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CASE REPORT

Malignant triton tumour of the distal femur: imaging features of a rare entity

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ABSTRACT

Malignant Triton tumour (MTT) is a rare rhabdomyoblastic variant of malignant peripheral nerve sheath tumour (MPNST), characterised by striated muscle differentiation, with an incidence of approximately 0.001%. We report a 40-year-old woman with a rapidly painful, enlarging mass of the left distal femur and associated palpable lesions in the left calf and abdomen. Multislice CT demonstrated multiple solid lesions with peripheral contrast enhancement infiltrating adjacent tissues. MRI showed a large (>5 cm) heterogeneous lesion measuring 9.4 × 8.4 × 9.2 cm with lobulated margins, heterogeneous contrast enhancement, and mass effect on surrounding structures. Diagnosis and treatment: Histopathology and immunohistochemistry confirmed the diagnosis of MTT. The patient underwent two wide tumour excisions followed by adjuvant chemotherapy. Despite treatment, chest radiography one and a half years later revealed pulmonary metastases. MTT is an aggressive tumour with rapid growth and a high propensity for early metastasis. Radiologic features that should raise suspicion include large size, lobulated margins, peripheral calcifications, and internal heterogeneity. Correlation of imaging with clinical and histopathological findings is essential for timely diagnosis and treatment planning.

1. Introduction

Malignant peripheral nerve sheath tumour (MPNST) is an uncommon soft-tissue sarcoma, accounting for approximately 5–10% of all sarcomas. MPNST may occur sporadically or in association with neurofibromatosis type 1 (NF1), a genetic disorder that predisposes to tumorigenesis in peripheral nerve tissue; NF1-associated lesions are usually benign but can transform into MPNST (Yao *et al.*, 2023). The overall incidence of sporadic MPNST is low, estimated at approximately 0.001% (Asahi *et al.*, 2018; James *et al.*, 2016; Yao *et al.*, 2023). MPNST arises from cells of the peripheral nerve sheath, including Schwann cells, perineurial cells, or fibroblasts (Yao *et al.*, 2023).

Malignant Triton Tumour (MTT) is a rare rhabdomyoblastic variant of MPNST, characterized histologically by malignant peripheral nerve sheath elements admixed with rhabdomyoblastic differentiation (Jack *et al.*, 2021). MTT represents about 5% of MPNST cases, with most reported patients being young to middle-aged adults; 25–50% of MTT cases are associated with NF1 (Jack *et al.*, 2021; Natalie Wu and Lu, 2019). Patients with NF1 carry an increased lifetime risk of MPNST—reported at approximately 8–13%—and MPNST accounts for a significant proportion of

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morbidity in this population (Natalie Wu and Lu, 2019).

MTT most commonly arises from peripheral nerves in the head and neck, extremities, or trunk, though rare sites such as the brain, buttock, internal organs, mediastinum, and retroperitoneum have been reported (Hou *et al.*, 2020; Chaudhry *et al.*, 2019; Zain *et al.*, 2021). Clinically and radiologically, MTT poses diagnostic challenges because of its rarity and nonspecific imaging appearance. Histologically, MTT is distinguished by rhabdomyoblastic differentiation and generally carries a worse prognosis and greater difficulty with complete resection than conventional MPNST (Jack *et al.*, 2021; Li *et al.*, 2022).

On MRI, MPNST and MTT commonly present as large, heterogeneous masses that are hyperintense on T2-weighted images and isointense to muscle on T1-weighted images; these features are not specific and often overlap with other soft-tissue tumours (Jiang *et al.*, 2024). On multislice CT (MSCT), lesions frequently appear as well-defined, lobulated soft-tissue masses with heterogeneous attenuation and peripheral post-contrast enhancement (Li *et al.*, 2022). Despite limited specificity,

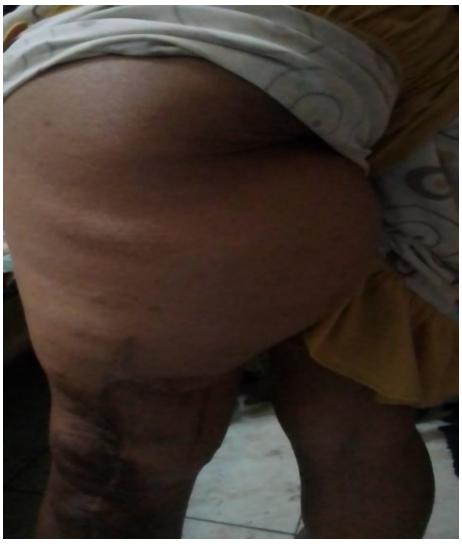

Figure 1. Clinical photograph showing a large, firm, fixed mass in the posterior left thigh measuring 11 × 12 × 16 cm

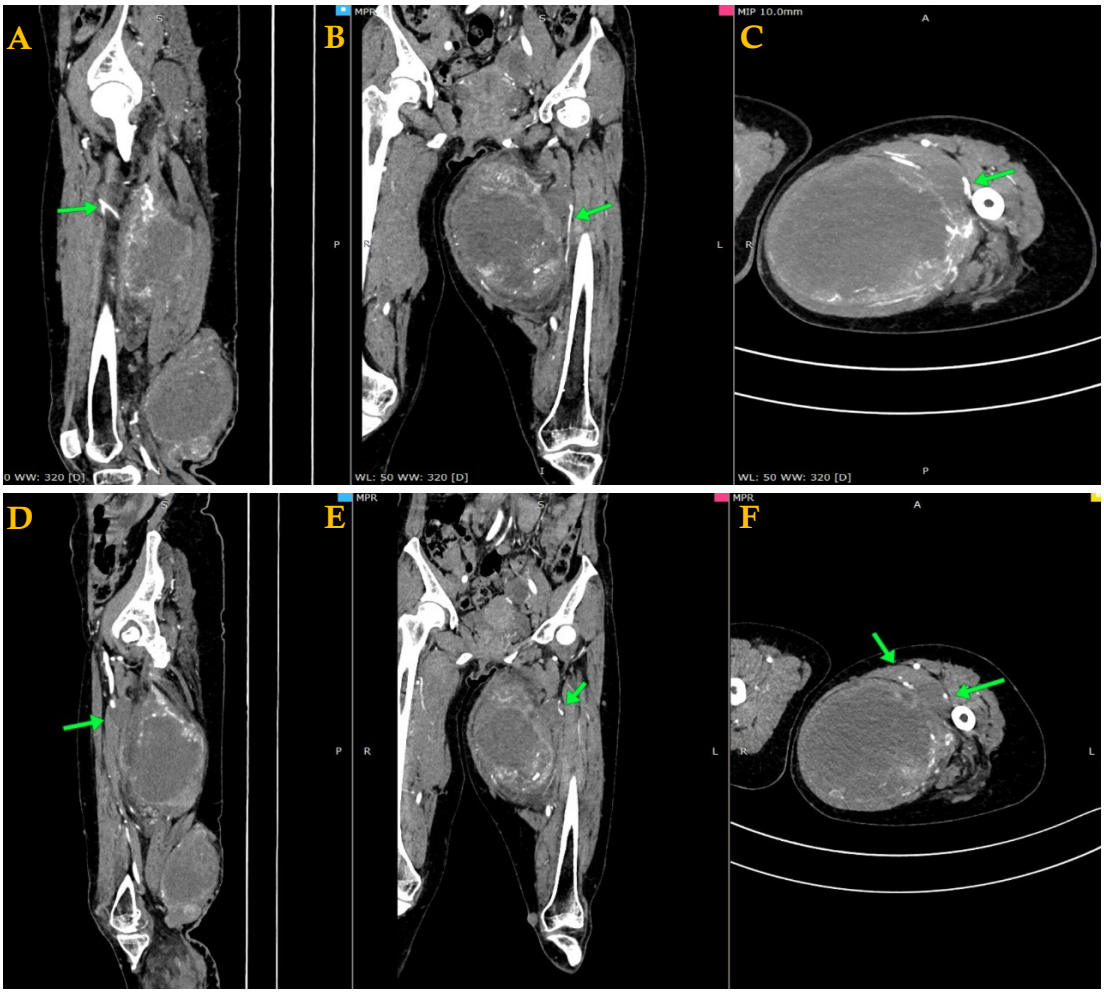

Figure 2. Contrast-enhanced MSCT: sagittal (A, C), coronal (B, D), and axial (E) views. The lobulated mass in the left distal femur demonstrates a solid component (31 HU) with peripheral contrast enhancement (75 HU), semisolid areas (26 HU), medial muscle infiltration (adductor longus, brevis, magnus, gracilis), and lateral displacement of the left deep femoral vessels and femur.

cross-sectional imaging with MRI and MSCT remains essential for lesion characterisation, preoperative planning, and assessment of local extension and metastatic disease.

Given the diagnostic and therapeutic implications, this study aims to evaluate the MRI and MSCT features of MTT arising in the distal femur and to highlight imaging signs that may facilitate recognition of this rare, aggressive entity.

2. Case

A 40-year-old woman was referred from an outside hospital with a progressive, painful mass involving the left thigh, left calf, and abdomen. She reported a two-year history of rapid enlargement and increasing pain from the posterior thigh to the calf, impairing daily activities. Her history dates back to age 6, when she first noticed that her left leg appeared larger than her right, with a palpable posterior-thigh lump. At age 18, the lesion enlarged further, spreading to the left forearm, left calf, and abdomen. There is no family history of tumours. She previously underwent resection of a left forearm lesion that was reported as diffuse neurofibroma on histopathology. On examination, a firm, fixed, skin-colored mass occupied the posterior left thigh, measuring approximately 11 × 12 × 16 cm, with tenderness radiating to the left calf (Figure 1).

On September 29, 2023, a multislice CT (MSCT) of the left lower extremity was performed with and without contrast. Images demonstrated multiple components: a solid portion with attenuation 31 HU and a semisolid component 26 HU, forming a well-defined, lobulated mass in the left distal femoral region. The lesion infiltrated the medial thigh muscles, including adductor longus, adductor brevis, adductor magnus, and gracilis. Post-contrast images showed peripheral enhancement of the solid component (peak ~75 HU). The mass displaced the left deep femoral artery and vein and caused lateral displacement of the left femur (Figure 2).

She underwent a chest radiograph on October 19, 2023, which showed no evidence of pulmonary metastasis. An initial wide excision was performed on October 24, 2023; the resected specimen measured 10 × 10 × 15 cm. Histopathology on October 31, 2023, reported a high-grade malignant mesenchymal tumour, with differential diagnoses including MTT,

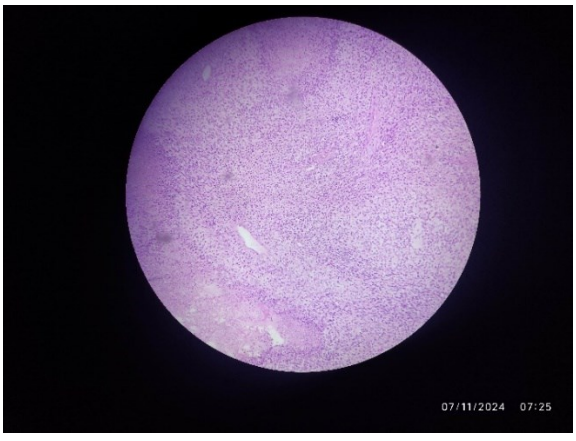

Figure 3. Histopathology (IHC) of the resected specimen demonstrates features of a high-grade malignant mesenchymal tumour.

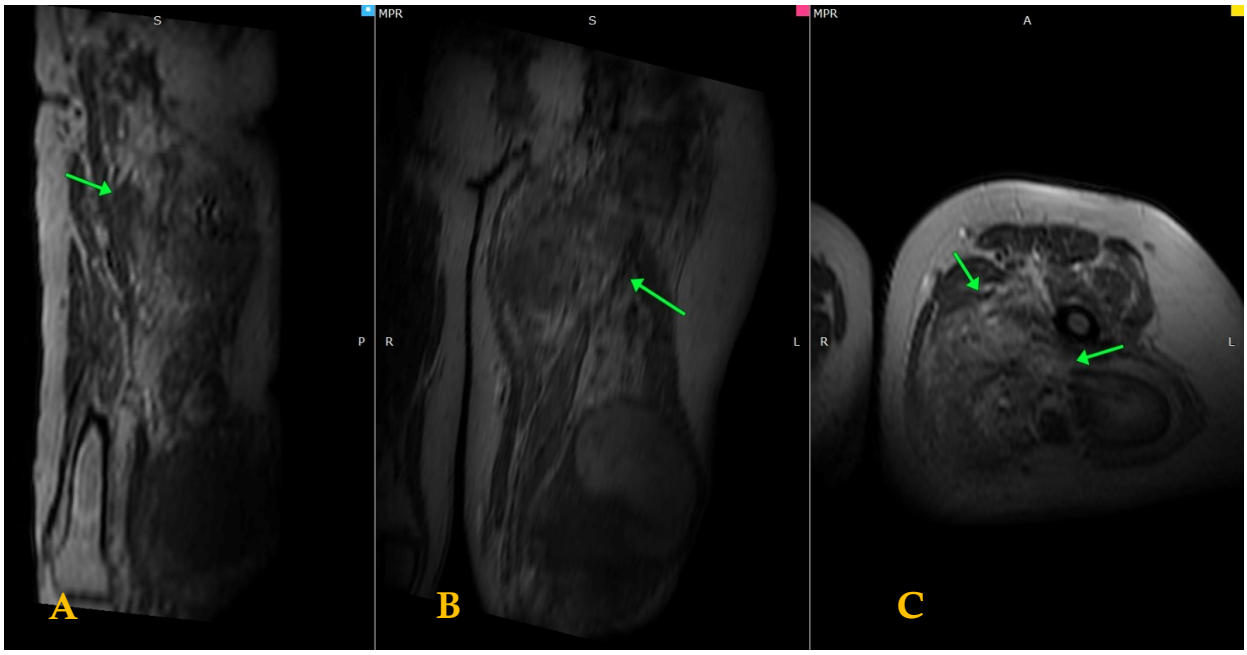

Figure 4. Contrast-enhanced MRI (T1WI) demonstrating multiple enhancing lesions in the left thigh infiltrating the adductor magnus, adductor longus, adductor brevis, and gracilis muscles (panels A–C).

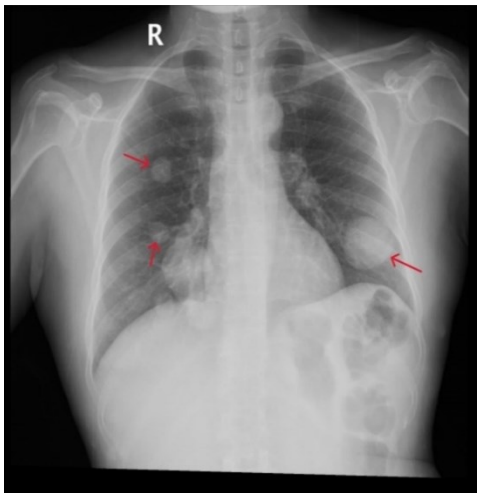

Figure 5. Chest X ray demonstrating multiple well defined, smooth margined, heterogeneous opacities of varying sizes in both hemithoraces, consistent with pulmonary metastases.

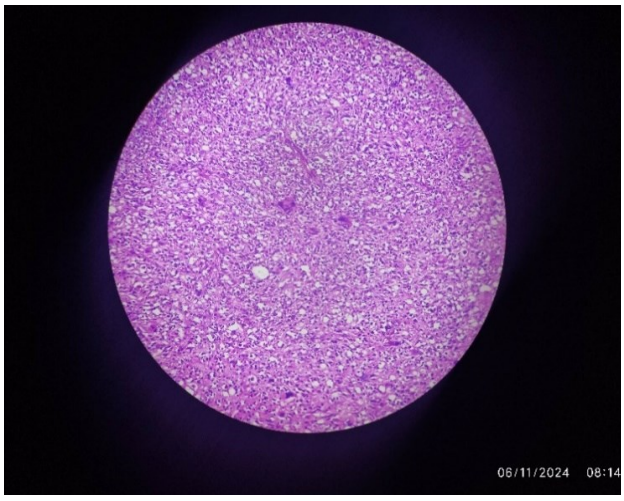

Figure 6. Histopathology (IHC) of the resected specimen from the second excision confirms a malignant Triton tumour with residual tumour at the resection margins.

MPNST, rhabdomyosarcoma, and liposarcoma. Immunohistochemical (IHC) analysis was therefore requested.

IHC performed on January 19, 2024, showed focal S-100 and desmin positivity, negative myogenin, and a Ki-67 index of approximately 40%, findings consistent with a malignant Triton tumour (Figure 3). The patient received eight cycles of adjuvant chemotherapy from February to July 2024 (Holoxan 6000 mg/m², Mesna 3600 mg/m², and epirubicin 50 mg/m² per cycle).

On September 5, 2024, an MRI of the left thigh was obtained for post-treatment assessment. Multiple well-defined, rounded soft-tissue lesions were identified along the mid to distal left femur, the largest measuring 9.4 × 8.4 × 9.2 cm (AP × CC × LL). The dominant lesion was iso- to mildly hyperintense on T1-weighted, T2-weighted, and T2-STIR sequences. Post-contrast images demonstrated heterogeneous enhancement. The mass infiltrated the left rectus femoris, vastus intermedius (lateral and medial portions), adductor magnus, adductor longus, adductor brevis, sartorius, and gracilis muscles, and it encased the left femoral artery and vein (Figure 4).

On October 19, 2024, chest radiography demonstrated multiple well-defined, smooth-margined, heterogeneous opacities of varying sizes in both hemithoraces, consistent with pulmonary metastases (Figure 5). Prior to this, she underwent a second wide excision of the left femoral mass on September 24, 2024; the resected specimen measured 20 × 15 × 16 cm. Histopathology on September 27, 2024, confirmed a malignant Triton tumour with residual disease (Figure 6). The patient was discharged with plans for clinical and radiologic follow-up at three months.

3. Discussion

Malignant Triton Tumour (MTT) is a rare rhabdomyoblastic variant of malignant peripheral nerve sheath tumour (MPNST) characterised by the coexistence of Schwann-cell–derived elements and rhabdomyoblastic differentiation (Jack *et al.*, 2021; Aykut *et al.*, 2016). The eponym originates from experimental work in Triturus salamanders demonstrating rhabdomyoblastic differentiation of nerve-derived tumour cells (Hashimoto *et al.*, 2025). Diagnostic criteria emphasise tumour development along peripheral nerves and histologic evidence of both Schwann cells and rhabdomyoblasts; identification relies on histopathology and immunohistochemistry (Jack *et al.*, 2021).

Epidemiologically, MTT accounts for a small proportion of MPNSTs, and may occur sporadically or in association with neurofibromatosis type 1 (NF1) (Zain *et al.*, 2021; Natalie Wu and Lu, 2019). Reported site distribution shows predilection for the trunk (32%), extremities (24%), and head and neck (20%), with less frequent involvement of mediastinum, heart, or lungs (Zain *et al.*, 2021). NF1-associated MTT tends to present in younger males, whereas sporadic cases more often affect older females (Zain *et al.*, 2021; Alwan *et al.*, 2024). A long latent period (often 10–20 years) between benign nerve-sheath lesions and malignant transformation has been described (Chaudhry *et al.*, 2019), consistent with our patient's prolonged history.

Cross-sectional imaging with MRI and multislice CT (MSCT) is essential for lesion mapping, staging, and surgical planning, although imaging appearances are

nonspecific and overlap with other high-grade soft-tissue tumours (Kazawa, 2018; Li *et al.*, 2022). Typical imaging features overlap substantially with high-grade MPNSTs: large size (often >5 cm), heterogeneous internal architecture reflecting necrosis, haemorrhage, or cystic change, lobulated or irregular margins, infiltration of adjacent musculature, and mass effect or encasement of neurovascular structures (Li *et al.*, 2022; Drews *et al.*, 2024). On MRI, lesions commonly show low-to-intermediate signal on T1WI and high signal on T2WI, with heterogeneous post-contrast enhancement (Jiang *et al.*, 2024). On CT, peripheral enhancement, heterogeneous attenuation, possible internal septations, and peripheral calcifications may be seen (Li *et al.*, 2022; Drews *et al.*, 2024). The absence of the classic target sign, rapid growth, necrosis, and aggressive invasion favours MPNST/MTT over benign peripheral nerve sheath tumours such as neurofibroma or schwannoma (Li *et al.*, 2022; Yao *et al.*, 2023). High-grade sarcomas without nerve continuity (e.g., undifferentiated pleomorphic sarcoma, synovial sarcoma) and metastatic disease are important differentials and must be excluded by correlation with clinical history and histopathology.

Definitive diagnosis of MTT requires histologic demonstration of a malignant peripheral nerve sheath tumour with rhabdomyoblastic elements. Immunohistochemistry typically shows focal S-100 positivity in Schwann-type cells and variable expression of muscle markers (desmin, myogenin) in rhabdomyoblastic components (Jack *et al.*, 2021). A high proliferative index (e.g., Ki-67) supports aggressive behaviour. In our case, focal S-100 and desmin positivity, along with elevated Ki-67, corroborated the diagnosis.

MTT has a poorer prognosis than conventional MPNST, with reported 5 year survival rates of 5–15% and a high propensity for local recurrence and distant metastasis (Zain *et al.*, 2021; Ban *et al.*, 2024; Chaudhry *et al.*, 2019). Radical surgical resection with wide margins remains the mainstay of treatment; adjuvant chemotherapy and radiotherapy are commonly employed, although their impact on long-term survival is uncertain (Bian *et al.*, 2019; Chaudhry *et al.*, 2019). Some series report limited chemoradiation responsiveness, while others describe tumour shrinkage after systemic therapy (Bian *et al.*, 2019). In our patient, chemotherapy produced a measurable reduction in tumour dimensions on MRI (from 11.1 × 12.6 × 17.1 cm to 9.4 × 8.4 × 9.2 cm) but did not prevent pulmonary metastases within 18 months despite repeated wide excisions, illustrating the aggressive natural history of MTT and the difficulty of achieving durable control.

This case underscores several key points: (1) a long history of nerve-sheath lesions with recent rapid growth should raise concern for malignant transformation; (2) MRI and MSCT are indispensable for mapping tumor extent, neurovascular involvement, and treatment response, even though imaging cannot replace histopathologic confirmation; and (3) early, aggressive surgical management with multidisciplinary oncologic input is critical given the high risk of recurrence and metastasis (Ghailane *et al.*, 2019; Tsagozis *et al.*, 2020). Radiologists should report features suggestive of high-grade nerve-sheath malignancy—large size, lobulated/irregular margins, internal heterogeneity, infiltration, and absence of the target sign—and recommend urgent tissue diagnosis when these are present (Li *et al.*, 2022; Yao *et al.*, 2023).

Limitations of this report include its single-case design and limited generalizability. Clinical outcomes may vary with tumour site, resectability, margin status, and adjuvant therapies. Further studies and pooled analyses are needed to better define optimal systemic treatments for MTT.

4. Conclusion

Malignant Triton Tumour (MTT) is a rare, aggressive rhabdomyoblastic variant of MPNST that commonly presents as a large, lobulated soft-tissue mass (>5 cm) with internal heterogeneity from necrosis, haemorrhage, or cystic change. MRI typically shows low–intermediate T1 and high T2 signal with heterogeneous postcontrast enhancement, while CT may demonstrate peripheral enhancement, septations, and occasional calcifications. Imaging features overlap with those of other high-grade sarcomas, so a definitive diagnosis requires histopathology and immunohistochemistry. Given the tumour's high risk of local recurrence and distant metastasis, and its limited responsiveness to systemic therapy, early recognition, prompt biopsy, and radical, wide surgical resection with multidisciplinary oncologic management are essential to optimise outcomes. Radiologists should report aggressive features (large size, lobulated/irregular margins, infiltration, heterogeneity, and calcifications) and recommend urgent tissue confirmation when present.

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Conflict of interest

We declare no conflict of interest in this article.

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