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A Rare Pediatric Case of Abdominal Malignant Triton Tumor

Rachida Saouab[1] and Rachida Dafiri[2]

[1]Departement of Radiology, Military Hospital HMIMV, Cadi Ayyad University, Faculty of Medicine and Pharmacy, Marrakech.

[2]Departement of Radiology, Pediatric Hospital, Mohammed V University, Faculty of Medicine and Pharmacy, Rabat.

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ABSTRACT

Malignant triton tumor (MTT) is an extremely rare variant of malignant peripheral nerve sheath tumor (MPNST) with rhabdomyo-sarcomatous differentiation. We report a case of a 14-year-old male without history of neurofibromatosis presented for post-traumatic abdominal pain. Ultrasonography and contrast-enhanced computed tomography (CT) revealed a large heterogeneous intraperitoneal mass near the vascular structures and repressing the adjacent organs. Surgery and anatomopathological examination revealed a malignant triton tumor. MTTs are highly aggressive tumors that are fast-growing with a tendency to recur locally and metastasize early. To date, there is no treatment consensus available yet and the overall prognosis remains dismal.

Introduction

Malignant triton tumor (MTT) is an extremely rare type of sarcoma that arises from peripheral nerve sheaths. It accounts for about 5 % of Malignant peripheral nerve sheath tumor (MPNST) [1, 2]. This tumor is thought to originate from Schwann cells of peripheral nerves or within existing neurofibromas and is characterized by the coexistence of malignant rhabdomyoblasts and malignant Schwann cells [3]. This is an interesting tumor not encountered very much in the neuro-oncology literature. It manifests itself more often in individuals with neurofibromatosis type I (NF-1) disease but also sporadically or post radiotherapy. We report a very rare case of intra-abdominal MTT in a child.

Case Report

A 14-year-old male patient, born in a family of healthy children with a history of scoliosis and psychomotor retardation without personal history of NF1, was referred to pediatric emergency following abdominal trauma, with epigastric pain and an abdominal mass. First abdominal ultrasonography and contrast-enhanced computed tomography (CT) of the chest and abdomen revealed a huge heterogeneous intraperitoneal mass that not involved adjacent organs and vessels measuring approximately 17 cm in diameter (Figure 1). Following contrast, heterogeneous enhancement of the mass was noted. Adjacent vessels, such as the common hepatic artery, the portal vein and the inferior vena cava, were compressed. There was no evidence of associated lymphadenopathy or distant metastases.

Biopsy and incomplete resection was undertaken. Histopathological examination revealed an MTT characterized by an MPNST with abundant rhabdomyosar comatous differentiation. Immunohisto chemical staining was negative for PS100, NSE and CD117 and the rhabdomyoblasts were positive for vimentine and desmine. The Ki67-proliferative index was 70–80% (Figure 2). Following surgery, the patient made a good recovery and received palliative intravenous chemotherapy (IVA1). The patient had no local recurrence and no metastasis and still alive after 21 months.

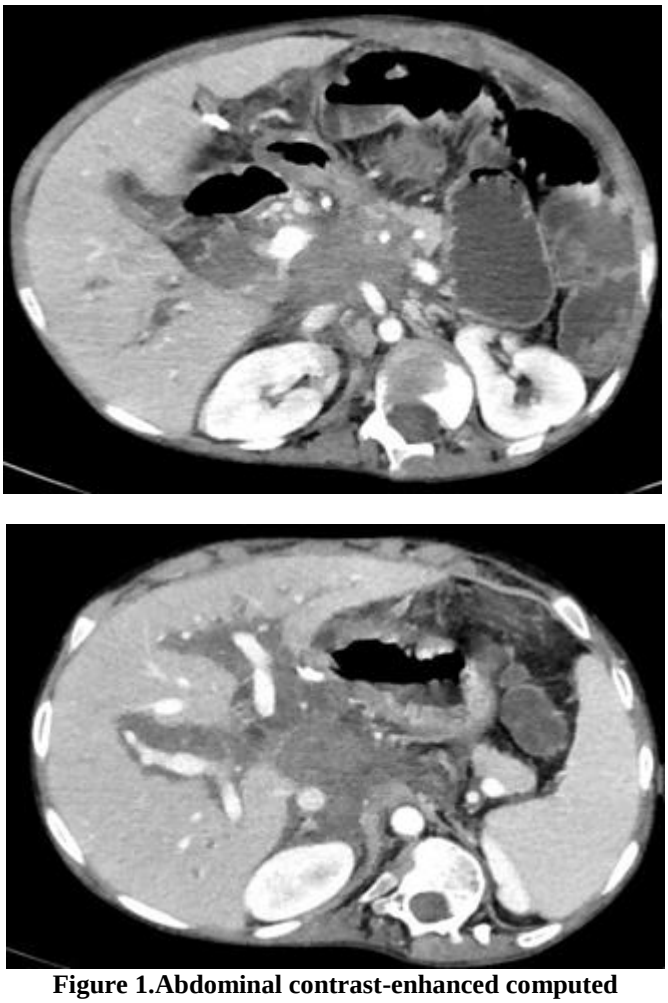

Figure 1.Abdominal contrast-enhanced computed tomography (CT) revealed a huge coeliac mass that not involved adjacent organs and vessels.

Tele:

E-mail address: rachida_sa@yahoo.fr

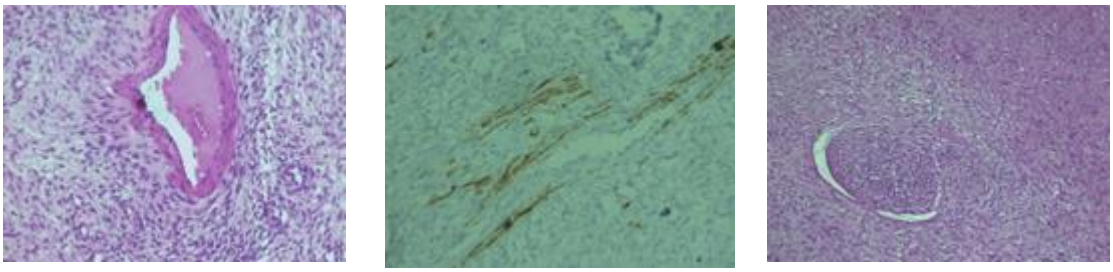

Figure 2. Histopathological examination showing the cellular proliferation of spindle-shaped cells with small slit-like vessels and chronic inflammatory infiltration. Immunohistochemical staining was negative for PS100, NSE and CD117 and the rhabdomyoblasts were positive for vimentine and desmine. The Ki67-proliferative index was 70–80%.

Discussion

The first description of MTT dates back to 1932, when Masson described rhabdomyosarcomatous elements within malignant peripheral nerve sheath tumors (MPNST) in patients with neurofibromatosis. To date, approximately 170 cases of MTT have been described in the literature of which more than 50% had previously been diagnosed with neurofibromatosis 1 (von Recklinghausen'sdisease, NF1) [4]. MTT is commonly seen in the head, neck, extremities and trunk [1,5]. Other rare locations are the buttock, visceras, retroperitoneum and mediastinum. Clinical findings are non-specific and depend largely upon the location; MTTs are often aggressive invading or repressing adjacent anatomic structures. The prognosis is dismal with a five-year survival rate of 12–26% [6, 7].

MTT commonly presents heterogeneous CT attenuation and MR signal intensity due to necrosis, hemorrhage or cystic change. The contrast enhancement pattern of MTTs is also frequently heterogeneous [8]. Larger lesion, irregular borders, infiltration of adjacent structures and rapid growth on interval imaging are radiologic criteria differencing MTT from benign nerve sheath tumors [9, 10][16,17]. Several studies demonstrated that FDG-PET was a sensitive and specific tool for diagnosis. The suggested cut- off values of SUVmax for the differentiation between malignant and benign peripheral nerve sheath tumor are 2.5 - 6.1, which is a relatively wide-range with some degree of overlap [11]. The high FDG uptake seems to correlate with the proliferation of spindle-shaped cells and less cellular myxoid stroma [8].

Woodruff *et al*. defined MTT with the following histo-pathological criteria: 1) the neoplasm is connected to peripheral nerves or occurs in patients with NF1; 2) most of the neoplasm consists of Schwann cells; and 3) the neoplasm contains rhabdomyoblasts [4].

Complete surgical resection with a wide margin and adjuvant radiotherapy are generally accepted in order to obtain the best outcome for an MTT. Systemic chemo-therapy is selected if there is evidence of metastatic disease [12, 13].

Conclusion

MTTs are rare, aggressive tumors with mixed nerve and muscle components, that tend to recur locally and metastasize, requiring a multimodal treatment. Imaging is Radical surgical excision is the only therapeutic intervention with curative intent at the moment.

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