Case Report

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Neglected Giant Schwannoma of the Sciatic Nerve. Case Report and Literature Review

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Abstract

Background Data: Schwannomas originating from the sciatic nerve are extremely rare and usually present as a pathological mass on palpable examination or pain located in the thigh. Motor and sensory deficits are observed more often with big size lesions.
Purpose: To describe a rare case of giant schwannoma of the sciatic nerve with 2 years delay in management.
Study Design: Case report and review of the literature.
Patients and Methods: A 19-years- old female patient was referred to Suez Canal University Hospital after 6 years of pain and 2 years of improper management. She presented with increasing swelling in the posterior aspect of her right thigh associated with paresthesia and weakness. After neurological examination an MRI of the right thigh was done and revealed huge mass attached to the right sciatic nerve.
Results: Surgical excision was undertaken, carefully dissecting the lesion from the sciatic nerve. Histopathological examination revealed a sciatic nerve schwannoma. The patient had marked postoperative recovery with marked pain reduction and improved neurological deficits.
Conclusion: Schwannomas of the sciatic nerve are eccentrically located on the nerve. they should be systematically suspected if thigh mass or persistent sciatica is reported. Surgical excision has good prognosis. (2018ESJ157)
Keywords: Schwannoma; sciatic nerve; sciatica

Introduction

Schwannomas originating from the sciatic nerve are extremely rare and usually present as a pathological mass in palpable examination or pain located in the thigh. Painful palpable mas on examination is the most common clinical symptom of sciatic nerve Schwannoma.[3,4,6] Motor and sensory deficits are observed more often when the size of a tumor is more than 40 mm. Schwannomas most commonly are observed in adults between 20 and 50 years old.[11,13]

We report a very rare case of giant schwannoma of the sciatic nerve in a 19-year-old female who presented with increasing swelling and discomfort in the posterior aspect of her right thigh. The surgical excision was delayed for 2 years due to lack of proper diagnosis. We demonstrate that even with such large tumors, surgical excision could be successfully carried out to resolve

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all symptoms while causing no permanent nerve damage.

Case Report:

Clinical Presentation:

A 19-year-old female patient had a 6-year history of increasingly severe right thigh pain. She had initially reported intermittent aching pain in the right posterior thigh after waking. After a 4-year period, the pain radiated from the right buttock down the posterior aspect of the right leg to the ankle. A mass appeared in the posterior upper right thigh and it gradual increased in size till became the size of an orange. Medical consultation was sought and a tru cut biopsy was taken. Biopsy result was sarcoma and chemotherapy was started for 6 months. Pain quietened for 2 months and then it recurred again. It was more severe than before and the mass increased in size. Another biopsy was taken and also the result was sarcoma. Radiotherapy was started and followed by another cycle of chemotherapy. Pain continued to increase in severity. The pain become agonizing and severe and lower limb weakness started. Muscle of the right lower limb started to decrease in size and limping gait was evident. She was referred to Suez Canal University Hospital in December 2017.

Clinical Examination:

On physical examination, a well - formed mass was palpated in the back of her upper right thigh. The mass was mobile, firm, tender to palpate and it also elicited a shooting pain down the leg on examination. The size of the mass was about 12 cm in length and 6 cm in width. There was progressive wasting of the right gluteal and calf muscles. The patient walked with a complete right foot drop, avoiding all contact of the right foot with the ground. Neurological examination of the right lower limb revealed marked muscle wasting of the glutei, left tibialis anterior and gastrocnemius muscles, decreased muscle tone, and severe weakness of ankle and toe dorsiflexion and plantar flexion (both were grade 1). The knee jerk was brisk but the ankle jerk was reduced. The right plantar response was not assessed because of hyperesthesia, but the left plantar response was flexor. There was altered soft touch, pin prick, and temperature sensation, with allodynia and hyperesthesia over the right L5 and S1 dermatomes.

Neuroimaging:

MRI of the lumbar spine was done to exclude any associated spinal pathology and was normal. MRI of the right thigh was done and demonstrated a well-defined well-encapsulated mass 12 cms×6 cms. It had heterogeneous high-signal intensity. There was a surgical plane accurately demonstrated the continuity between the mass and the sciatic nerve. (Figure 1)

Operative Treatment:

The patient was operated in the prone position. (Figure 2) The patient underwent a linear midline skin incision on the posterior surface of the thigh and pass laterally around gluteus maximus muscle. The schwannoma was seen in relation to the sciatic nerve. The tumor had originated from the main sciatic nerve trunk. (Figure 3-A) We made a proper exposure of structures adjacent to and both proximal and distal to the lesion. The tumor had displaced, thinned out, and “blanketed” the fascicles so that they encircle the lesion. (Figure 3-B, C) Under microscopic magnification, we made a longitudinal incision between the fascicles that are spanned or blanketed around the tumor. The tumor was enucleated from its capsule without any damage to the sciatic nerve branches (Figure 3-D). Complete excision of the tumor was performed. Macroscopically, the tumor was characterized by an encapsulated nodule 12 cm in length with a firm greyish cut surface. (Figure 3-E) Repair of epineurium of tibial and peroneal nerves after total excision of the mass was done. (Figure 3-F)

Postoperative Course:

Postoperatively, the patient demonstrated marked improvement of her agonizing pain. No added motor deficit was evident. Immediately post operatively she got some improvement of her foot weakness that might be from pain limitation. Physiotherapy was started. The patient returned to her follow up visit at 1 month completely asymptomatic (grade 5 in all muscle groups). The histopathological report confirmed the diagnosis of a sciatic nerve schwannoma, owing to the presence of Antoni A and B areas and Verocay bodies.

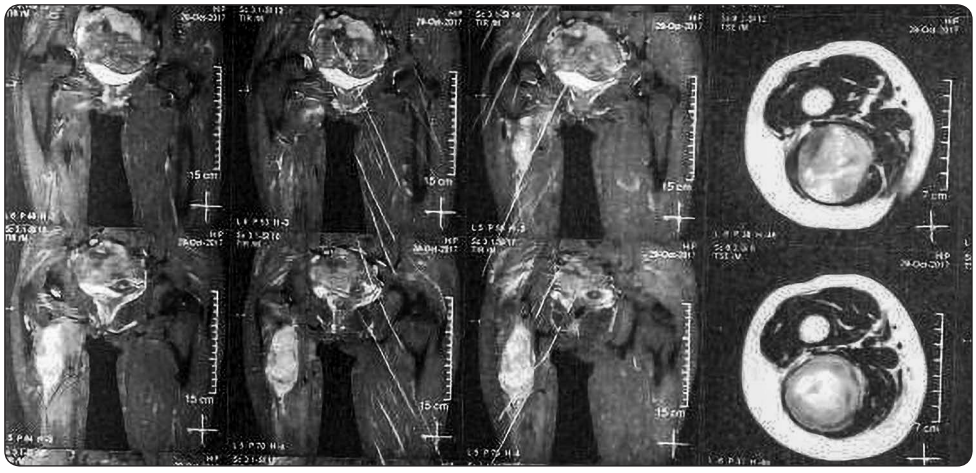

Figure 1. Magnetic Resonance Imaging (MRI) image of the posterior aspect of the right thigh: well-defined, homogeneous fusiform mass.

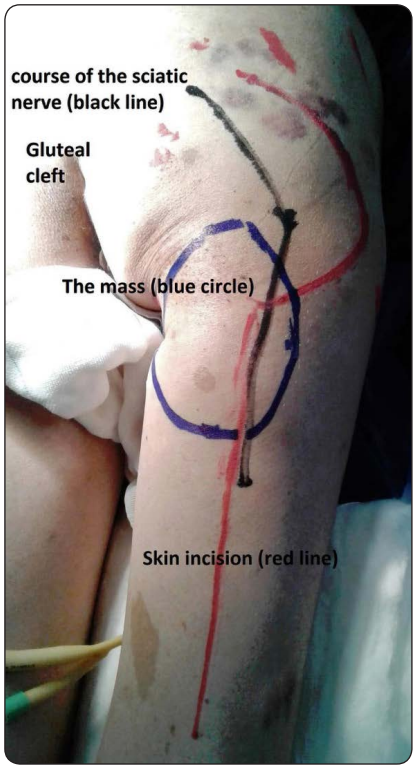

Figure 2. Preoperative view showing surgical planning. The mass (blue circle), anatomical course of the sciatic nerve (black line) and surgical incision (red line).

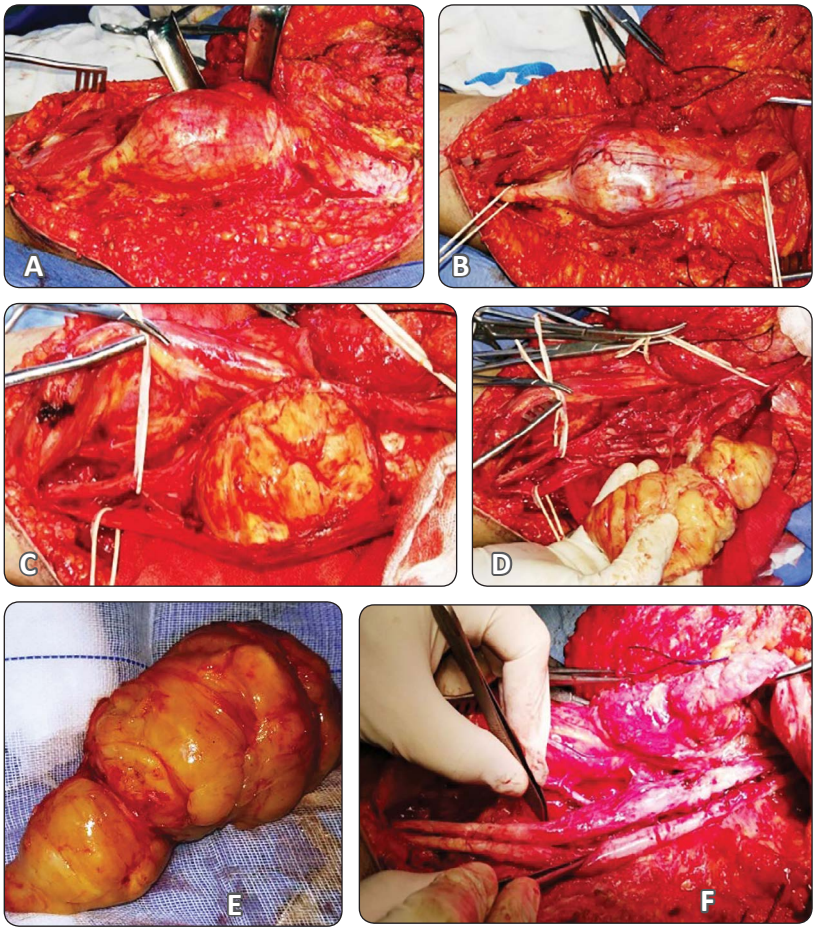

Figure 3. (A) Intraoperative view showing initial exposure of the mass and the sciatic nerve. (B) Further dissection with isolation of both ends of the sciatic nerve. (C) After dissection of the perineurium the mass appeared bisecting the two sciatic nerve branches. (D) The mass attached to its origin nerve fascicle. (E) The mass after total separation. (F) Repair of perineurium of tibial and peroneal nerves after total excision of the mass.

Discussion

The sciatic nerve is the largest nerve in the human body. Schwannomas, also called neurinomas or neurilemmomas, are tumors arising from the Schwann cells of the neural sheath. Peripheral nerve sheath tumors are rare conditions. Frequent locations for schwannomas are the head, neck and main nerve trunk.[1,7] Schwannomas in the sciatic nerve are rare and usually present as a mass or pain in the thigh. Sciatic schwannoma frequency is less than 1%.[3,4,6] Schwannomas occur in any age group; there is no sex predilection. The posterior tibial nerve at the tarsal sinus is the most frequently involved nerve of the lower limb. The most common clinical presentation of sciatic nerve schwannoma consists of a painful palpable mass.[9,11,13] Schwannomas most commonly occur in adults between 20 and 50 years of age. Schwannomas are usually homogeneous on both T1- and T2-weighted images but a neurofibroma is usually heterogeneous. Sciatic schwannoma has a good prognosis and a low incidence of recurrence and malignant transformation. The risk of malignant transformation is approximately 18% in neurofibromatosis type 1 and 5% in schwannomas. Patients with von Recklinghausen disease carry a worse prognosis.[2,10,12,16]

Surgical excision is the treatment of choice. Schwannomas are theoretically removable since they repulse fascicular groups without penetrating them thus allowing their enucleation while preserving nerve continuity,[8,11,14,15] as reported in our patient. Microsurgical excision should be performed using electrical stimulation to facilitate detection of motor fascicles. The sciatic nerve fascicles might sometimes be incorporated peripherally on the tumor capsule thus requiring to be sacrificed.[8]

Our patient had delay in treatment and wrong exposure to chemotherapy and radiotherapy due to improper sampling of her mass. Biopsy is often necessary to diagnose a mass that is indeterminate based on history, physical, laboratory, and imaging studies alone. The goal of biopsy is to obtain diagnostic tissue while minimizing morbidity, limiting potential tumor spread, and avoiding interference with future treatments. Techniques that have evolved to accomplish these goals include open surgical biopsy, core biopsy, and fine-needle aspiration (FNA). Open surgical biopsy was 100% accurate on all accounts. With regard to determining malignancy, fine-needle aspiration and core biopsy had 79.17% and 79.2% sensitivity, 72.7% and 81.8% specificity, 67.9% and 76% positive predictive value, 82.8% and 84.4% negative predictive value, and an overall accuracy of 75.4% and 80.7%, respectively.[5] In regard to determining exact diagnosis, fine-needle aspiration had 33.3% accuracy and core biopsy had 45.6% accuracy. With regard to eventual treatment, fine-needle aspiration was 38.6% accurate and core biopsy was 49.1% accurate. In soft tissue mass diagnosis, core biopsy is more accurate than fine-needle aspiration on all accounts, and open biopsy is more accurate than both in determining malignancy, establishing the exact diagnosis, and the guiding appropriate treatment.[5]

Conclusion

Schwannomas of the sciatic nerve are eccentrically located on the nerve. Schwannoma of the sciatic nerve should be systematically suspected if persistent sciatica or thigh mass is reported in young adults. Surgical excision has good prognosis.

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The authors report no conflict of interest

الملخص العربي

ورم شوان كبير مهمل متصل بعصب النسا, دراسة حالة

البيانات الخلفية: نستعرض فى هذا البحث حاله نادره الحدوث لحدوث ورم شوان متصل بعصب النسا و الذى يظهر فى صوره كتله بالفخد و يمكن ان يسبب ضعف حركى او اعراض حسيه اذا زاد حجم الورم عن 4 سم

الغرض: توضيح الخصائص الاكلينيكيه و النتائج الجراحيه لحاله نادره الحدوث لسيده تبلغ من العمر تسع عشر عاما تعانى من كتله متصله بعصب النسا و قد تاخر العلاج لمده سنتين لوجود خطاء فى تشخيص الحاله

تصميم الدراسة: دراسه لحالات اكلينيكيه لحاله نادره و مراجعه للدراسات العلميه

المرضي و الطرق: تم اجراء الجراحه فى فى ديسمبر 2017 فى قسم جراحه المخ و الاعصاب فى مستشفى جامعه قناه السويس بالاسماعيليه . و ذلك بعد الفحص الاكلينيكى العصبى و عمل اشعه رنين مغناطيسى على الفخد الايمن.

النتائج: تم استئصال الورم المتصل بعصب النسا و اظهر الفحص النسيجى ورم حميد شوانى و تحسنت حاله المريضه بدرجه كبيره جدا بزوال الالم و الاعراض العصبيه

الاستنتاج: الورم الشوانى المتصل بعصب النسا هى حالات نادره و تحتاج لتوقع عند وجود الم و كتله بمسار عصب النسا و يمكن علاجها بنجاح.