A rare case of retroperitoneal malignant Triton tumor invading renal vein and small intestine

Redak slučaj retroperitonealnog malignog tumora Triton sa invazijom renalne vene i tankog creva

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Abstract

Introduction. Malignant Triton tumor is a very rare malignant peripheral nerve sheath tumor with rhabdomyosarcomatous differentiation. Most of those tumors occur in patients with von Recklinghausen’s disease or as a late complication of irradiation and commonly seen in the head, neck, extremities and trunk. Case report. We reported retroperitoneal malignant Triton tumor in a 57-year-old female patient. Skin lesions were not present, and there was no family history of neurofibromatosis or previous irradiation. The presented case is one of a few recorded in the specialized literature that occurs in the retroperitoneal space in sporadic form. In this case, tumor consisted of a multilobular mass was in close relation with the abdominal aorta and inferior vena cava and involved the renal vein with gross invasion of the small intestine. The patient underwent total resection of the tumor and left nephrectomy. The small intestine 10 cm in length was also resected and end-to-end anastomosis was conducted. The postoperative course was uneventful and the patient was discharged from the hospital ten days after the surgery.

Conclusion. Diagnostically, it is crucial to recognize this uncommon histological variant because malignant Triton tumor has a worse prognosis than classic malignant peripheral nerve sheath tumor does. The use of immunohistochemistry is essential in making the correct diagnosis. Only appropriate pathological evaluation supported by immunostaining with S-100 protein and desmin confirmed the diagnosis. Aggressive surgical management treatment improves the prognosis of such cases with adjuvant radiotherapy.

Key words: peripheral nervous system neoplasms; diagnosis; immunohistochemistry; surgical procedures, operative; treatment outcome.

Apstrakt


Ključne reči: živci, periferni, neoplazme; dijagnoza; immunohistohemija; hirurgija, operativne procedure; lečenje, ishod.

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Introduction

Malignant Triton tumor (MTT) is a malignant peripheral nerve sheath tumor (MPNST) with rhabdomyosarcomatous differentiation. MTT constitutes about 5% of all MPNSTs. MTT arises in two principal forms: sporadic or in association with neurofibromatosis type 1 [von Recklinghausen’s disease (NF-1)]. Slightly more than half of the cases of MTT have been reported in conjunction with NF-1. MTT is commonly seen in the head, neck, extremities and trunk 1. The fact that the presence of this unusual tumor in the retroperitoneal space is extremely rare has prompted the authors to report this case. We presented a 57-year-old female patient in whom a retroperitoneal paravertebral mass was postoperatively diagnosed as MTT and described the histomorphological and immunohistochemical features of this uncommon tumor. In this case, tumor developed outside the setting of NF-1.

Case report

A 57-year-old female patient presented with a 2-month history of abdominal pain radiating to the back. Physical examination revealed a painful abdomen in the region of umbilicus. Full blood cell count, serum urea levels, and electrolyte levels were within normal limits. Chest X-ray was normal. At laparotomy, a multilobular paravertebral mass the size of two male fists was found occupying the left abdominal region. Tumor was in close relation with the abdominal aorta and inferior vena cava and appeared to involve the left renal vessels (artery and vein) with gross invasion of the small intestine. The patient underwent total resection of the tumor and left nephrectomy was performed. Upon resection, thrombosis was present on part of a renal artery and there was no need for further pathological analysis. The small intestine 10 cm in length was also resected and end-to-end anastomosis was conducted. The postoperative course was uneventful and the patient was discharged from the hospital ten days after the surgery.

On the basis of histological findings and immunohistochemistry, a malignant Triton tumor, an uncommon subtype of peripheral nerve sheath tumor with rhabdomyosarcomatous elements, was diagnosed. Thereafter, the patient was referred to the Oncology Department for Radiotherapy.

During the 8-month follow-up, the patient showed no evidence of recurrence or metastasis.

Macroscopically, the tumor was received in three fragments measuring 7, 5 and 4.5 cm. The cut sections showed solid, firm and yellow whitish tissue with areas of necrosis (Figure 1). The left kidney measured 11 x 6 x 5 cm. Cut section showed no gross abnormalities. Samples of left renal vein measured 0.6 and 0.7 cm, and part of small intestine was 10 cm in length.

Microscopically, the tumor was composed of spindle cells arranged in a fasciculated pattern and whorls (Figure 2). Spindle cells showed wavy, hyperchromatic nuclei with indistinct light staining cytoplasm. Hypercellular and hypocellular zones with areas of palisading necrosis and myxoid stroma were also present. Scattered large round cells with eosinophilic cytoplasm and hyperchromatic nuclei were seen admixed with neoplastic spindle cells.

On histology, the kidney showed no significant morphological abnormalities. The lesion involved renal vein and a part of the small intestine were similar, displaying poorly differentiated spindle cells.

Immunohistochemically, the spindle cells were focally positive for S-100 (Figure 3), strongly positive for vimentin, and negative for cytokeratin 7, EMA, HMB-45, and CK AE1/AE3. The large, round eosinophilic pleomorphic cells

Fig. 1 – A fragment of primary malignant Triton tumor. The cut surface was solid, firm and yellow whitish with the areas of necrosis

Fig. 2 – Malignant Triton tumor (spindle cells arranged in interlacing fascicles, sheaths and whorls; H&E, ×100)

Fig. 3 – Focal staining of spindle cells for S-100 protein (peroxidase-antiperoxidase technique, ×200)
were immunoreactive for muscle-specific actin and desmin (Figure 4). Immunohistochemical staining with S-100 and desmin indicated that the tumor cells originated from Schwann cells and showed rhabdomyosarcomatous differentiation. Ki-67 was expressed in a large number of cells (30%).

Fig. 4 – Staining for desmin was strongly positive in the area of malignant Triton tumor (peroxidase-antiperoxidase technique, ×100)

Based on these findings, the histopathological diagnosis was malignant peripheral nerve sheath tumor with rhabdomyosarcomatous differentiation, namely malignant Triton tumor.

Discussion

MPNSTs are neoplasms derived from the cellular constituents of the peripheral nerve sheath. This term replaces the earlier terms malignant schwannoma, neurofibrosarcoma, and neurogenic sarcoma. The majority arise from Schwann cells, but some could develop from fibroblasts and supporting cells known as perineural cells. The capacity of MPNSTs to undergo focal divergent differentiation to rhabdomyosarcoma, chondrosarcoma, osteosarcoma, angiosarcoma, epithelial elements, or a combination thereof is well known. MTT is a variety of this type of tumors which presents a rhabdomyoblastic differentiation. This composite neoplasm was first described in 1938 by Masson and Martin, who suggested that the neural elements in the tumor induced differentiation of skeletal muscle in much the same fashion as normal nerve was believed to induce the regeneration of skeletal muscle in the Triton salamander. This tailed amphibian displays the ability to regenerate limbs after the cut end of the sciatic nerve is implanted into the soft tissue of its back. Although Masson believed that one cell line induced the other, it seems more likely that both cell lines originate from less well-differentiated neural crest cells. The term “malignant Triton tumor” was first introduced by Woodruff et al. in 1973.

MTTs are extremely rare, with less than 100 cases documented world-wide to date. Regarding location, MTT occurs predominantly in the head, neck, trunk regions and lower extremities. To the best of our knowledge, there are only a few reports of these tumors developing in the retroperitoneal space. MTT shows marked male predominance with more predilections for younger age groups. The sporadic forms mostly occurring in females of older age groups, as the case reported herein. These tumors may also arise in sites of previous radiation therapy. We reported the case of paraaortic MPNST occurring in the patient without clinical setting of NF-1 or previous irradiation.

MPNST is one of the most histologically variable soft tissue tumors, and use of immunohistochemistry is essential in making a correct diagnosis. The fasciculated, spindled cell growth pattern may cause confusion with leiomyosarcoma, fibrosarcoma, or monophasic synovial sarcoma. In addition, MPNST must be distinguished from melanoma malignum. In the present case, histology and immunohistochemical staining revealed a typical pattern of MPNST with the additional features of rhabdomyoblastic differentiation supported by positive staining with desmin and musclespecific actin.

Retroperitoneal localization MTT has the most unfavorable prognosis due to the delayed diagnosis but also due to the relation to adjacent organs. This case of MTT was presented as a large abdominal mass with invasion of the left renal vein and the small intestine since these retroperitoneal tumors are often asymptomatic in the earlier stages. The natural history of MTT is much more aggressive than MPNST. The tumor has a high propensity for early local recurrence, rather than metastatic disease. The prognosis is poor with a 5-year survival rate around 12%. Location has been correlated with survival. Tumors occurring in the head and neck, upper and lower extremities have a better prognosis than tumors located in the retroperitoneum, buttocks or trunk. Cytogenetic studies have revealed a break-point in 11p15, considered a region of myogenic differentiation. Amplification of c-myc oncogene is probably responsible for aggressive biologic behavior of MTT. As MTT is a very aggressive tumor, behaving like a high grade sarcoma, it is believed that to obtain the best outcome a full surgical resection with as wide a margin as possible is vital followed by adjuvant radiotherapy.

Conclusion

Retroperitoneal malignant Triton tumor is extremely rare, but an important pathological condition. This uncommon histological variant has the worse prognosis than classic malignant peripheral nerve sheath tumor does. Immunohistochemistry is an essential tool for ruling out differential diagnostic considerations. Radical surgical excision of the tumor followed by radiation therapy is the treatment of choice.

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