

Case Report

Surgical intervention for peripheral plexiform schwannoma of the sciatic nerve: a pediatric case study

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ABSTRACT

Plexiform Schwannoma, or neurilemmomas, is a rare and benign manifestation of peripheral nerve sheath tumors. The tumor mostly arises from the gradual growth of Schwann cells. These nerve tumors occur in several sites, with the most prevalent being the head, or neck, or even upper extremities. This tumor predominantly manifests in individuals aged 30 to 40 years and is seen equally in both sexes, with these tumors generally measuring less than 2 cm in size. A 14-year-old boy was referred to Oriana Specialty Hospital, Sharjah, UAE, with a one-year history of vague pain in the middle third posterior aspect of the right thigh accompanied by mild tenderness but no peripheral neurological deficit. Magnetic resonance imaging (MRI) of the right thigh revealed a lesion, and a true-cut biopsy confirmed the diagnosis of a peripheral plexiform Schwannoma arising from the sciatic nerve. On 29 December 2024, a surgical excision of the right thigh sciatic nerve Schwannoma was carried out. A posterior midline thigh incision was made, and the procedure was done with advanced techniques, including the use of a microscope and intraoperative nerve stimulator, to ensure precise tumor dissection from the sciatic nerve. The entire Schwannoma, measuring 24×5.5 cm, was successfully excised. There were excellent recoveries post operatively with no neurological deficits or complications of surgery. He was mobilized with full weight-bearing three hours after the surgery and discharged the next day on oral medications. This particular case testifies to the complete excision of a tumor through proper surgery techniques, preserving the neurological function, with promising outcomes for similar cases.

Keywords: Schwannomas, Lumbar disease, Head, Plexiform, Sciatic nerve, Surgical techniques

INTRODUCTION

Schwannomas are benign nerve sheath tumors, that originate from Schwann cells, accounting for about 5% of all soft tissue tumors.¹ Although more than half of these tumors are found in the head and spine, their appearance in the extremities, particularly in the thighs, is rarely seen.² Schwannomas are known to grow slowly and often lead to a lack of symptoms at early stages. That especially becomes the case when they find themselves in locations highly muscular which renders their visualization so difficult due to lack of identifiable symptoms.³

As their sizes increase these masses may eventually compress the lying nearby nerves producing some symptoms; either localized aching, paresthesia, or even frank paralysis.⁴ However, Schwannomas in diagnosis becomes increasingly tricky when concomitant, especially with pathology such as the lumbar intervertebral discs' hernial rupture. In such cases, thigh pain—a potential symptom of a sciatic Schwannoma—is often misattributed to lumbar disc herniation. Pain management strategies aimed at treating lumbar disc herniation can further obscure the clinical picture, masking the underlying Schwannoma and delaying its diagnosis.

Sciatic nerve Schwannoma is a rare but significant cause of sciatica, which usually presents with pain that radially courses along the path of the sciatic nerve, usually resulting from a herniation in the lumbar spine.^{5,6} But in a case of sciatica where there is no evidence of a prolapsed disc in the lumbar spine and as confirmed by imaging, crossed straight leg raise test negative, suggests that the cause of pain is not a herniated disc but rather another pathology such as a Schwannoma of the sciatic nerve. The crossed straight leg raise test is commonly utilized to evaluate nerve root irritation secondary to disc herniation. A negative test in this context may suggest that the problem resides outside the lumbar spine and may be located in the sciatic nerve.

Sciatic nerve Schwannomas are responsible for the formation of myelin sheaths around peripheral nerves. These tumors are rare and tend to present with compression of the sciatic nerve, producing pain, tingling, and numbness in the distribution of the nerve. It can, therefore, simulate symptoms of sciatica due to lumbar disc herniation. The diagnostic work-up, however, should focus on determining the specific nature of the lesion, especially if the symptoms of sciatica persist without evidence of lumbar disc pathology.

Magnetic resonance imaging is the best diagnostic tool for identifying Schwannoma of the sciatic nerve. Magnetic resonance imaging (MRI) is highly sensitive for visualizing soft tissue tumors and can obtain detailed images of the sciatic nerve along its entire course. In suspected Schwannoma, MRI helps to detect the presence of a mass along the nerve, assesses its size, location, and relationship to surrounding structures, and confirms the diagnosis. The imaging characteristics of a Schwannoma usually consist of an encapsulated well-defined mass which often shows features like homogeneous enhancement following the injection of contrast agent and a "target" or "onion skin" appearance secondary to alternating regions of hyperintensity and hyperintensity on T2 weighted images.^{7,8}

This report documents a rather unusual and puzzling case where there was occurrence of sciatic Schwannoma concurrently with lumbar disc herniation. In this case, the diagnosis was significantly delayed by a decade since the onset of symptoms to final identification of the Schwannoma. This case emphasizes that even in cases of common diseases such as lumbar disc herniation, adequate diagnostic work-up should be conducted in patients who have persistent or atypical symptoms to avoid missing rarer but clinically relevant pathologies like Schwannomas.

CASE REPORT

A 14-year-old male patient presented with a newly diagnosed Schwannoma located in the right thigh—a notably uncommon occurrence for this age group and anatomical location. Schwannomas are typically benign

nerve sheath tumors, and their presence in the extremities, especially in pediatric cases, is rare. To determine the metabolic activity of the lesion and exclude the possibility of hypermetabolic foci or distant metastases, a comprehensive PET/CT scan was undertaken as part of the diagnostic and staging process.

The procedure began with a pre-imaging assessment of the patient's blood glucose level, which was recorded at 91 mg/dl, confirming the suitability for 18-fluoro-2-deoxyglucose (FDG) administration. A radiotracer dose of 1 ml FDG, with an activity of 133.94 MBq, was administered intravenously into the patient's right arm. After a standardized uptake period of 54 minutes, the patient was asked to void in a "hot toilet" to reduce urinary FDG concentration, thus minimizing interference during imaging.

The imaging process involved acquiring a non-contrast CT scan followed by a PET scan, covering the entire body from the head to the feet. The non-contrast CT images were utilized for anatomical localization of the lesion and to provide photon attenuation correction for the PET images. The entire procedure was completed without the need for ancillary medications.

The PET/CT scan provided crucial insights into the metabolic behavior of the thigh Schwannoma, allowing for an accurate evaluation of the lesion and aiding in disease staging. However, the absence of previous PET/CT imaging limited the ability to compare findings over time, which could have been beneficial for assessing disease progression. Despite this, the detailed imaging findings played a vital role in guiding the clinical team's approach to management and treatment planning for this rare presentation of a pediatric thigh Schwannoma.

The findings from the PET/CT scan confirm a hypermetabolic intermuscular soft tissue mass lesion in the posterior compartment of the right thigh, measuring 5.6×5.7×22.6 cm. This lesion has a maximum standardized uptake value (SUV max) of 4.78, which is consistent with a neoplastic pathology, likely Schwannoma. There is no evidence of distant metastases or hypermetabolic foci in the axial or proximal appendicular skeleton, nor any FDG-avid lesions in the skin, subcutaneous tissues, or other organs.

In the head and neck region, no abnormal FDG uptake was seen in the brain or neck structures, and no FDG-avid lymphadenopathy was detected in the cervical or supraclavicular lymph nodes. Physiologic FDG uptake was noted in brown fat across various regions, including the cervical, occipital, and axillary areas. Additionally, mild to moderate frontal sinusitis was observed incidentally.

The lesion is well-circumscribed and lobulated, occupying the posterior compartment of the thigh. It appears hyperintense on T2-weighted imaging, which is consistent

with Schwannoma characteristics. The lesion shows an elongated, fusiform morphology, a hallmark of peripheral nerve sheath tumors like Schwannomas.



Figure 1: MRI scan of a soft tissue mass in the thigh, specifically a sagittal view.

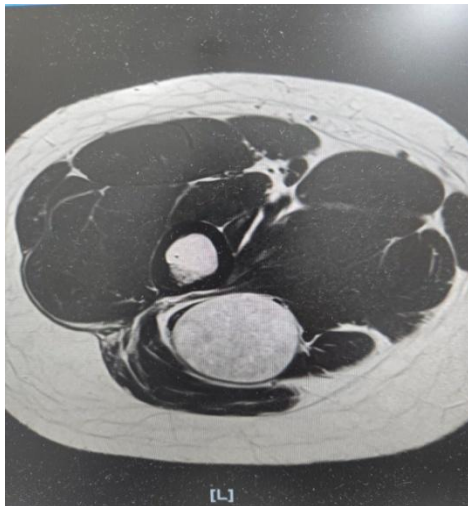


Figure 2: MRI scan appears to be an axial section of the thigh, showcasing a soft tissue lesion in the posterior compartment.



Figure 3 (a and b): Surgical treatment and complications of deep-seated nodular plexiform neurofibromas.

The chest imaging showed no significant findings, with unremarkable lung parenchyma and no signs of pleural effusion or pneumothorax. Similarly, there were no FDG-avid masses or lymphadenopathy in the chest. Hence The PET/CT and MRI results revealed the existence of a well-defined, lobulated soft tissue mass in the posterior compartment of the right thigh, which is consistent with a Schwannoma. The brain, neck, or chest showed no aberrant FDG absorption; likewise, no FDG-avid lymphadenopathy was seen, thereby excluding metastatic involvement. Brown fat showed evidence of physiologic FDG absorption; incidental mild to moderate frontal sinusitis was found. MRI images further defined the lesion as hyperintense on T2-weighted imaging with an elongated, fusiform appearance, therefore supporting its diagnosis as a peripheral nerve sheath tumor.

DISCUSSION

The discussion of this study focuses on the diagnostic, imaging, and surgical challenges associated with peripheral plexiform Schwannomas of the sciatic nerve, particularly in pediatric patients. Given their rarity and slow-growing nature, these tumors often present with delayed symptoms, leading to late-stage diagnoses. Advanced imaging modalities such as MRI and PET/CT play a crucial role in identifying these lesions and distinguishing them from malignant counterparts. This study highlights the clinical presentation, histopathological findings, and surgical approaches for managing sciatic Schwannomas, emphasizing the importance of early detection and precise intervention to optimize patient outcomes.

In the abdomen and pelvis, the organs were normal, with no abnormal FDG uptake in the liver, spleen, kidneys, or other abdominal structures. No FDG-avid lymphadenopathy or abnormal fluid collections were seen.

Studies suggest that sciatic nerve Schwannomas exhibit a gender and age predilection, with middle-aged women being more frequently affected. While hormonal factors may contribute to this trend, further research is necessary to establish a definitive pathophysiological basis. These tumors are relatively rare, and their diagnosis is often delayed due to their slow-growing nature. Symptoms, such as pain or neurological deficits, tend to be subtle or intermittent in the early stages, leading to misdiagnosis or delayed referrals for advanced imaging. The average time from symptom onset to diagnosis is approximately 33 months, but this delay has been decreasing in recent years. The increased availability and utilization of MRI have played a critical role in earlier detection. MRI not only helps identify Schwannomas at an earlier stage but also provides detailed imaging features that assist in differentiating them from other nerve-related tumors. Prompt and precise diagnosis is crucial for timely intervention along with improved patient outcomes.

Pain is the most frequent symptom prompting patients with sciatic Schwannomas to seek medical attention. Among 33 patients reviewed, 31 reported pain, underscoring sciatica as the predominant clinical presentation. However, variations in symptomatology exist, as two patients did not report any pain. The pain is typically associated with compression of the sciatic nerve, leading to nerve irritation and symptoms such as shooting, burning, or sharp aching pain radiating along the nerve's distribution. Clinical assessment plays a vital role in the diagnostic process. In more than half of the cases, a thorough physical examination can identify signs like a palpable mass or neurological symptoms indicative of nerve compression. For tumors that are not palpable, a positive Tinel's sign can serve as a valuable diagnostic clue. This sign, characterized by a tingling sensation elicited by gentle percussion over the affected nerve, suggests irritation or compression. These clinical findings emphasize the importance of detailed examination and targeted diagnostic approaches in the timely identification and management of sciatic schwannomas.^{9,10}

From a morphological point of view, most sciatic Schwannomas grow in isolation, presenting themselves as solitary lesions. The presence of multiple Schwannomas along the sciatic nerve is very rare, and only a few cases have been reported in which more than three Schwannomas were present in the same patient. These tumors are usually small, with an average diameter of 5.29 cm, and generally remain benign. The possibility of Schwannomas deteriorating into malignant tumors is extremely low, with only one reported case of a sciatic Schwannoma transforming into a malignant tumor.⁹

Schwannomas, benign nerve sheath tumors, usually show characteristic MRI features that help in the diagnosis. On T1-weighted images, Schwannomas are isointense to hypointense because of their high content of dense fibrous tissue. They are hyperintense on T2-weighted images due to myxoid or cystic components. Enhancement post-contrast is usual, because of the vascularity and permeability of the tumor's capsule. Heterogeneity of signal may be seen in the presence of internal hemorrhage or cystic change, with enhancement post-contrast in these regions.¹¹

While such MRI characteristics may be common for Schwannomas, they are not entirely specific, because neurofibromas and malignant peripheral nerve sheath tumors (MPNSTs) may sometimes present similarly. Other imaging signs, such as the entering and exiting nerve signs the peripheral nerve passes through the mass-and the target sign, in which Schwannomas feature a central low-to intermediate-signal surrounded by a higher T2 signal, can be useful in further narrowing the diagnosis. Another hallmark of Schwannomas is the fascicular sign, which represents multiple ring-like structures that correspond to fascicular bundles in normal nerves. Furthermore, muscle atrophy within the distribution of the nerve might suggest the presence of a neurogenic neoplasm, although again this

is more nonspecific and can be seen in other nerve-associated tumors.¹¹ Overlapping features may make it challenging to distinguish between benign Schwannomas and malignant soft-tissue tumors. Characteristics such as ill-defined margins and larger tumor size (>5 cm) may suggest malignancy.^{13,14} On the other hand, the split fat sign and the bright rim sign, which show a high T2 signal around the periphery of the mass, are suggestive of Schwannomas. Benign Schwannomas are usually not lobulated, have minimal peritumoral edema, and have a well-defined capsule, which are features that reliably identify Schwannomas when two or more are present simultaneously.

Intra-capsular excision preserves the tumor capsule and is used when the tumor is closely associated with the nerve, helping maintain nerve integrity and minimizing postoperative neurological deficits. Extracapsular excision involves the removal of the tumor along with its capsule, which is necessary when the tumor is deeply embedded in the nerve and surrounding tissues. The preservation of the capsule during resection has been shown to maximize nerve function preservation, reducing the risk of nerve damage and improving postoperative outcomes. While some patients may experience temporary neurological symptoms such as weakness, numbness, or tingling, these usually improve within a year as the nerve regenerates.¹⁵

According to Merritt et al, the histopathological examination in the case confirmed the lesion as a Schwannoma, although it was initially misdiagnosed as Morton's neuroma based on clinical and ultrasonographic findings.¹⁶ Schwannomas are histologically characterized by distinct regions known as Antoni A and Antoni B areas. Antoni A areas are hypercellular, exhibiting nuclear palisading and the formation of Verocay bodies, while Antoni B areas are hypocellular and composed of loosely arranged spindle cells in a myxoid stroma. Plexiform Schwannomas, a rare subtype of Schwannoma, differ in their morphological configuration. These tumors exhibit a plexiform architecture with multiple interconnected fascicles and nodules primarily composed of Antoni A regions. Additionally, plexiform Schwannomas demonstrate immunoreactivity for the S100 protein, a specific marker found in Schwann cells, which aids in distinguishing these tumors from other nerve sheath tumors.

This study highlighted the importance of histopathological evaluation in accurately diagnosing Schwannomas, particularly in distinguishing them from other similar-appearing lesions, such as Morton's neuroma, which can have overlapping clinical and imaging features.¹⁷

The discussion summary the significance of PET/CT imaging in evaluating a rare pediatric case of sciatic Schwannoma, emphasizing its role in determining metabolic activity and staging. The use of FDG-PET/CT enabled precise localization of the lesion and

differentiation from malignant processes, aiding clinical decision-making.

Despite the absence of prior imaging for comparison, the study underscores the diagnostic value of PET/CT in assessing peripheral nerve sheath tumors. The findings contribute to the understanding of pediatric Schwannomas and support the integration of advanced imaging in their evaluation and management.

CONCLUSION

Sciatic nerve Schwannomas, though rare, can now be diagnosed more promptly due to advancements in MRI technology. Pain is the predominant symptom, and a thorough physical examination, along with tests like Tinel's sign, plays a crucial role in early detection. Most sciatic Schwannomas are solitary and small, with a benign prognosis and an extremely low risk of malignant transformation. Increased awareness and improved imaging techniques have led to earlier diagnoses, ultimately resulting in better treatment outcomes for patients. This case highlights the positive outcome following complete excision of the tumor with proper surgical techniques, preserving neurological function and ensuring rapid recovery. The patient experienced no postoperative complications or neurological deficits and was able to resume full weight-bearing activities shortly after surgery, demonstrating the efficacy of timely diagnosis and intervention for this condition.

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