SCIATIC NERVE SCHWANNOMA – CASE REPORT

PROF M. GORGAN MD PHD, A.M. SANDU, MD, N. BUCUR MD PHD, A. NEACSU MD, V. PRUNA MD, A. VOINA MD, A. GIOVANI MD, D. MARTIN MD, A. DEDIU MD

Emergency Clinical Hospital "Bagdasar-Arseni", Fourth Department of Neurosurgery, Bucharest

Background. Schwannomas are common, benign tumors of the sheath of peripheral nerves. Sciatic schwannomas are rare. PURPOSE: We report a case of a large sciatic schwannoma, emphasizing the role of nerve preservation. Material & methods. A 54 years old female was admitted in our clinic for persistent right sciatalgia, and right L5 and S1 motor loss, symptoms lasting for almost 2 years. Patient underwent surgery in a different department, but the operation was stopped, because of excessive bleeding. Results Neurological examination revealed right sciatalgia, right L5 and S1 motor loss and diminished ankle jerk reflex, but failed to reveal relevant symptoms for disc herniation clinical diagnostic. Thigh CT-scan and MRI showed a large (70/80/75 mm) encapsulated, contrast-enhancing tumor, located in the middle third of the posterior thigh, in intimated relation with the sciatic nerve. Associated chronic myeloproliferative syndrome required oncological treatment and surgery postponement. We performed a subtotal resection of a large, solid tumor, infiltrating the sciatic nerve on 4 cm length, with anatomic integrity of sciatic preservation. Histopathological examination found cellular schwannoma. Outcome was favorable, and on follow-up examination the patients presents no additional deficits and thigh CT-scan showed no evidence of tumor regrowing. Discussions & Conclusions. Diagnostic is challenging, in many symptoms are attributed to degenerative spinal pathology. Any persistent sciatlagia, with irrelevant lumbar MRI must raise the suspicion of a peripheral sheath tumor, and must be further investigated. Surgery is very demanding and associated co-morbidities must be treated prior surgery. Preserving the integrity of the sciatic nerve, assure favorable outcome, with no additional neurological deficits.

Keywords: sciatic schwannoma, peripheral nerve surgery

INTRODUCTION

Peripheral nerve tumors are benign or malignant, and neural or non-neural in origin.[1] Most common benign neural peripheral nerve sheath tumors are schwannoma (neurilemoma) and neurofibroma.[2-4]

Schwannomas are common, slow growing benign tumors of the sheath of peripheral nerves, arising from the Schwann cells. Involvement of the sciatic nerve is rare.[2]

Diagnosis can be delayed for a long period of time, usually symptoms being attributed to lumbosacral degenerative pathology.[3;5;6]

CASE REPORT

A 54 years old female, was admitted into the 4th Department of Neurosurgery, Emergency Clinical

Hospital "Bagdasar-Arseni", for persistent right sciatalgia, and right L5 and S1 motor loss. Symptoms last for almost 2 years. During this period of time the patient sought medical advice several times, but symptoms were mistakenly attributed to lumbosacral discal radiculopathy. Finally, the patient was diagnosed with right sciatic nerve tumor, and underwent surgery twice in a different department, 2 and 3 months before admission in our clinic. Both times the operation was stopped, because of the excessive bleeding.

On admission, clinical examination revealed an over-height patient, having gross thighs. Palpatory examination was difficult because of the gross adipose tissue.

Neurological examination revealed right sciatalgia, right L5 and S1 motor loss and diminished ankle jerk reflex. Lasègue's test was negative, the pain was not exacerbated by back movements or coughing, and there was no pain relief under conservative therapy or rest

Thigh CT-scan and MRI showed a large tumor, 70/80/75 mm in diameter, well-defined, encapsulated, having a thin fibrous capsule, contrast-enhancing mass, located in the middle third of the posterior thigh. The tumor had intimated relation with the sciatic nerve. The tumor displaced, but did not infiltrate surrounding muscles. (Fig. 1a, 1b, 1c)

Inferior limb angiography also confirmed the existence of a thigh tumor. (Fig. 2a, 2b)

Laboratory tests revealed thrombocytosis. The patient was further investigated, and she was diagnosed with chronic myeloproliferative syndrome. Surgery was postponed and the patient underwent oncologic therapy.

After proper treatment and normalization of blood test, the patient underwent surgery.

Patient received general anesthesia. Patient's positioning was supine position.

We performed a linear midline skin incision on the posterior surface of the thigh. Muscles were divided, and we found a well-circumscribed, encapsulated, tumor, originating from the sciatic nerve, growing along the sciatic nerve fascicles. The tumor was solid, greywhite, measuring 70/80/75 mm in diameter. The sciatic nerve was infiltrated by the tumor on 4 cm length. We exposed the sciatic nerve proximal and distal and isolated the tumor. The tumor had no propensity for surrounding tissue infiltration. We opened the sciatic sheath in a longitudinal manner, and carefully dissected the tumors along the nerve fascicles, which were incorporated within the tumor capsule and displaced circumferentially by the tumor.

We preserved the anatomic integrity of the sciatic nerve. We achieved a subtotal resection (aproximatively 90%) in order to avoid supplementary nervous fascicles sacrifice, dislocated by the tumor and to reserve the radical solution for regrow or malignization.

Histopahological examination found cellular schwannoma, associated with muscular, fibrous and adipose tissue; surrounding muscular tissue presented chronic inflammation (chronic miositis) and peritumoral vascular changes.

Outcome was slowly favorable. Right L5 and S1 motor deficits persisted postoperative.

On follow-up examination in 1 year, the patient presented no residual pain, no sensory loss, no trophic disturbances, and with residual motor loss (presented preoperative). The patient was able to walk by means of orthosis.

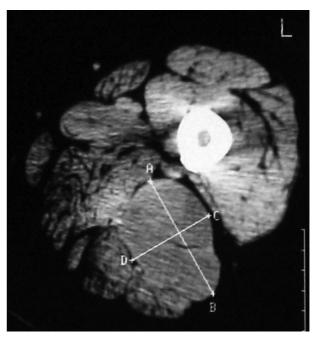


FIG. 1A

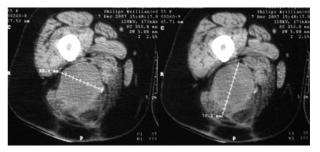


FIG. 1B

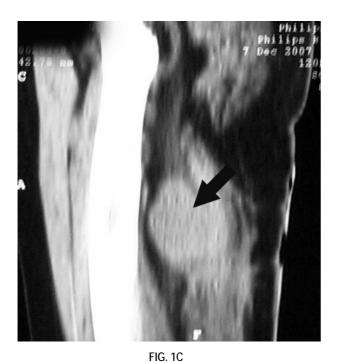


FIG. 1A, 1B, 1C Preoperative contrast CT-scan of the tight,

revealing anatomical position and tumor size

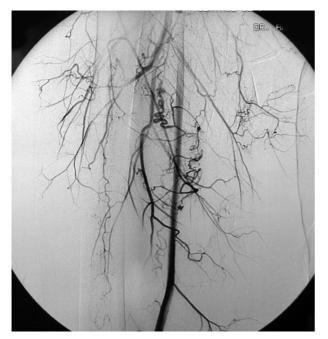


FIG. 2A



FIG. 2B

FIG. 2A, 2B Preoperative femoral angiography, revealing tumor blush and arterial pedicles

Control thigh MRI at six and twelve moths showed no evidence of tumor regrowing. (Fig. 3a, 3b)

DISCUSSION

Clinical presentation

Schwannomas have a long subclinical course and their clinical presentation is usually misleading. Radicular pain or sensory or motor loss are often misdiagnosed for radicular pain secondary to degenerative spine pathology.[7;8]



FIG. 3A

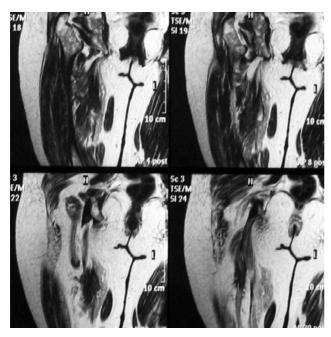


FIG. 3B

FIG. 3A, 3B Postoperative MRI with contrast one year after surgery, revealing the absence of the tumor mass and the sciatic nerve in continuity

Detailed history, with long lasting radicular pain, unresponsive to anti-inflammatory therapy and rest, and physical examination absence of lumbar contracture, a negative Lasègue's test, and lumbar spine IRM showing no signs of disc herniation, raise suspicion of non-discogenic sciatica and peripheral nerve pathology.[5;7;9] In such cases MRI imaging of the entire limb confirm the diagnosis of a peripheral nerve tumour.[2] Unresponsive pain and an irrelevant spinal MRI exam must conduct investigations in searching other nerve pathology.

EMG and ultrasound may, also be useful in establishing the diagnostic of sciatic schawannoma.[10]

Tumor characteristics

The real tumor incidence is unknown, being approximated by some studies at 0.6 cases per 100,000 people. From all benign sciatic tumors 60% are represented by neurofibromas and 38% schwannomas, the rest having other histological types.

Cellular schwannomas are slow growing tumors, and can achieved large size in time. The patients had a

long history of radicular pain and motor loss prior diagnostic.[6]

Associated pathology

Patient was operated twice in a different department, and both times the operation was stopped due to excessive bleeding.

After this unfortunate event, the patient is found on routine preoperative laboratory examination with elevated thrombocytes level and was further diagnosed investigated and with chronic myeloproliferative syndrome. This association is very unfortunate, carrying surgical pitfall. In spite of high thrombocytes level, coagulation was improper and did not allow surgery. In this situation, surgery must be postponed, and stabilizing the patient should be the treatment of choice. After stabilizing the patient surgery can be performed safely and effective. This association is casual, but it highlights the fact that patients carry a multitude of diseases, many having surgical consequences. Preoperative careful clinical and paraclinical examination is mandatory. The operation is extremely demanding and any comorbidity can compromise the result.

Differential between schwannoma and neurofibroma

Usually, schwannomas are solitary, well-circumscribed, encapsulated tumors, eccentrically located on nerve roots, originating from proximal nerves or spinal nerve roots. Neurofibromas are multiple, they lack tumor capsule, originate from distal nerves, causing fusiform enlargement of distal nerves.[2] Schwannomas arise from a single fascicle, and grow displacing circumferentially the other fascicles within the nerve sheath. Neurofibromas arise from perineural fibrocytes, cells having many histological similarities to Schwann cells.[1]

Schwannomas usually originate from the sensory fascicles in mixed nerves, while neurofibromas arise from motor parts. Plus, in neurofibromas fascicular bundles are more intimately involved, unlike schwannomas.[1]

Outcome

Malignant transformation in schwannomas is rare, the risk of malignization being approximated at 18% in neurofibromatosis type 1, and 5% in schwannomas.[11]

Patients with von Recklinghausen disease carry a worse outcome. Von Recklinghausen disease, neurofibromatosis associates multiple nerve sheath tumors. Patients may have simultaneous different histological types of nerve tumors, like schwannomas, neurofibromas, and malignant nerve sheath tumors.[1]

Surgery

Surgery is the treatment of choice in peripheral nerve schwannomas. During surgery, after finding the tumor, adequate exposure of the nerve, proximal and distal to the tumor, and tumor poles are mandatory. Nerve and tumor must be careful isolated and dissected from all adjacent neurovascular and soft tissue structures. Only after this wide exposure, the nerve sheath is opened and nerve fascicles are divided and tumor is resected. Nerve fascicles stretch over the tumor capsule. The surgeon must identify all intact fascicles over the capsule. Longitudinal dissection of the nerve sheath is prohibited because it cuts off vascular supply.

In small tumors, extracapsular resection, (under magnification) with gentle dissection of nervous fibers from the tumor capsule is the treatment of choice. In large tumors extracapsular excision cannot be achieved due to tumor size and an intracapsular debulking is needed first. In cases in which functional fascicles cannot be dissected from the capsule is wiser to leave a residual tumor than produce postoperative deficits. We prefer performing a subtotal resection with neurologic function preservation and reserve the radical surgery for tumor regrowth or malignant evolution.

Surgery in peripheral nerve sheath tumors, tend to be extremely satisfactory, when strict surgical principles are applied.[12] In the great majority of peripheral nerve schwannomas, total tumor resection without residual significant neurologic deficits can be achieved.[1;6;12]

CONCLUSIONS

Diagnostic is challenging, in many cases late, often symptoms are attributed to degenerative spinal

pathology. Any persistent sciatalgia, with irrelevant lumbar MRI must raise the suspicion of a peripheral sheath tumor, and must be further investigated. Surgery is very demanding and any associated comorbidities must be treated prior surgery. Preserving the integrity of the sciatic nerve, together with fascicles which are not infiltrated by the tumor, assure favorable outcome, without any additional neurological deficits.

REFERENCES

- 1. Lamond RC, Fox B. Management of peripheral nerve tumors. Limb Preservation 11. 2004.
- 2. Maraziotis T, Panagiotopoulos T, Panagiotopoulos E, Panagiotopoulos K. Neurilemoma of the popliteal fossa: report of two cases with long subclinical course and misleading presentation. Acta Orthop Belg 71, 496-499. 2005.
- 3. Ghaly RF. A posterior tibial nerve neurilemoma unrecognised for 10 years: case report. Neurosurgery 48, 668-672. 2001.
- 4. Gabhane SK, Kotwal MN, Bobhate SK. Morphological spectrum of peripheral nerve sheath tumors: A series of 126 cases. Indian J Pathol Microbiol 52, 29-33. 2009.
- 5. Blanchard C, Dam-Hieu P, Zagnoli F, Bellard S. Chronic sciatic pain caused by sciatic nerve schwannoma. Rev Med Interne 29, 748-750. 2008.
- Nawabi DH, Sinisi M. Schwannoma of the posterior tibial nerve: the problem of delay in diagnosis. J Bone Joint Surg Br 89, 814-816. 2007.
- 7. Kralick F, Koenigsberg R. Sciatica in a patient with unusual peripheral nerve sheath tumors. Surg Neurol 66, 634-637. 2006.
- 8. Rafai MA, El Otmani H, Rafai M, Bouhaajaj FZ, Largab A, Trafeh M, Adil A, Kadiri R, Slassi I. Peroneal nerve schwannoma presenting with a peroneal palsy. Rev Neurol
- 9. Kulcu DG, Naderi S. Differential diagnosis of intraspinal and extraspinal non-discogenic sciatica. J Clin Neurosci 15, 1246-1252. 2008.
- 10. Padua L, Commodari I, Zappia M, Pazzaglia C, Tonali PA. Misdiagnosis of lumbar-sacral radiculopathy: usefulness of combination of EMG and ultrasound. Neurol Sci 28, 154-155. 2007.
- 11. Weiss SW, Glodblum JR. Benign tumors of peripheral nerves and malignant tumors of peripheral nerves. In Enzinger and Weiss Soft Tissue Tumors. Mosby: St. Louis;
- 12. Oberle J, Kahamba J, Richter HP. Peripheral nerve schwannomas an analysis of 16 patients. Acta Neurochirurgica 139, 949-953. 2005.