misdiagnosed as nonspecific S1 radiculopathy and psychogenic chronic pain syndrome. The patient's presentation and initial management are unique. CLINICAL PRESENTATION: A 40-year-old woman reported severe left foot and calf pain, numbness, and weakness. The symptoms were evident during three pregnancies, and they gradually progressed. The neuropathic pain was protracted, despite implantation of a dorsal column stimulator and administration of a wide variety of medications and therapies. The symptoms were unresponsive to both inpatient and outpatient treatments, which resulted in a misdiagnosis of psychogenic pain for more than a decade. Diagnostic scans obtained by computed tomography, ultrasonography, and nuclear scintigraphy confirmed a popliteal fossa mass. INTERVENTION: A high, large posterior tibial nerve neurilemoma was found intraoperatively, positioned just below the sciatic nerve bifurcation with extensive degenerative features and hemorrhages. Surgical resection provided immediate recovery. CONCLUSION: Peripheral nerve tumors are rarely acknowledged clinical entities. Chronic unexplained foot and calf pain and a positive Tinel's sign should raise suspicion of posterior tibial nerve neurilemoma. Even in patients who have had such tumors for a decade, surgical resection remains the treatment of choice.
Radiology Department, Hospital de La Princesa, Madrid, Spain. (1990)

Tibialis posterior nerve schwannoma mimicking Achilles tendinitis
Two cases of deep-seated neurilemmoma that arose from plantar branches of the posterior tibial nerve and caused chronic heel pain are described. At the initial examination, one case was misdiagnosed as tarsal tunnel syndrome and the other was overlooked as plantar fasciitis; both cases were treated for long periods prior to operation. Deep-seated neurilemmomas in the foot can easily be overlooked and misdiagnosed as tarsal tunnel syndrome or plantar fasciitis because of the rarity, absence of palpable mass, and similarity of symptoms to those of other frequently encountered foot disorders. Magnetic resonance imaging provides the best modality for differential diagnosis. In the present cases, surgical excision of the tumors resulted in immediate and complete relief of chronic heel pain. Surgeons should consider neurilemmoma as a cause of persistent chronic heel pain despite the rarity of the disease.
The incidence of neurilemoma in the foot is uncommon. Neurilemoma of the medial plantar nerve distal to the tarsal tunnel, with symptoms isolated to the foot, is rare. A case presentation of neurilemoma arising from the distal portion of the medial plantar nerve that was identified and excised is presented with a 19-month follow-up.
Neurilemoma of the tibial nerve causing intermittent claudication.
Tibial Neuroma Presenting as a Baker Cyst

1991 A CASE REPORT* (BY PETER F. DELUCA, M.D.†, AND ARTHUR R. BARTOLOZZI, M.D.†, PHILADELPHIA, PENNSYLVANIA,†, †, †, †)

Investigation performed at

Fig. 3

An intraoperative photograph revealing a 2.5-centimeter-diameter grayish mass in continuity with the tibial nerve (arrow).
Case Report

(An Occult Schwannoma of the Deep Peroneal Nerve Presenting with Neuralgia Mimicking Sciatica)

From the National Neurosurgery Center, Khoula Hospital (Drs. Sharma and Pawar), Muscat, Oman, and Department of Neurosurgery

Accepted for publication 1 November 1999. Received 3 July 1999.
Schwannoma of the Superficial Peroneal Nerve

G. P. Thomas, MBBS, BSc

Senior House Officer
Department of Orthopaedics
St Mary's Hospital
London
The authors report two cases of neurilemoma localised in the popliteal fossa. Both patients experienced non-specific symptoms such as painful numbness and burning dysaesthesia, involving the lower extremity. Tinel's sign was positive over the popliteal fossa. The patients sought medical advice and underwent conservative treatment without any relief, for a long time before the right diagnosis was made. Magnetic resonance imaging revealed in both patients a well-circumscribed mass posterior to the sciatic nerve, occurring in the popliteal fossa. Following surgical excision of the neurilemoma, the patients experienced immediate relief of their chronic symptoms. In similar situations, ultrasound or magnetic resonance imaging of the whole sciatic nerve should be performed if this is indicated by detailed physical examination. Once the diagnosis is made, neurilemomas should be surgically removed, in order to exclude malignancy, prevent neurological deficits.
Compressive lesions of the suprascapular nerve produce weakness and atrophy of the supra- and infraspinatus muscles and a poorly defined aching pain along the posterior aspect of the shoulder joint and the adjacent scapula. Entrapment neuropathy of the suprascapular nerve is fairly common whereas tumorous lesions are rare; among the latter ganglion cysts are frequently seen. An isolated solitary schwannoma of the suprascapular nerve presenting with atypical neuralgic pain is exceptional. The location of a schwannoma under the firm deep cervical fascia in the posterior triangle of the neck is implicated in the genesis of neuralgic pains mimicking the suprascapular entrapment syndrome. One such case is reported with discussion of the relevant clinical features.
OBJECTIVE AND IMPORTANCE: Intraosseous schwannomas (neurilemomas) are rare neoplasms. In one large series, schwannomas accounted for less than 0.2% of all primary bone tumors. These schwannomas most commonly arise in the mandible and have also been observed, very rarely, in the vertebra. We present a patient with an unusual intraosseous schwannoma in the lumbar vertebra. CLINICAL PRESENTATION: A 58-year-old man who was admitted to Cathay General Hospital had complained of numbness and pain in his lower extremities for approximately 1 year. At the time of admission, a neurological examination revealed mild motor weakness and sensory numbness bilaterally in his lower legs. Magnetic resonance images showed a neoplastic lesion occupying the entire L4 body and the superior part of the L5 body, with marked perivertebral protrusion and compression of the thecal sac and bilateral neuroforamina. INTERVENTION: The patient underwent a computed tomography-guided needle biopsy and a two-stage operation. During the initial surgical procedure, the tumor mass was totally removed via an anterolateral approach. Fixation and fusion of the third to fifth lumbar vertebral bodies was accomplished using a full-thickness iliac bone graft with an anterior locking plate and screws. During the second operation, posterior fusion of L2, L3, L4, L5, and S1 with Luque wires and a Hartshill rectangle rod was performed to increase the spinal stability. CONCLUSION: Intraosseous schwannomas are rare neoplasms that are not commonly observed in the lumbar vertebral bodies. Schwannomas in the lumbar spine commonly originate from the nerve passing through the neural foramina. We report a rare case of L4 intraosseous schwannoma, the magnetic resonance imaging, computed tomographic, and histological findings, and the surgical procedures.
A case of solitary schwannoma and one of solitary neurofibroma originating from the spinal accessory nerve in the posterior triangle of the neck are described. Location of such neoplasms in this region is exceptional. The authors emphasize the importance of accurately enucleating the mass; when it is impossible to preserve the continuity of the neural pathway, nerve repair should be considered.
We report a patient with neurinoma of the spinal accessory nerve, who complained of intermittent occipital headaches, nausea, vomiting, blurred vision and unsteady gait. Neurological examination revealed papilledema, bilateral horizontal nystagmus and right cerebellar signs. Computed tomography revealed mild hydrocephalus, a low-attenuated lesion with a faint capsule after enhancement and partial compression of the 4th ventricle in the right posterior fossa. Vertebral angiography revealed no definite tumor vessels or stains. Under the impression of a posterior fossa tumor, a suboccipital craniectomy with a C1 and C2 laminectomy was performed. A 4 x 4 x 2.5 cm³ dumbbell tumor arising from the left spinal accessory nerve at the C2 level was found 4 x 4 x 2.5 cm³. The tumor extended upward through the foramen magnum with upward displacement of both tonsils to the right jugular foramen with slight adhesion to the right IX, X and XI cranial nerves. The left spinal accessory nerve was severed from the pedicle at the C2 level, and the tumor was totally removed. Diagnosis was made during the operation. The pathological examination showed neurinoma with cystic degeneration. During the following year, atrophy of the left sternocleidomastoid and trapezius muscles were noticed. Up to the time of this writing, there had been no clinical recurrence.
Primary tumors of the brachial plexus are unusual. We describe a patient with a large schwannoma of the lower trunk of the brachial plexus that had the radiologic appearance of an apical lung mass. Use of a posterior subscleral approach as well as intraoperative nerve action potential recording permitted resection with spared function.
Giant sacral schwannoma is a very rare tumor (25 cases reported). The authors report 3 cases of giant sacral schwannoma treated by curettage through posterior approach and discuss symptoms and treatment. These tumors were characterized by their minimal symptoms compared to radiographic findings. Magnetic resonance imaging must be performed in order to detect extraosseous tissue component and intradural invasion. A biopsy was performed to confirm the diagnosis before definitive treatment. Wide resection was proposed by many authors because of the high recurrence rate. We believe that a wide resection is too severe as it causes neurologic sacrifices. A curettage through posterior approach preserves nerve function, and if a local recurrence occurs it remains possible to perform a wide resection. When sacroiliac joint instability is detected, a lumboiliac arthrodesis is indicated. Osteosynthesis could be performed with spine device (using pedicular and iliac screws).
Nerve sheath tumors that involve the sacrum are rare. Delayed presentation is common because of their slow-growing Nature.

The authors discuss a case of a sacral nerve sheath tumor. They also propose a classification scheme for these tumors based on their location with respect to the sacrum into three types (Types I-III). Type I tumors are confined to the sacrum; Type II originate within the sacrum but then locally metastasize through the anterior and posterior sacral walls into the presacral and subcutaneous spaces, respectively; and Type III are located primarily in the presacral/retroperitoneal area. The overwhelming majority of sacral nerve sheath tumors are schwannomas. Neurofibromas and malignant nerve sheath tumors are exceedingly rare. Regardless of their histological features, the goal of treatment is complete excision. Adjuvant radiotherapy may be used in patients in whom resection was subtotal. Approaches to the sacrum can generally be classified as anterior or posterior. Type I tumors may be resected via a posterior approach alone, Type III may require an anterior approach, and Type II tumors usually require combined anterior-posterior surgery.
OBJECTIVE: To review the features of spinal schwannoma in a case that mimicked a lumbar disc herniation.

FEATURES: A 37-yr-old woman suffered from a 4-yr history of progressive low back and leg pain, with neurological involvement of several nerve roots. Noteworthy symptomatology included increased pain when lying recumbent and urinary and fecal incontinence. Several health care practitioners diagnosed her with a lumbar disc herniation. Investigations with myelography, computed tomography (CT) with myelographic contrast and magnetic resonance imaging (MRI) revealed the presence of an intradural tumor at the T12-L2 region. INTERVENTION AND OUTCOME: The tumor was surgically resected via laminectomies at T12-L2. The patient experienced a decrease of pain intensity but continued to complain about the low back and posterior thigh pain and has been unable to return to work. She continues to suffer from urinary incontinence, which is controlled by medication. There has been no recurrence of the tumor. CONCLUSION: Spinal pathology such as schwannoma of the cauda equina can mimic common complaints of low back pain seen in clinical practice. Differentiating features of cauda equina tumors from lumbar disc prolapse include: pain on lying recumbent, progressive nature of the pain and neurological deficit, involvement of several nerve root levels and intractability of the condition to conservative therapy. The most appropriate methods for imaging these tumors are CT with myelography or MRI. Patient prognosis is improved with early detection and removal. Chiropractors can play a pivotal role in the care of these patients by being suspicious of patients who do not respond as expected to a course of manipulative therapy, by investigating or referring appropriately and by aiding in active rehabilitation postoperatively.
A case of cauda equina neurinoma associated with intracranial hypertension is reported. A 59-year-old female with a history of disturbed orientation was admitted. A neurological examination upon admission revealed the disorientation and gait disturbance. Superficial sensation under L3 was impaired. A computed tomographic (CT) scan presented the enlargement of ventricles and the slightly poor description of cerebral sulci. Since the patient had a high fever up to 40.1°C, meningitis was suspected. Cerebrospinal fluid revealed that cell count was only 2/3, while the protein concentration was markedly elevated (389mg/dl). Froin reaction was extremely positive and fibrin was observed. Based upon these findings, the spinal tumor was considered. Plain lumber film showed the posterior scalloping of the L5 and S1 vertebral bodies. Gd-DTPA enhanced MRI showed a high signal intensity area at the cauda equina. Diagnosed as a cauda equina tumor, the total resection of the tumor was performed via laminectomy of L3-S1. The tumor was involved with nerve filaments at the cauda equina. The pathological diagnosis was neurinoma. After the operation, her symptoms improved and a CT scan revealed the reduction of the ventricular size. However, the protein concentration of cerebrospinal fluid did not normalized.
Department of Orthopaedic Surgery, Fukui Medical School, Japan. (1996)

The case history of a patient with a dumb-bell neurilemmoma arising from the L5 nerve root is described. The tumour extended into the outlet of the neural foramen at L5-S1 on the left and was also compressing the dural sac and the S1 nerve root. A limited laminotomy at L4-5 and L5-S1, with preservation of the neural arch, was followed by microsurgical medial foraminotomy at L5-S1 which allowed visualisation of the nerve root and enucleation of the tumour. The technique maintains stability of the posterior elements and spinal movement, and is recommended for the removal of this type of tumour.
A group of 42 patients with C-1 and C-2 neurinomas treated during the 10-year period 1982 to 1992 has been collected, including 25 cases from 20 French neurosurgical departments and 17 personal cases from the Neurosurgical Department of the Lariboisiere University Hospital, Paris. Analysis of this series reveals some interesting findings relating to multiplicity of tumors, extradural extension, and neurofibromatosis. There were seven patients with multiple lesions (bilateral C-2 neurinomas in six cases and two neurinomas at C-2 and one at C-1 in one case). In the 35 other cases, 16 lesions were entirely extradural and 19 had an hourglass configuration. Thirteen patients presented signs of neurofibromatosis. One lesion had a melanotic form and another was a radiation-induced schwannoma. Surgical results were excellent in most cases with no immediate postoperative death. Best results in terms of complete removal and neurological condition were achieved with posterolateral or anterolateral surgical approaches (17 cases) as compared with the standard midline posterior route (25 cases).
We report on a case harboring a **cervical dumbbell type neurinoma**. The tumor was completely removed by a modified posterior approach, consisting of partial hemilaminectomies of C2 and C3 with preservation of the facet joint. The operative field under microscope was limited by the preserved facet joint of C2/3. However, sufficient bulk reduction of the epidural and paravertebral mass enabled us to obtain a good operative field. The paravertebral mass, which extended anteriorly to just beside the posterior aspect of the carotid sheath, was removed through the lateral space. The operative field was easily widened beside the right facet joint of C2/3 with partial removal of the posterior part of the transverse process of C2 and C3.

The transit portion of the tumor to the normal nerve fiber was also identified through this space. The intracanalicular mass was removed by the partial hemilaminectomies of C2 and C3 without compressing the dural sac. Following sufficient reduction of the bulk, the right vertebral artery was identified at the anteromedial margin of the enlarged intervertebral foramen. Finally the intradural part of the tumor was removed through this space. Our modified posterior approach is a less invasive method to the bony elements of the cervical vertebrae and may minimize the incidence of postoperative instability and angular deformity. This approach also eliminates the necessity of long postoperative immobilization using a rigid cervicothoracic brace.
The case of a 52-year old patient is described who was operated on for pronator-teres syndrome. Eight months postoperatively reoperation was necessary because of recurrent pain. The median nerve showed haemorrhagic necrosis in the region of the pronator teres muscle with an extensive intrafascicular swelling of the adjacent proximal and distal nerve segments. After resection of the median nerve, the histological examination revealed a benign schwannoma. Reconstruction of the median nerve was performed by interfascicular nerve grafting. Three months later severe pains recurred in the forearm which had to be opened in an emergency operation. Extensive haemorrhagic necrosis was evident. The histological diagnosis was now malignant schwannoma. Upper arm amputation was performed and later a functional prosthesis was fitted. Three years after the amputation multiple metastases occurred; the patient died within three months.
Endobronchial metastasis from malignant Schwannoma.
Malignant schwannoma of the medial plantar branch of the posterior tibial nerve
Malignant schwannoma in the foot is extremely rare. The authors present a case involving the posterior tibial nerve in a patient with neurofibromatosis, without previous history of malignancy.

Following full excisional biopsy, the diagnosis was made using immunohistochemical studies and electron microscopy. A review of the literature also is presented.
Intraosseous Epithelioid Malignant Schwannoma

By Branko Sestan PhD FRCS; Damir Miletic PhD; Nives Jonjic, PhD; Gordan Gulan, PhD, FRCS

Figure 4: Epithelioid malignant peripheral sheath tumor showing a vague nodular pattern (A, ×100) of a large epithelioid cells with polymorphic nuclei (B, ×200).

Note irregular cortical disruption and anterior extension of the lesion into the subcutaneous fat tissue.
Conclusion

- Consider schwannomas as a cause of persistent chronic heel pain despite the rarity of the disease.
- Deep-seated schwannomas in the foot can easily be overlooked and misdiagnosed as tarsal tunnel syndrome or plantar fasciitis.
- Chronic unexplained foot and calf pain and a positive Tinel's sign should raise suspicion of posterior tibial nerve neurilemoma.
- Magnetic resonance imaging provides the best modality for differential diagnosis.
- Surgical excision is the golden standard modality for Rx.