



## Case report

# Malignant Triton tumor: a rare cause of sciatic pain and foot drop<sup>☆</sup>



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## ABSTRACT

Malignant peripheral nerve sheath tumors (MPNST) are very rare and are frequently localized in the buttocks, thigh, arm, or paraspinal region; one variant is the malignant Triton tumor, with rhabdomyosarcomatous differentiation. The authors present a challenging differential diagnosis of a sciatic pain and foot drop in a woman with history of lumbar disk herniation, which was found to be caused by a Triton tumor of the sciatic nerve. She underwent surgical excision, followed by radiation and chemotherapy. Malignant Triton tumor cases have rarely been described and reported in the literature. The recommended treatment is radical excision followed by high-dose radiotherapy and chemotherapy. The prognosis, although poor, depends on the location, grade, and completeness of surgical margins.

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## Tumor de Triton maligno: uma causa rara de dor ciática e pé pendente

## RESUMO

Os tumores malignos da bainha dos nervos periféricos (TMBNP) são muito raros e localizam-se mais frequentemente na região nadegueira, paraespinal, coxa ou braço; uma variante é o tumor de Triton maligno, com uma diferenciação rhabdomyosarcomatosa. Apresentamos um diagnóstico diferencial desafiante de dor ciática e pé pendente em uma paciente com antecedentes de hérnia discal lombar, que se descobriu que era causada por um tumor de Triton do nervo ciático. A paciente foi submetida a excisão cirúrgica, seguida de radio e

## Palavras-chave:

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quimioterapia. Poucos casos de tumores de Triton malignos foram descritos e relatados na literatura. O tratamento recomendado é a excisão radical, seguida de radioterapia em alta dose e quimioterapia. O prognóstico, embora mau, depende da localização, do grau e das margens cirúrgicas da exérese.

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## Introduction

Malignant tumors that originate or differentiate from the various nerve sheath lineages are collectively termed malignant peripheral nerve sheath tumors (MPNST); they represent a small percentage of soft tissue sarcomas, and may be sporadic or associated with neurofibromatosis type 1.<sup>1,2</sup>

They are often located in the buttocks, thighs, arms, or the paraspinal region. Most are high-grade sarcomas, which often metastasize to the lungs or bone; a variant of this rare type is the malignant triton tumor, which has rhabdomyosarcomatous differentiation.<sup>1,3</sup>

## Case report

This study reports the case of a 45-year-old female patient who had a history of low back pain and sciatic pain with progressive aggravation in the last six months. Axial CT scan indicated left L5-S1 disk herniation (Fig. 1). Her physician referred her to an Orthopedic Surgery specialist and while awaiting for the visit she got intramuscular non-steroidal anti-inflammatory drugs (NSAIDs) prescriptions.

However, in the previous two weeks, the patient developed a painful swelling in the left buttock that she believed to be related to the NSAIDs injections, attending the Orthopedic Emergency Room.

Physical examination indicated a hard swelling in the left buttock, hypoesthesia of the left leg with no specific territory, a positive Lasègue sign, foot drop gait, and absence of Achilles reflex.

An ultrasound examination of the buttock was performed, which indicated a vascularized mass in the left gluteal region. Magnetic resonance imaging (MRI) confirmed a heterogeneous lesion of 10.5 × 4.5 cm, deep in the gluteal muscles, that entered the pelvic cavity through the greater sciatic notch (Figs. 2 and 3).

The patient underwent *tru-cut* biopsy, which revealed a malignant triton tumor; one month later, she underwent *en bloc* resection of the tumor and sciatic nerve roots. Two months after surgery, adjuvant radiotherapy (RT) and chemotherapy (doxorubicin) were initiated.

Histopathology indicated that the surgical resection was complete; together with the imaging data, the lesion was staged as a grade-III pT2bN0M0 tumor.

Four months after surgery, hypoesthesia and drop foot persisted; electromyography confirmed neurotmesis of the sciatic nerve. She was referred for physical medicine and

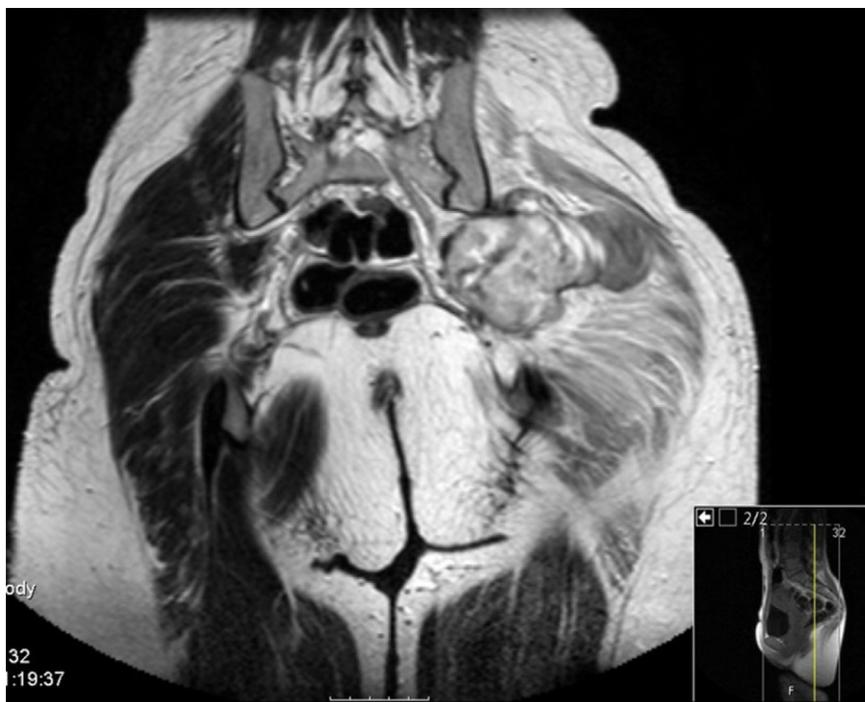


**Fig. 1 – Sagittal section of a T2-weighted lumbar spine MRI disclosing of L5-S1 disk herniation.**

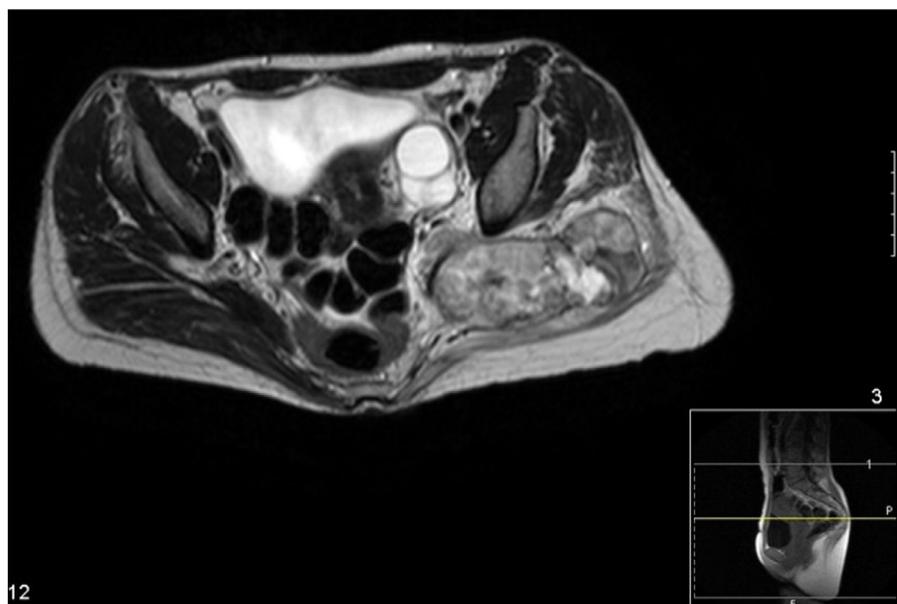
rehabilitation, followed the physiotherapy plan, and walked with an anti-equinus orthosis.

At eight months postoperatively, she presented dysesthesia and complained of pain at the level of the pelvis, irradiating to the left lower limb and refractory to analgesia. She also presented fever, dyspnea, constipation, and urinary retention. The patient was admitted; a new lumbar spine and hip MRI were requested, as well as a thoracic CT.

MRI identified structural changes at the level of the sacrum and iliac bones that could have been RT sequelae, but it was not possible to exclude metastatic lesions; nonetheless, no images compatible with local recurrence were observed (Fig. 4). The thoracic CT scan disclosed an image compatible with acute respiratory distress syndrome, as well as nodular lesions; once again, it was not possible to rule out metastatic disease (Fig. 5). A bone scan was requested, but it was not performed due to the increased respiratory insufficiency after an alveolar hemorrhage, requiring admission to the Intensive Care Unit.



**Fig. 2 – Coronal section of a T2-weighted pelvic MRI disclosing the lesion in the left gluteal region.**



**Fig. 3 – Axial section of a T2-weighted pelvic MRI identifying the lesion in the left gluteal region.**

At nine months post-operatively, she died of progressive respiratory failure and amine-refractory shock.

## Discussion

Traditionally, the diagnosis of MPNST has been difficult among soft tissue tumors due to the lack of standardized diagnostic and histological criteria. To date, there are no specific biomarkers that can help establish the diagnosis. The divergence in the

diagnostic criteria has also contributed to the variability of the incidence of such lesion described in the literature.<sup>3</sup>

MPNST occur mostly between the ages of 20 and 50, affecting both genders equally. In patients with neurofibromatosis type 1, these tumors tend to occur in males, at an earlier age, and to have larger dimensions.<sup>1-3</sup>

They present as swellings with progressive growth, and several months of evolution elapse prior to the diagnosis. Pain is an inconsistent finding. Lesions originating from the major nerves typically cause radiating pain, as well as motor and sensory deficits, which rarely precede the detection of

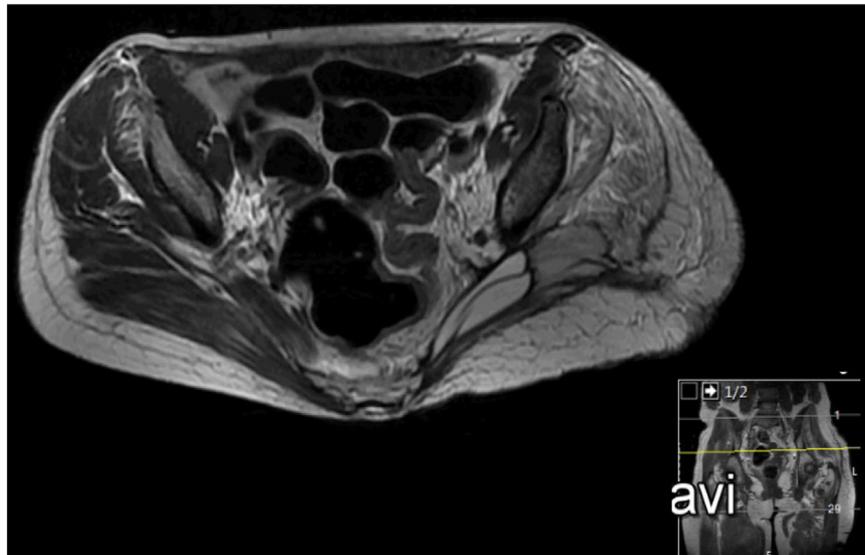


Fig. 4 – Axial section of a T2-weighted pelvic MRI in the postoperative period.



Fig. 5 – Axial cut of thoracic CT in the postoperative period.

swelling. The association of MPNST with the main nerve trunks explains its greater incidence in the proximal regions of the limbs and the trunk (sciatic nerve, brachial plexus, and sacral plexus).<sup>3</sup>

Histologically, these tumors are composed of hyperchromatic fusiform cells arranged in a fasciculated pattern. High-grade tumors usually contain areas of necrosis and increased mitotic activity.<sup>2,3</sup>

The capacity of MPNST to undergo mesenchymal differentiation is well known. The malignant triton tumor is a rare variant with rhabdomyosarcomatous differentiation, first described in 1938 by Masson and Martin. It consists of a stroma

typical of MPNST, with additional rhabdomyoblasts, which usually arise spherically and with eosinophilic cytoplasm.<sup>2,3</sup>

Few cases of malignant triton tumors have been described in the literature. As these are lesions with a high probability of local and distant recurrence, the recommended treatment is radical excision followed by high-dose radiation therapy, although more recent studies suggest that chemotherapy may eradicate micrometastases and extend survival.<sup>1,3</sup> During follow-up, CT and positron emission tomography are the exams of choice in the evaluation of relapses.<sup>2,3</sup> The prognosis, albeit poor, depends on the location, degree, and surgical excision margins; the five-year survival rate is 10%.<sup>1-3</sup>

The present case has some particularities that may have caused a delay in diagnosis and made it even more challenging. The history of low back pain with a documented left L5-S1 disk herniation could perfectly explain the symptoms, leading the attending physician to refer the patient to an orthopedic surgeon.

The distinctive feature of this clinical case was the swelling in the left buttock, which may have raised the clinical suspicion of another cause for clinical presentation.

*En bloc* resection of the tumor as well as the sciatic nerve and its roots contributed to postoperative motor and sensory deficits, but was a necessary procedure, since the tumor invaded the nerve. Complete surgical resection and absence of nodular or distant metastases would represent good prognostic factors. However, this type of tumor has a generally poor prognosis, which was confirmed in this case.

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### Conflicts of interest

The authors declare no conflicts of interest.

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