



GRAND ROUNDS

Malignant triton tumor: Grand Round presentation of a rare aggressive case thoracolumbar spine tumor

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Abstract



Introduction We report a rare and aggressive case of malignant triton tumor (MTT) at the thoracolumbar junction with foraminal extension mistreated as schwannoma.

Materials and methods A 70-year-old man with a 2-year history of lower back pain and left L4 radiculopathy with no history of neurofibromatosis.

Results Pre-operative MRI suggested a typical schwannoma. Upon complete marginal resection, histological findings revealed a MTT. The patient presented with a local and regional recurrence and died 10 months after surgery. MTTs are a subgroup of malignant peripheral

nerve sheath tumors, which develop from Schwann cells of peripheral nerves or within existing neurofibromas, and display rhabdomyoblastic differentiation.

Conclusion Based on the Grand Round case and relevant literature, we present a case of a highly aggressive and fast-growing tumor with a very high local and distant recurrence. There is no consensus treatment plan available and patients usually die shortly after diagnosis.

Keywords Malignant triton tumor · Malignant peripheral nerve sheath tumor · Spinal tumor

Case presentation

A 70-year-old man with a 2-year history of lower back pain and left L4 radiculopathy was admitted to our department. His radicular pain had increased gradually. The patient had a history of prostatic adenocarcinoma 7 years before, treated by surgery and hormone therapy. There was no personal or family history of neurofibromatosis. Upon examination, we found no systemic signs of neurofibromatosis (NF). The rest of the neurological physical examination was strictly normal and no signs of myelopathy or cauda equina syndrome were found.

Diagnostic imaging section

The CT-scan showed a degenerative lumbar spine and a left T12–L1 foraminal mass. An MRI of the thoracolumbar spine revealed an intradural mass at the T12–L1 level extending through the left T12 foramen into the paraspinal muscles (Fig. 1). The tumor size was 32 mm × 11 mm × 2 mm. The mass was homogenous with an intermediate signal on T1 and T2-weighted images. However, there was heterogeneous

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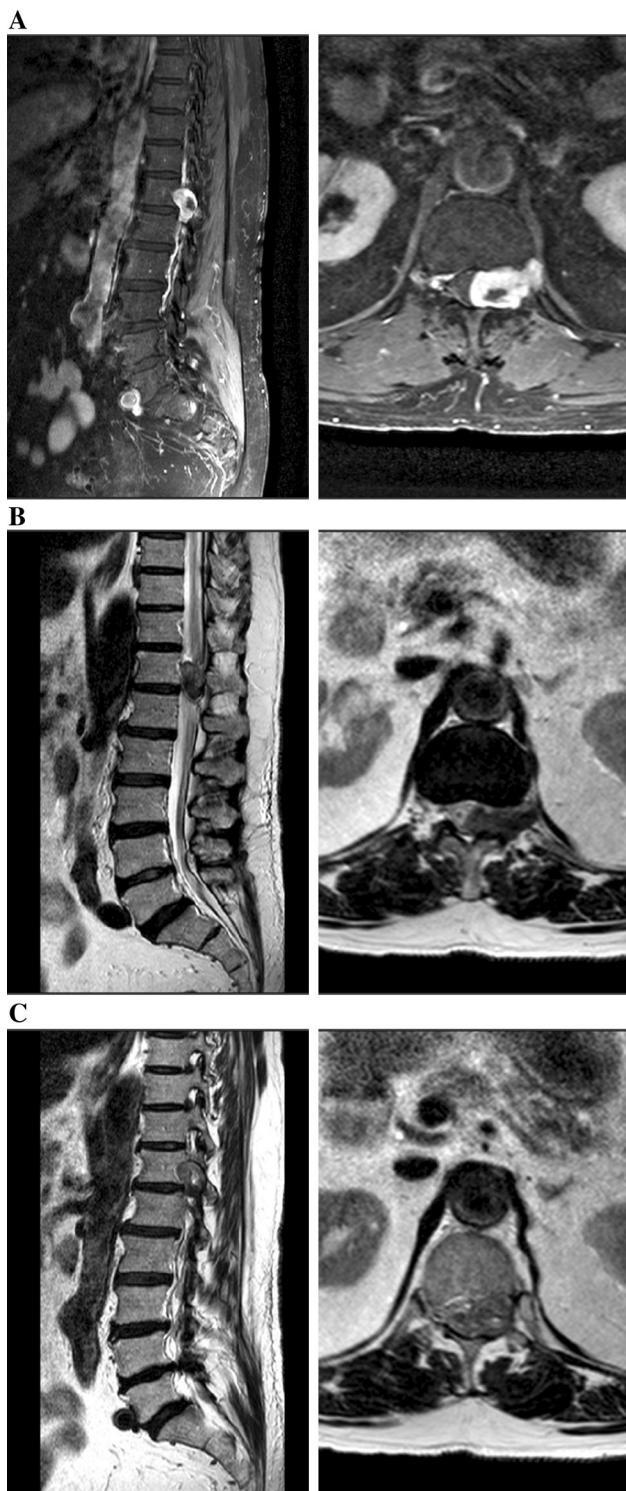


Fig. 1 Preoperative MRI: **a** T1-weighted FAT-SAT axial and sagittal views showing a hyperintense lesion with heterogeneous enhancement in the left T12-L1 foramen, **b** T2-weighted axial and sagittal views and **c** T2-weighted axial and parasagittal views through the invaded foramen

enhancing after IV gadolinium administration. Radiologist suggested the diagnosis of schwannoma developing from the left T12 root.

Procedure

Under general anesthesia, a posterior surgical approach to the tumor was performed. The patient underwent a T12 laminectomy and a medial T12-L1 facetectomy on the left side. Once the dura mater was opened, a well-encapsulated mass arising from the left T12 root was found. The gross tumor appearance resembled benign schwannoma. A complete marginal resection followed by a T12-L1 posterolateral fusion with autograft was performed (Fig. 2).

Histological analysis

The histological examination revealed a highly cellular cell proliferation (Fig. 3a). The tumoral cells are mainly fusiform, atypical, sometimes showing a nuclear pleomorphism. We observed ten mitoses for ten high power magnification and necrosis areas. Admixed in the proliferation, there were large cells with rhabdoid features such as central abundant eosinophilic cytoplasm and peripheral atypical nuclei (Fig. 3b). Immunohistochemical study showed that focally the fusiform cells expressed S100 protein and that rhabdoid cells expressed desmin (Fig. 3c, d). These cells did not express CKAE1/AE3, CD31, and EMA. The overall features of the lesion supported those of a malignant peripheral nerve sheath tumor with rhabdomyosarcomatous differentiation: malignant triton tumor.

Outcome, follow-up

The postoperative course was uneventful and the patient's back and leg pain were significantly reduced. As the patient's performance status (World Health Organization score) was favorable (0/5) postoperatively, he underwent adjuvant chemotherapy. A lumbar spine MRI performed at 3-month follow-up revealed a local and regional tumoral recurrence with multi-metastatic dissemination to the entire cauda equina (Fig. 4). PET-CT displayed hypermetabolism at the T12 and L4 levels. A rectal hypermetabolism was also detected (with no further endoscopic exam). Concomitantly, the patient developed febrile neutropenia due to the adjuvant treatment complicated with septic shock. Consequently, chemotherapy was discontinued and was substituted for palliative radiotherapy. The patient died 10 months after diagnosis.

Fig. 2 **a** Postoperative A-P X-ray. **b** Postoperative lateral X-ray

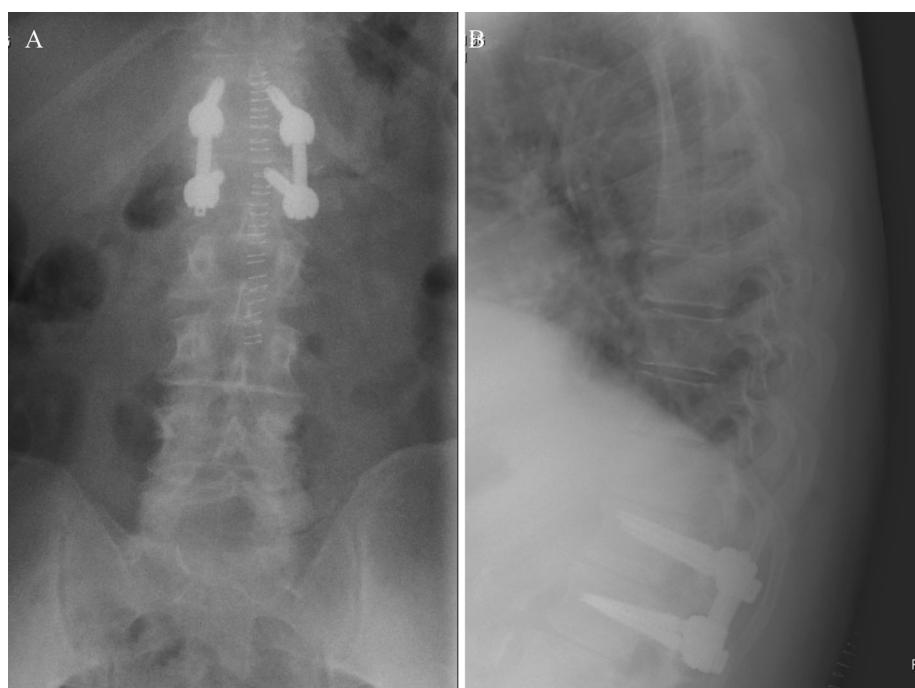
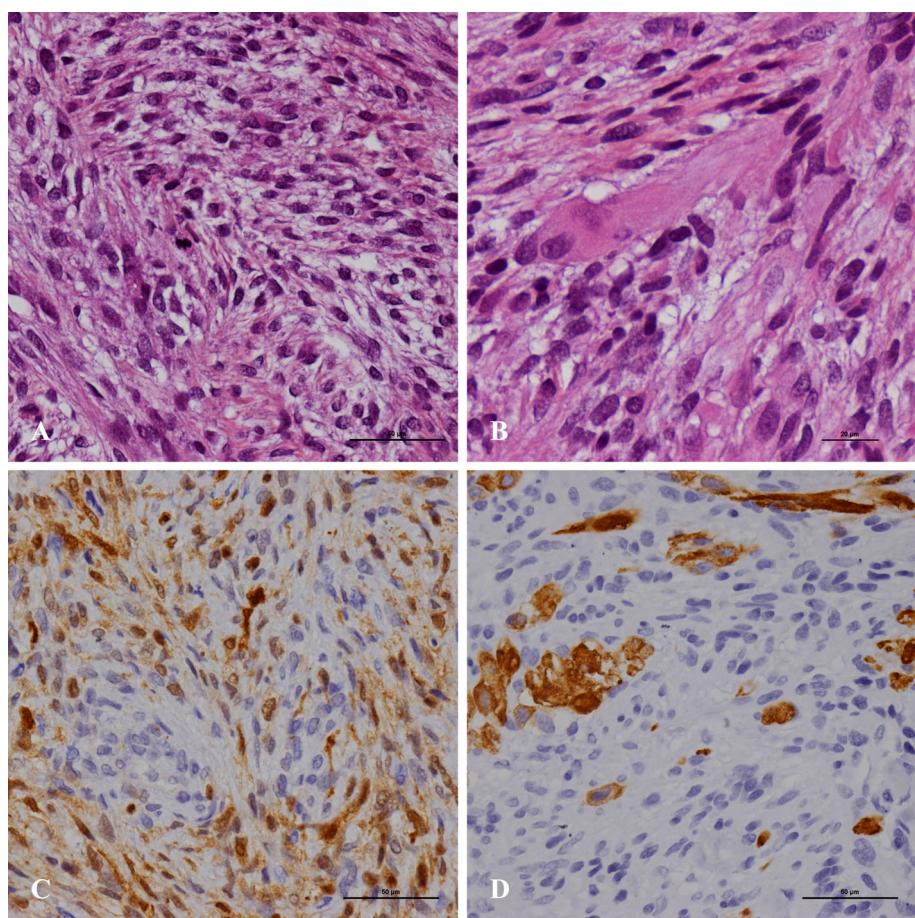


Fig. 3 **a** Malignant triton tumor: spindle cell component within a fibrous stroma. Hyperchromatic pleomorphic and plump nuclei and occasional mitoses (hematoxylin–eosin saffron). **b** Some cells have rhabdoid features (hematoxylin–eosin saffron). **c** Numerous cells express S-100 protein (immunohistochemistry). **d** Cells with rhabdoid features express desmin (immunohistochemistry)



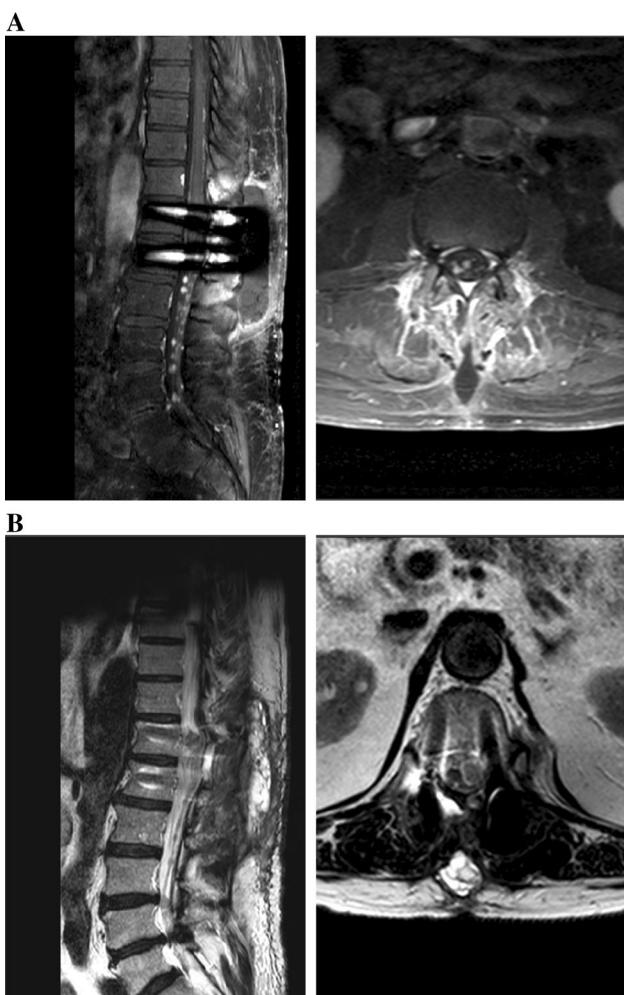


Fig. 4 MRI at a 3-month follow-up: **a** T2-weighted enhanced axial and sagittal views showing a regional recurrence, with multi-metastatic dissemination to the entire cauda equina and **b** T2-weighted enhanced axial and sagittal views showing a local recurrence

Description about the condition (condition, epidemiology, diagnosis, pathology, differential diagnosis)

Malignant triton tumor (MTT) is an extremely rare and aggressive type of sarcoma that arises from peripheral nerve sheaths with high local recurrence rate. It is a subgroup of malignant peripheral nerve sheath tumors (MPNSTs), which develop from Schwann cells of peripheral nerves or within existing neurofibromas, and display rhabdomyoblastic differentiation [1–3].

In 1932, Masson described rhabdomyosarcomatous elements within malignant peripheral nerve sheath tumors (MPNST) in patients with neurofibromatosis [4]. To date, less than 170 cases of MTT have been described in the literature. They represent only 5–10% of MPNSTs and are

associated to NF-1 disease in 50% of cases. The other 50% seem to be sporadic [5].

The histopathogenesis still remains unknown. Most common locations are the head, neck, extremities, and trunk [5, 6]. Mediastinal and retroperitoneal locations are less commonly described [5, 7].

Spinal MTT is difficult to manage, and is typically associated with a pejorative prognosis, similar to MTT in the trunk and buttocks [8].

The diagnosis of MTTs is made according to the histological analysis [5]. The clinical or radiological suspicion is very difficult due to its scarcity. In our case, we report a rare spinal location with foraminal extension in a 70-year-old man. Usually, patients with MTT are under 40 years old (mean age 39 years old) [6]. The clinical presentation was usual with back pain and leg pain, which was compatible with degenerative radiculopathy with no sensory or motor impairment. Imaging (CT scan and MRI) revealed an intradural mass at the T12–L1 level extending through the left T12 foramen. The radiologist wrongly suggested the diagnosis of schwannoma developed from the left T12 root because the tumor displayed benign imaging criteria.

Indeed, schwannoma commonly originates from the dorsal nerve root. Schwannoma is a nerve sheath tumor that is usually benign, although malignant subtypes are described. The peak incidence of these tumors occurs between the fourth or fifth decades of life, equally among men and women. This tumor may be associated with NF-2 with high risk of malignant transformation. The spinal location is variable: cervical spine (31%), cauda equina (24%), thoracic spine (22%), upper cervical spine (16%), conus medullaris (7%) [9]. The MRI appearance of schwannoma is the following: a solitary, well-circumscribed mass arising from a dorsal nerve root. Schwannoma can also feature intradural and extradural components. Classically, T1-weighted MR images display iso-intensity while T2-weighted images appear hyperintense; contrast enhancement is variable.

Yimaz et al. [8] reported a similar case of MTT in a 58-year-old man, which was mistreated as a benign tumor. Following histologic analysis, the patient was treated by radiotherapy with no recurrence at an 8-month follow-up. We believe that exhaustive and systematic reports of all cases worldwide are critical for the improvement of medical knowledge of MTTs (clinical course, treatment outcome, and prognosis).

Currently, there is no consensus regarding the treatment guidelines for MTTs. As for sarcomas, several authors recommend a first radical excision with wide margins followed by radiotherapy. Chemotherapy did not show high efficacy.

In this case, after close discussion between the radiologist and the referring clinician, a percutaneous biopsy

seemed not necessary. The clinical and radiological data suggested a benign diagnosis. Therefore, another preoperative investigation was not necessary. A complete marginal resection was decided and improved both back pain and leg pain. However, the patient relapsed 3 months later. Retrospectively, the results of a primary percutaneous biopsy could affect the subsequent management of the patient and could influence the surgical management (en bloc resection surgery). Despite palliative chemotherapy and radiotherapy, he died 10 months after diagnosis. James et al. reported a median survival of 12 months. Admittedly, the variation of outcome is due to a variation in location, tumor grade, and resectability [10].

While treatment guidelines for MTT are not consensual yet and in spite of initial diagnostic difficulties, we strongly advise radiologists and surgeons to rule out this diagnosis whenever possible before the surgical resection of schwannoma.

Compliance with ethical standards

Conflict of interest No funds were received in support of this work. No benefits in any forms have been or will be received from a commercial party related directly or indirectly to the subject of this manuscript.

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