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Malignant triton tumor in the abdominal wall: A case report

Ho Jik Yang, Donghyun Kim, Won Suk Lee, Sang-Ha Oh

Corresponding author: Sang-Ha Oh, MD, PhD, Professor, Surgeon, Department of Plastic and Reconstructive Surgery, Chungnam National University College of Medicine, 282, Munwha-ro, Jung-gu, Daejeon 35015, South Korea. djplastic@cnu.ac.kr

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Abstract

BACKGROUND

Malignant triton tumor (MTT) is a subgroup of malignant peripheral nerve sheath tumors (MPNSTs) that exhibits rhabdomyosarcomatous differentiation and follows an aggressive course. MTTs are primarily located along peripheral nerves. Cases of MTTs in the abdominal wall have not been reported. MTT has a poorer prognosis than classic MPNSTs, and accurate diagnosis necessitates a keen understanding of the clinical history and knowledge of its differential diagnosis intricacies. Treatment for MTTs mirrors that for MPNSTs and is predominantly surgical.

CASE SUMMARY

A 49-year-old woman presented with a subcutaneous mass in her lower abdominal wall and a pre-existing surgical scar that had grown slowly over 3-4 months before the consultation. She had previously undergone radical hysterectomy and concurrent chemo-radiotherapy for cervical cancer approximately 5 years prior to the consultation. Abdominal computed tomography (CT) showed a 1.3 cm midline mass in the lower abdomen with infiltration into the rectus abdominis muscle. There was no sign of metastasis (T1N0M0). An incisional biopsy identified sporadic MTT of the lower abdomen. A comprehensive surgical excision with a 3 cm margin, inclusive of the peritoneum, was executed. Subsequently, the general surgeon utilized an approach akin to the open peritoneal onlay mesh technique. The skin defect underwent additional treatment with an excision shaped as a mini-abdominoplasty. No complications arose, and annual follow-up CTs showed no signs of recurrence or metastasis.

CONCLUSION

An abdominal MTT was efficaciously treated with extensive excision and abdominal wall reconstruction, eliminating the need for postoperative radiotherapy.

Key Words: Malignant triton tumor; Abdominal wall; Surgical excision; Reconstruction; Case report

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Core Tip: Malignant triton tumor is an uncommon condition characterized by a poor prognosis. Cases emerging in the abdominal wall are especially rare. Swift differential diagnosis and comprehensive surgical removal play a pivotal role in management. In this instance, we managed to treat the patient without the necessity for postoperative radiotherapy, thanks to a wide excision complemented by suitable reconstruction.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) constitute approximately 5%-10% of soft tissue sarcomas and are believed to originate from Schwann cells or adjacent cells with perineural differentiation[1]. Malignant triton tumor (MTT), a subtype of MPNST displaying rhabdomyoblastic differentiation, presents in two primary forms: sporadic, and neurofibromatosis type 1 (NF-1)-associated forms. MTT represents approximately 5% of all MPNSTs, and its nomenclature references early experiments with the amphibian Triton, which can regenerate limbs containing muscle following ectopic implantation of the transected end of the sciatic nerve[2].

MTTs are predominantly located along peripheral nerves, frequently near the spine, in the head and neck region, or within the upper and lower extremities. Other uncommon sites include the buttocks, viscera, retroperitoneum, mediastinum, and intracranial locations such as the parieto-occipital lobe, lateral ventricle, and cerebellopontine angle[2-6]. The extreme rarity of this tumor in the abdominal wall motivated this case report.

The presentation, progression, and response to treatment of MTTs remain largely undefined as published data predominantly consist of case reports and limited patient

series. MTT displays aggressive biological behavior, with various factors like location, size, and tumor stage potentially impacting patient prognosis and survival. Non-extremity MTTs often result in a poor prognosis. The metastasis rate for MTT stands at 31.4% and correlates with reduced survival in its sporadic form[7]. Adjuvant radiotherapy plays a crucial role in treatment, with complete surgical excision of MTT providing a survival advantage[8]. However, no studies have explored the relationship between the quality of surgical margins and treatment outcomes.

To shed light on the progression and prognosis of this infrequent neoplasm in the abdominal wall, we detail a case of sporadic MTT originating in the abdominal wall with a distinct prognosis.

CASE PRESENTATION

Chief complaints

A 49-year-old woman presented with a subcutaneous mass in her lower abdominal wall alongside a previous surgical scar.

History of present illness

A subcutaneous mass had enlarged over the 3-4 months prior to consultation.

History of past illness

Approximately five years before this consultation, she had undergone a radical hysterectomy and concurrent chemo-radiotherapy (135 Gy) for cervical cancer.

Personal and family history

The patient did not display café-au-lait spots or cutaneous neurofibromas. Both her medical and family histories related to NF-1 were unremarkable.

Physical examination

A firm, painless subcutaneous mass measuring approximately 1 cm was identified. No inguinal lymphadenopathy was detected upon palpation.

Laboratory examinations

There were no specific findings.

Imaging examinations

Abdominal computed tomography (CT) showed a 1.3 cm midline mass in the lower abdomen with infiltration into the rectus abdominis muscle. Comprehensive CT scans (encompassing the chest, abdomen, and pelvis), combined with positron emission tomography/CT and colonoscopy, found no signs of metastasis (T1N0M0) (Figure 1).

FINAL DIAGNOSIS

Combined with the patient's medical history, the final diagnosis was MTT in the lower abdomen.

TREATMENT

Under general anesthesia, a wide surgical excision of the mass, ensuring a 3 cm margin that included the peritoneum, was carried out. The resected margins were found to be clear. Following this, the general surgeon employed a technique akin to the open peritoneal onlay mesh method to prevent herniation of abdominal wall defects. For reinforcement, a composite mesh (Parietene composite mesh; COVIDIEN, Dublin, Ireland) was introduced and anchored to both pelvic symphyses. The peritoneum was subsequently advanced and repaired on both sides. The anterior fascia's deficit was addressed using an acellular dermal matrix (SureDerm; Hans BioMed, Seoul, South Korea) with further reinforcement and rectification. To achieve a minimal scarring effect, the skin defect underwent an additional excision in the pattern of a mini-abdominoplasty and was sutured in layers (Figure 4).

OUTCOME AND FOLLOW-UP

The post-operative period was uneventful, and annual follow-up CT scans showed no recurrence or metastasis. Three years post-diagnosis, the patient remains alive and without any sign of recurrence (Figure 5).

DISCUSSION

MTT presents in two principal forms: sporadic, and associated with NF-1. In its sporadic form, other spindle cell sarcomas, such as fibrosarcoma, malignant fibrous histiocytoma, and rhabdomyosarcoma, might be considered in the differential diagnosis[2]. The most widely accepted view suggests that MTT originates from Schwann or ectodermal cells of the neural crest. More than 50% of patients with MTT develop NF-1, with the remainder being disseminated[5]. MTT associated with NF-1 is notably more prevalent in men, particularly in younger age groups (28-36 years old). The sporadic form tends to appear in older women (40-44 years old). Divergent differentiation strongly suggests NF-1, and 57% of patients with MTT, which manifests after a prolonged latent period of 10-20 years, have NF-1[9,10]. As many as 6% of malignant or atypical peripheral nerve tumors, inclusive of MTT, have associations with prior radiotherapy[11], a crucial factor highlighted in our case report. In this instance, the tumor emerged sporadically in a 49-year-old woman and was linked to prior radiotherapy.

The diagnosis of MTT chiefly rests on the histopathological and immune-histochemical attributes of the tumor. There is consensus that an MPNST diagnosis can be rooted in morphological features, bolstered by S-100 protein positivity. Morphological characteristics include alternating hypocellular and hypercellular regions, the presence of thin wavy comma-shaped or bullet-shaped nuclei in hypocellular areas, nuclear palisading, nerve whorls or tactoid bodies resembling Wagner-Meissner corpuscles, prominent thick-walled vasculature, and heterologous elements like rhabdomyoblasts, cartilage, and bone. Such tumors exhibit focal positivity

for the S-100 protein in 50%-90% of cases, pointing to a nerve sheath derivation. Rhabdomyoblasts test positive for desmin, myogenin, and myo-D1[2].

MTTs typically have an unfavorable prognosis, contingent on the tumor's location, grade, and the completeness of surgical margins. Cases located in the head, neck, and extremities are less intricate than those found in other sites, like the buttocks. Past research indicates that MTT in conjunction with NF-1 has a worse prognosis relative to its sporadic counterparts[9,12]. There was no discernible difference between MTT and MPNST groups regarding rates of local recurrence or metastasis. Nonetheless, the 5-year survival rate for MTTs stands at a mere 5%-15%, compared to 50%-60% for MPNSTs. Several factors underlie these disparate survival rates. MTT proliferates swiftly and, in its nascent stages, is susceptible to local recurrence and hematogenous metastases. In comparison to MPNST, MTT often arises in more advanced-aged patients, predominantly manifests in the trunk, and results in larger tumors. Furthermore, patients diagnosed with MTT experience a reduced metastasis-free interval and diminished overall survival. Thus, MTT's aggressive nature sets it apart from MPNST, underscoring the significance of differential diagnosis[13]. Although the association of genomic alteration and prognosis of MTT has not been fully investigated, there have been a few studies that evaluated such a relationship, and associated poorer survival with tumors having gains and loss of particular genes, suggesting promise for understanding this disease[14-16].

The treatment approach for MTT mirrors that of MPNST and is predominantly surgical. The Oncology Consensus Group endorses postoperative radiotherapy as a standard treatment protocol for MTT. Typically, radical excision followed by high-dose radiotherapy is employed, and recent studies hint at the potential of neoadjuvant therapy and adjuvant chemotherapy in eliminating micro-metastases. However, this treatment remains controversial. A MTT case of gluteal underwent full resection with postoperative chemoradiotherapy and there were no recurrence symptoms for 4 years after treatment[17]. Whereas another case adopted the same treatment for a patient with rectum MTT, who passed away under palliative therapy and survived only

9 months[4]. Therefore, it is necessary to seek effective methods for MTT treatment[6]. In the discussed case, the MTT developed post-radiotherapy, a modality that can induce significant side effects when aimed at the abdomen[18]. Given the comprehensive nature of the surgical excision of the abdominal wall tumor, postoperative radiotherapy was deemed unnecessary. While no recurrence or metastasis has been observed, consistent follow-up is imperative, especially since the post-surgery window is under 5 years.

MTT is a rare condition with an unfavorable prognosis. Prompt differential diagnosis and thorough surgical removal are crucial. However, in the presented case, extensive surgical removal was deemed sufficient without the need for postoperative radiotherapy.

CONCLUSION

MTT is an uncommon condition characterized by a poor prognosis. Cases emerging in the abdominal wall are especially rare. Swift differential diagnosis and comprehensive surgical removal play a pivotal role in management. In this instance, we managed to treat the patient without the necessity for postoperative radiotherapy, thanks to a wide excision complemented by suitable reconstruction.

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