Malignant Triton Tumor: Role of Electron Microscopy in Determining Differentiation

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Abstract

Malignant triton tumors (MTTs) are peripheral nerve sheath tumors: Perivascular origin is extremely rare. We report a sporadic perivascular MTT in a 37-year-old man. He presented with a swelling in the right popliteal fossa. Imaging revealed a solid lesion adherent to the popliteal artery. He underwent en bloc excision of the tumor with repair of the popliteal artery. We studied the histopathology and electron microscopy (EM) features of this tumor. He received local radiotherapy and remains disease-free at 2 years. EM may help prognosticate tumor behavior when these tumors occur in rare/nonneural locations.

Key Words: Differentiation, electron microscopy, perivascular, triton tumor

Introduction

Malignant triton tumors (MTTs) are peripheral nerve sheath tumors: Perivascular origin is extremely rare. We describe the management of such a case.

Case Report

Clinical presentation and operative details

A 37-year-old male presented with a single, nontender, gradually progressive swelling in the right popliteal fossa for 3 months. He did not complain of paresthesia, weakness, limb swelling, or claudication. There was no history of trauma, allergies, drug abuse, or other systemic diseases. On examination, there was an 8 cm × 6 cm firm nonpulsatile swelling in the right popliteal fossa. There was no inguinal lymphadenopathy, and the neurovascular examination was normal.

Magnetic resonance imaging (MRI) revealed a uniformly hyperintense mass adherent to the right popliteal artery. The differentials considered were a soft tissue tumor or a thrombosed pseudoaneurysm - the former was confirmed on duplex ultrasound. He underwent excision of the tumor. A posterior approach was used for control of the popliteal artery - the tumor was densely adherent to the artery. It was completely separate from the tibial nerve which coursed more laterally [Figure 1a]. The tumor was excised en bloc with the artery. This was reconstructed with an interposition vein graft [Figure 1b]. The patient’s postoperative recovery was uneventful. He did not develop any neurological deficit, and his distal pulses were normal. He received local radiation therapy postprocedure. He had a good clinical outcome with no tumor recurrence or metastasis at 2-year follow-up. He consented for the publication of this report.

Histopathology and electron microscopy

The tumor did not have intraluminal extension [Figure 1c]. Histopathology of the tumor revealed fascicles of neoplastic spindle cells in a myxoid stroma with rhabdomyoblastic differentiation [Figure 2]. Lipoblasts were not seen. Mitotic activity was 6/high-power fields. Immunohistochemistry showed diffuse positivity of the tumor cells for S100. There was focal positivity for CD34 and bcl2. The rhabdomyoblasts were positive for desmin and myogenin. The absence of SYT-SSX1 and SYT-SSX2 translocation by quantitative reverse transcriptase polymerase chain reaction ruled out synovial sarcoma. A diagnosis of malignant peripheral nerve sheath tumor (MPNST) with rhabdomyoblastic differentiation (triton tumor) was made. However, the origin of this rare tumor was not clear. Hence, we proceeded for transmission electron microscopic examination, Tecnai T12 Spirit, FEI which showed predominantly tumor cells with long interdigitating cytoplasmic processes [Figure 3], some with...
Malignant triton tumors (MTTs) were first described by Masson and named by Woodruff and Locatelli. Clinical presentation is similar to other soft tissue tumors. They occur in all parts of the body, but the involvement of the extremities is less commonly reported than the head and neck or retroperitoneum. Preoperative diagnosis of the tumor is by multimodality imaging (computed tomography/MRI/ultrasound). Imaging assesses the local extent and helps plan resection but does not allow differentiation from other MPNSTs. Treatment is radical excision and postoperative radiotherapy. Recurrence rates are high with a dismal 5-year survival rate (about 11%), which is worse when compared to other MPNSTs. Large size, site (retroperitoneum, buttock), histological subtype, and distant metastases at diagnosis are poor prognostic factors.

MTTs are diagnosed in 10% of MPNSTs. They arise from Schwann cells or pluripotent neural crest cells; 25%–50% are associated with neurofibromatosis (NF). Sporadic tumors in non-NF patients can occur. Nerve origin may not be identified in 44%–61% of cases even when the patient is a diagnosed case of NF. Histopathological diagnosis is by Daimaru criteria – the presence of Schwann cells with rhabdomyoblastic differentiation in the tumor. The exact pathogenesis as to why the rhabdomyoblastic differentiation worsens prognosis is not understood. The behavior and features of those arising from atypical (nonneural sites) are thought to be less aggressive, but due to the rarity of these tumors, literature is sparse. Demonstration of perineural differentiation is associated with an overall better survival. This may be seen even in sporadic tumors atypical/perivascular sites, which have no connection to a peripheral nerve. Electron microscopy (EM) has been used for confirming the diagnosis of MTT in tumors arising from nerves and in differentiating benign from malignant tumors.

We performed EM to study tumor origin and differentiation in this patient with sporadic malignant MTT. EM helped establish the tissue of origin of this MTT to definitively differentiate it from other perivascular tumors. EM features were of a well-differentiated tumor with luse bodies which was suggestive of perineural differentiation. This was proved in this patient only by EM as the intraoperative, and histopathology had not been able to demonstrate relation to a peripheral nerve. The good clinical outcome in a malignant tumor with such a differentiation has to be studied in larger cohorts to determine if EM should be used in all sporadic MTTs as a prognostic factor.

**Conclusion**

The use of EM to determine origin and differentiation in sporadic MTT as seen may help in prognosticating...
response to therapy, especially in rare instances of origin in nonneural locations.

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**Conflicts of interest**

There are no conflicts of interest.

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