Intra Peritoneal Malignant Triton Tumor: A Case Report and Literature Review

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ABSTRACT

Background: Malignant triton tumor (MTT) is a variant of the typical malignant peripheral nerve sheath tumors with rhabdomyoblastic differentiation what mostly occur in the context of neurofibromatosis type 1. Case presentation: Here, we report on a case of a 15 year-old male with a clinical history of neurofibromatosis type 1 who presented to the pediatric emergency department with an intra peritoneal tumor of about 20 cm of diameter. The biopsy of this tumor confirmed histology of rhabdomyosarcoma. The patient received six cycles of chemotherapy. After that, resection of the tumor was performed. Histological findings revealed a malignant peripheral nerve sheath tumors (MPNST) of Triton type. Then he received 28 sessions of radiation therapy. The child died 11 months after radiation therapy. Conclusion: Malignant triton tumor is a very aggressive tumor. Multidisciplinary approach is required to diagnosis and treatment of this rare pediatric tumor.

Keywords: Triton tumor, Intra-peritoneal, neurofibromatosis

1. INTRODUCTION

Malignant triton tumor (MTT) is a very rare and highly invasive variant of the typical malignant peripheral nerve sheath tumors that occur in the context of neurofibromatosis type 1 (Von Recklinghausen’s disease NF1) in more than 50% of cases [1]. The treatment of this entity involved radical surgical resection, adjuvant chemotherapy and radiation therapy. We report on a rare case of intra peritoneal MTT in a patient with NF1.

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2. **CASE REPORT**

A 15 year-old male presented to the pediatric emergency department with abdominal pain, vomiting and expansion of his abdomen. Clinical examination revealed a mass of ~ 15 cm of diameter in the left hypochondrium and signs of neurofibromatosis (diffused café-au-lait spots on the thorax and abdomen and dorsolumbar scoliosis with thoracic deformity) (Fig.1).

*Fig.1. Clinical photograph of our patient*

(A) Diffused café-au-lait spots on the thorax and abdomen.

(B) Dorsolumbar scoliosis

Abdominal CT scan showed an intra peritoneal mass located between the small gastric curvature and the inner border of the liver of about 20.5×19×15 cm (Fig.2).

*Fig.2. Tomography (CT) scan result of our case of malignant triton tumor (MTT) revealing an enormous intra peritoneal tumor of about 20X19X15 mm*

(A) Coronal CT image

(B) Axial CT image
Microscopic examination of this tumor after surgical biopsy revealed a Rhabdomyosarcoma (with Desmin(+), Myogenin(+), Vimentin(+), ProteinS 100(+), NSE(-), CD117(-) and a Ki67-proliferative index of 70% on the immunohistochemistry results) (Fig.3).

Fig.3. Initial biopsy

(A) GX10 proliferation of fusiform cells on a mucus background

(B) GX40 Cells with reduced cytoplasm and atypical nuclei (characteristics of rhabdomyoblastic cells)

(C) IHC some cells myogenin positive

A bone scintigraphy, Thoracic CT and myelogram didn't find other lesions. The patient received six courses of chemotherapy, 3 courses of combined Ifosfamide (2500mg), vincristine (1.2mg) and Actinomicyne D (120mg), 2 courses combined Carboplatine (450mg), Doxorubicine (25mg) and vincristine (1.2mg) and one cycle of Ifosfamide (2500mg), vincristine (1.2 mg ) and Etoposide (120mg). The tumor volume regressed by 70%. After that, the patient was posted for exploratory laparotomy, the resection R1 of the tumor was performed. The specimen was sent for histopathological examination which gave a diagnosis of malignant peripheral nerve sheath tumor (MPNST) of Triton type (Immunohistochemically, the tumor cells homogenously expressed ProteinS100 and Myogenin without evidence of expression of Aktin and Desmin) (Fig.4).

Fig.4. Tumor

(A) GX10 Proliferation of fusiform cells

(B) GX40 Atypical cells
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Patient received 3 other courses of chemotherapy (CEV-IVE-CEV) followed by radiation therapy for the total abdomen including the tumor residue, at a total dose of 50.4 Gy (5 X 1.8Gy / week). The child died 11 months after radiation therapy (local progression located on the liver).

3. DISCUSSION

Malignant triton tumor (MTT) is a variant of malignant peripheral nerve sheath tumor (5–10% of MPNSTs)[2]. In 1973, Woodruff introduced the term MTT when he reported a series of ten cases of malignant schwannoma with rhabdomyoblastic differentiation [3]. MTT are associated to the NF-1 in 50-70% of the cases [4], occurring mainly in young males with a median age of 30 years [5, 6]. The most commons anatomical sites for this tumor are head and neck and extremities [7]. These two locations have a better prognosis than other sites such as the buttocks, trunk, and retroperitoneum [6].

The immunohistochimical profil of the MTT tumor find S-100 protein positive and Leu-7 (CD57) positive (confirming Nerve sheath differentiation) and desmin, actin, and myogenin positives (confirming rhabdomyoblastic differentiation) [3].

The studies suggest that the standard of treatment for this tumor is a radical excision followed by postoperative radiotherapy and chemotherapy. Radiotherapy is recommended for the MTT with positives margins after surgical resection (according to the standard protocol of soft tissue sarcoma)[10]. The role of adjuvant chemotherapy has not been clearly defined [11].

MTT is a very aggressive tumor with a poor prognosis (The Five-year survival rate for MTT is 10-20% [4]). We report here a rare case of an intra peritoneal MTT in the context of NF1 (Fig.1). Because of surgical resection R1, we followed the standard of treatment of soft tissues sarcomas and we decide to give radiations (high dose 50,4Gy), and 3 others courses of adjuvant chemotherapy. The patient dies from local progression 11 months later despite an aggressive treatment. The further studies are needed to make guidelines for treatment of this rare but highly and rapidly invasive tumor.

COMPETING INTERESTS
The authors have declared that no competing interest exists.

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ETHICAL APPROVAL
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CONSENT

REFERENCES