

Schwannomatosis in pelvic region topography with neuro-muscular complications: A case report

Marco Orsini, Antônio Marcos da Silva Catharino, Valéria Camargo Silveira, Maria Luiza Santos Costa, Mauricio de Sant' Anna Junior, Roberta Araújo de Arruda Camara

ABSTRACT

Introduction: Schwannomatosis is a rare type of neurofibromatosis, usually affects people between ages 25 and 30 years old. Schwannomatosis causes tumors to develop on the cranial, spinal, and peripheral nerves. The most common symptoms are: chronic pain, which can occur anywhere in the body and can be disabling, numbness or weakness in various parts of the body, and muscle paresis. Two genes are known to cause schwannomatosis. Mutations of the genes SMARCB1 and LZTR1, which suppress tumors, are associated with this type of neurofibromatosis. We report the case of a patient victimized by schwannomas in the pelvic region and, as a consequence, neuromuscular injuries.

Case Report: Patient under investigation of lesions in the lumbosacral plexus and clinical picture marked by pain in the iliopsoas muscle in the left pelvic region and ipsilateral involvement of the obturator nerve (mild paresis in the adduction of the thigh and allodynia in the anterolateral region). A positron emission tomography-

computed tomography (PET-CT) study was carried out to assess the nodular findings. Elongated and nodular lesions were observed, with attenuation of soft tissues, located laterally to the left iliopsoas muscle. In the biopsy of one of the nodules, the result was compatible with schwannomas. Immunohistochemical assay confirmed the schwannomas to be positive for Sox10.

Conclusion: The patient, in this case, will be surgically approached due to the topography of the lesion, which currently causes refractory pain and impaired mobility.

Keywords: Neuromuscular injuries, Pelvic topography, Schwannomatosis

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Marco Orsini¹, Antônio Marcos da Silva Catharino², Valéria Camargo Silveira², Maria Luiza Santos Costa³, Mauricio de Sant' Anna Junior⁴, Roberta Araújo de Arruda Camara²

Affiliations: ¹Department of Neurology – UNIG and Coordinator of the Academic Master's Degree in Neurology at the University of Vassouras, RJ, Brazil; ²Professor at Iguaçú University – UNIG – Hospital Geral de Nova Iguaçu, RJ, Brazil; ³Medical student and member of the editorial team of the Journal of Biological and Health Sciences of the Universidade Iguaçú, RJ, Brazil; ⁴Federal Institute of Education, Science and Technology of Rio de Janeiro – IFRJ, Rio de Janeiro-RJ, Brazil.

Corresponding Author: Antônio Marcos da Silva Catharino, Rua Gavião Peixoto 70, Room 811, CEP 24.2230-100, Icaraí, Niterói-RJ, Brazil; Email: neurocurso@globo.com

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INTRODUCTION

Benign schwannoma is a tumor arising from Schwann cells (forming the neural sheath of peripheral nerves). The retroperitoneal location is unusual (0.5–5% of cases). The most common locations are cranial nerves (especially the vestibular nerve) and, in the peripheral nervous system the neck, mediastinum, and extremities [1].

Primary multiple obturator nerve schwannomas originate from Schwann cells and are extremely rare. Patients with schwannomas are asymptomatic and a retroperitoneal schwannoma is often misdiagnosed as

an adnexal mass [2]. An association between obturator nerve injury and iliopsoas muscle is also poorly reported.

Pelvic computed tomography (CT) or magnetic resonance (MR) imaging helps to limit diagnostic hypotheses. The resection of this tumor is the appropriate treatment, even though it is really a complex one. Prognosis is quite good since post-surgical recurrences are unusual. Complete resection is the best treatment for retroperitoneal pelvic schwannoma and **nowadays** it can be performed also by laparoscopy. Partial resection can be used when the mass is strongly connected to essential organs in order to prevent iatrogenic harms (neural deficit, vessel lesions); this may occur in 10% of cases [1].

Differential diagnosis must always be considered; in view of this, biopsy and histopathological report are necessary [3]. We report the case of a patient with two schwannomas in the pelvic region and neuromuscular involvement. Clinical control was not efficient in managing neuropathic pain and in slowing muscle weakness in the iliopsoas muscle.

CASE REPORT

A 51-year-old female patient was under investigation for lumbosacral plexus injuries. The clinical picture was marked by pain in the topography of the iliopsoas muscle on the left and involvement of the obturator nerve (mild paresis in the adduction of the thigh and allodynia in the anterolateral region) in the same limb. A PET-CT study was performed to assess the lesions. Despite the possibility of false positive result, the tomographic images of the whole body were acquired in a hybrid PET-CT device with a 16-channel multislice CT, after the administration of venous contrast.

Metabolic findings: The fusion of metabolic images (PET) to anatomical images (CT) showed increased contrast enhancement in elongated and nodular lesions, with soft-tissue attenuation: two lesions located laterally to the left iliopsoas muscle, the largest measuring 10 × 8 mm; medially to the left iliopsoas muscle, measuring 24 × 23 mm (Figure 1A and B). It presents heterogeneous enhancement by the iodinated contrast medium. There was also a small lesion in the intraperitoneal fat on the left flank, in the plane of the iliac crest, measuring about 20 × 12 mm (Figure 1C).

Impression: Glycolytic hypermetabolism on elongated and nodular images, most adjacent to the left iliopsoas muscle, that may correspond to neurofibromas or schwannomas.

She was treated the use of duloxetine 60 mg and gabapentin 400 mg each 8 hours. Occasionally, it is necessary to use anti-inflammatory drugs due to the clinical condition of "psoriasis" and, consequently, difficulties in performing flexion of the thigh over the hip. She also has neuropathic pain due to the involvement of the obturator nerve.

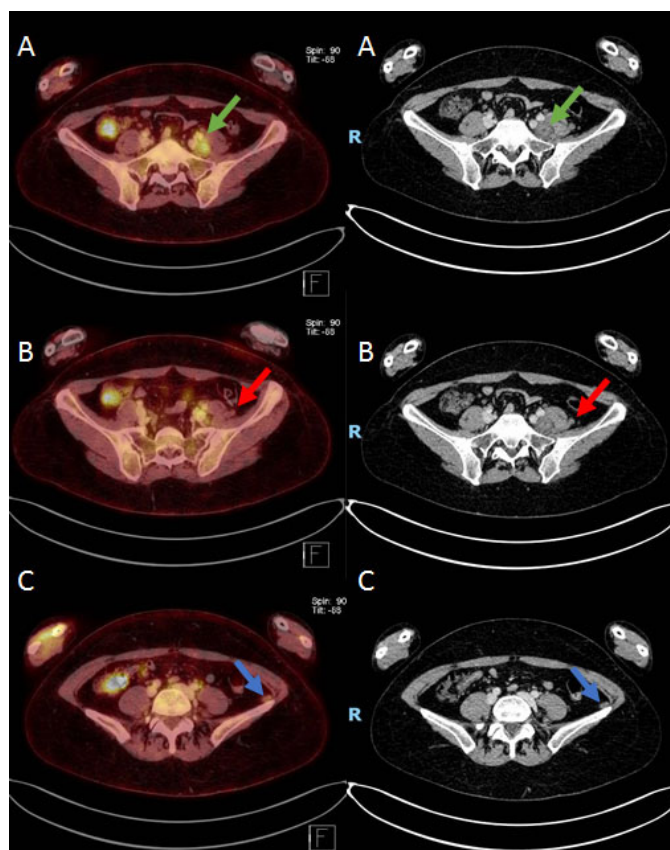


Figure 1: Left side – Fusion of metabolic images (PET), Right side – anatomical images (CT). (A) and (B) Two lesions with increased contrast enhancement, located medially (green arrows) and laterally to the left iliopsoas muscle (red arrows). (C) Small lesion in the intraperitoneal fat on the left flank, in the plane of the iliac crest (blue arrows).

DISCUSSION

Schwannomas are usually common benign neoplastic lesions that occur due to the proliferation of Schwann cells in the cranial nerve sheath, spinal roots, and peripheral nerves. Pelvic location in the region of the obturator nerve and iliopsoas muscle is uncommon [4]. We present the case of a patient with associated findings of peripheral nerve and muscle involvement and, unfortunately, refractory to conservative treatment.

Differential diagnosis includes fibromatosis, spindle cell sarcomas, and eventually ependymoma. However, the histological picture combined with the immunohistochemical study is essential for diagnostic elucidation. Immunohistochemical assay of our patient confirmed the schwannomas to be positive for **Sox10** [3, 4].

Reports in the literature have presented the feasibility of a minimally invasive resection of retroperitoneal or pelvic schwannomas. However, there are only a few reports in the literature about a robot-assisted nerve-sparing approach toward obturator schwannomas [5]. The patient in this case was instructed to perform this type of surgical intervention, although we have other techniques that are no less satisfactory.

CONCLUSION

The presence of schwannoma is an uncommon retroperitoneal pelvic tumor, and it can be misdiagnosed as an adnexal mass. To our knowledge, this is a rare case of two schwannomas arising from the obturator nerve and iliopsoas muscle. It usually does not have any neurological deficit, unlike the present clinical case (marked by difficulties in flexing the thigh due to injury and adduction of the hip).

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Author Contributions

Marco Orsini – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Antônio Marcos da Silva Catharino – Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Valéria Camargo Silveira – Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be

accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Maria Luiza Santos Costa – Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mauricio de Sant' Anna Junior – Acquisition of data, Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Roberta Araújo de Arruda Camara – Acquisition of data, Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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ABOUT THE AUTHORS

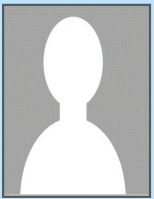
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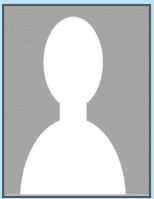
Marco Orsini, Full Professor at Universidade Iguaçú and Vassoras University, Rio de Janeiro, Brazil. He earned the undergraduate degree in Medicine from Rio de Janeiro Federal University/IPUB/Rio de Janeiro/Brazil and postgraduate degree from Fluminense Federal University, Department of Medicine Niterói, RJ, Brazil. He has published 420 articles/papers in national and international academic journals and authored 2 books. His research interests include Motor Neuron Diseases, Pain, and Peripheral Neuropathies. Now he is continuing his PhD in Rio de Janeiro from Federal University and Baylor. Email: orsinimarco@hotmail.com



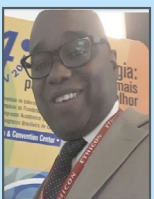
Antônio Marcos da Silva Catharino is a Physician, Neurologist, PhD student in Neurology at the Federal University of the State of Rio de Janeiro - UNIRIO, Adjunct Professor of the Medicine course at the Iguaçú University, Iguaçú University - UNIG/RJ, Nova Iguaçú, Brazil. Orcid: <https://orcid.org/0000-0002-5736-1486> Email: neurocurso@globo.com



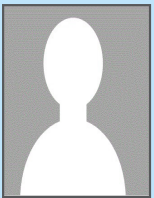
Valéria Camargo Silveira is a Professor at Iguaçú University – UNIG – Hospital Geral de Nova Iguaçú, RJ, Brazil.



Maria Luiza Santos Costa is a Medical student and member of the editorial team of the Journal of Biological and Health Sciences of the Universidade Iguaçú, RJ, Brazil.



Mauricio Sant' Anna Junior is a Physiotherapist, Doctorate in Medical Sciences from the Fluminense Federal University - UFF. Adjunct Professor at the Federal Institute of Education, Science and Technology of Rio de Janeiro (IFRJ), Lead researcher of the Study Group on Rehabilitation in High Complexity (GERAC) working in the line of research Physical-functional aspects in In-hospital rehabilitation.



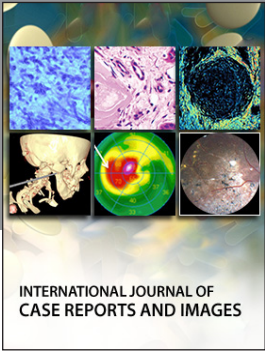
Roberta Araújo de Arruda Camara is a Professor at Iguaçú University – UNIG – Hospital Geral de Nova Iguaçú, RJ, Brazil.

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