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A series of 10 malignant triton tumors in one institution

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Abstract

Malignant triton tumor (MTT) is an extraordinarily uncommon and aggressive tumor which have poor prognosis. Malignant peripheral nerve sheath tumors with additional rhabdomyoblasts are found in MTT histologically. The prognosis of patients is poor. The goal of our study is to describe the largest number of cases characteristic and outcome, to our knowledge, such a presentation was not described in the English-language literature until now.

From 1999 to 2014, 10 patients (5 women and 5 men) with a malignant triton tumor were treated at our institution. All these cases were followed-up and patient charts were analyzed for outcome.

In our study, 3 cases of the Malignant triton tumors originate in the head, 2 cases in the joints, 2 cases in the retroperitoneum, 2 cases in the soft tissues of the thoracic wall, and 1 case in the prostate. Neoplasm associated with pain was the main manifestation. Patients have a poor prognosis. Completely surgical excision of the tumor is the only treatment. Additional radiation or chemotherapy show little effect.

Malignant triton tumor is a rare sarcoma. The high probability of developing local recurrence and distant metastases could account for its poor prognosis.

Abbreviations: AWD = alive with disease, CTx = chemotherapy, DOD = died of disease, LOST = lost to follow-up, MPNST = malignant peripheral nerve sheath tumor, MTT = malignant triton tumor, NF1 = neurofibromatosis type 1, RTx = radiotherapy, STs = surgical times.

Keywords: malignant peripheral nerve sheath tumor, malignant triton tumor, rhabdomyoblastic differentiation, treatment

1. Introduction

Malignant triton tumor (MTT) is a relatively rare subtype of malignant peripheral nerve sheath tumors (MPNST), which presents rhabdomyosarcomatous differentiation and has a particularly aggressive clinical course. Less than 10% of MPNSs could be attributed to MTTs. The immunohistochemical staining of MTT is positive for desmin, focal positive for actin, and myogenin of skeletal muscle and myoglobin. Up to now, approximately 100 cases of MTTs have been reported, but most are case reports. Our study displays the largest cases with MTT treated in our hospital. As far as we know, this is the largest report regarding MTTs that has been recorded in the literature worldwide.

2. Patients and methods

We reviewed the medical charts of 10 patients treated for malignant triton tumor between 1999 and 2014 in Peking Union Medical College Hospital, China. There were 5 female and 5 male patients with a mean age of 42.4 years (range 13–62, Table 1).

Pathologic biopsy revealed malignant peripheral nerve sheath tumor together with rhabdomyosarcomatous differentiation. Definite histological analysis indicated spindle cell tumor with prominent mitotic figures. Immunohistochemical stains revealed positive for S-100 in malignant cells, and positive for desmin in rhabdomyoblasts (Fig. 1).

Our study shows that 3 cases of the malignant triton tumors originate in the head, 2 cases in the joints, 2 cases in the retroperitoneum, 2 cases in the soft tissues of the thoracic wall, and 1 case in the prostate.

In 2 cases both chemotherapy and radiation therapy were given postoperatively, in 2 cases only local radiation therapy postoperatively was adopted, only 1 case received chemotherapy postoperatively. Patients' demographics, course of disease, tumor location, primary symptom, data on surgical procedure, adjuvant therapy, and follow-up data are presented in Table 1.

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Table 1

Patients' demographics presenting sex, age, nationality, course of disease, location, primary symptom, surgery, followed-up, recurrence, surgical times, status, chemotherapy, radiotherapy.

NO.	Sex	Age	Nationality	Location	Course of disease	Symptom	Surgery	Follow-up (M)	Recurrence	STs	Status	CTx/RTx
1	F	13	Hui	Retroperitoneum	2M	Diet and weight loss	Rescetion	1M	Thoracic vertebra	3	DOD	Y/Y
2	M	31	Han	Head	2M	Left facial swelling	Rescetion	4M	In situ	2	LOST	N/Y
3	M	34	Han	Prostate	2Y	Micturition and acute urinary retention	Rescetion	6M	-	1	LOST	N/N
4	M	38	Han	Thoracic wall	2Y	Chest tumor	Rescetion	8M	-	1	LOST	N/N
5	F	40	Hui	Left hip	6M	Tumor and pain	Rescetion	12M	In situ	3	AWD	N/N
6	F	45	Han	Right nasal cavity	1M	right nasal obstruction symptom	Rescetion	12M	Lymph nodes	2	DOD	Y/Y
7	F	46	Han	Nasal cavity	5Y	Headache and nasal bleeding	Rescetion	6M	-	1	DOD	N/N
8	F	57	Han	Retroperitoneum	10D	Right Abdominal mass	Rescetion	24M	In situ	1	AWD	N/Y
9	M	58	Han	Right knee/femur	6M	Right knee pain	Biopsy	18M	-	1	AWD	Y/N
10	M	62	Han	Thoracic wall	2M	Chest tumor and pain	Rescetion	12M	-	1	DOD	N/N

AWD = alive with disease, CTx = chemotherapy, DOD = died of disease, LOST = lost to follow-up, RTx = radiotherapy, STs = surgical times.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

3. Results

Overall, in 9 of 10 patients wide resection were achieved (Fig. 2). In 1 patient a biopsy was received. With a mean follow-up of 10 months (range 1–24), 3 were alive with disease, 4 died of disease, 3 were lost of follow-up. Five recurrences were detected after the wide resection of MTT, local recurrence in 2 cases, distance metastasis in 3 cases. Two patients received 3 times resection, 2 patients received 2 times resection (Table 1).

4. Discussion

In 1932, Masson[1] was the first to describe the disease. In 1973, this disease was name after malignant triton tumor (MTT) by Woodruff et al,[2] because Titron salamander owns the similar ability to regenerate extra limbs consisting of nerve, muscle and bone tissue,[3] and 3 basic diagnostic criteria were established that must be fulfilled to diagnose a sarcoma as MTT. The tumors which derived from the peripheral nerve, in ganglioneuroma, or in patients with type 1 neurofibromatosis, or represents metastasis from the above-mentioned tumors. The tumor has growth characteristics of Schwann cell tumors. Rhabdomyoblastic elements can be observed within the body of tumors. Immunohistochemical staining assists the identification of the origination of tumor cells. S-100 protein positivity is the confirmation of nerve sheath differentiation, whereas the positivity of desmin, actin, and myogenin is the confirmation of rhabdomyoblastic differentiation. In these present cases, the neoplasm was both positive for S-100, and desmin. This indicated that nerve sheath as well as rhabdomyoblastic components were contained in tumor.[4] MTTs can display as sporadic cases, in young patients, approximately 60% to 70% accompany with neurofibromatosis type 1 (NF1). And deregulation of p53 may play a part in the genesis of MTT.[5]

MTT is a very rare entity, few case reports and series have been documented. This study is the largest study in one institute to date, total consists of 10 MTT cases. Literature published previously has demonstrated the poor prognosis of MTT,[6] because local recurrence and distance metastasis was common following resection, while the lymph node involvement was infrequent. Our study report 2 cases recurrence in situ and distance metastasis in 3 cases, the rate of recurrence was equal to the study by Woodruff et al[2] which presents the overall local recurrence rate of MTT was 50%. Previously study reported the 5-year survival rate of MTT was 11% to 12%, the median survival time of MTT patients was about 13 months.

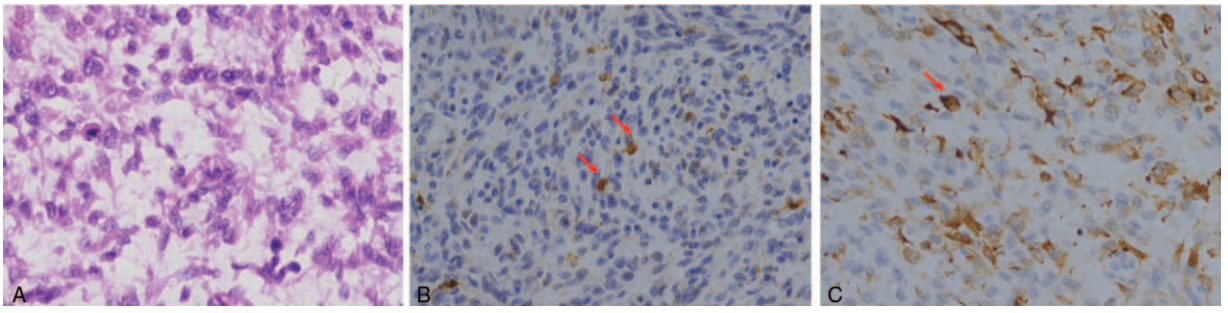

Figure 1. A. Pathologic biopsy revealed malignant peripheral nerve sheath tumor together with rhabdomyosarcomatous differentiation. (Hematoxylin-eosin, original magnification 200). B. Immunohistochemical stains revealed that the malignant cells were positive for S-100 (arrows), C. and the rhabdomyoblasts were positive for desmin (arrow).

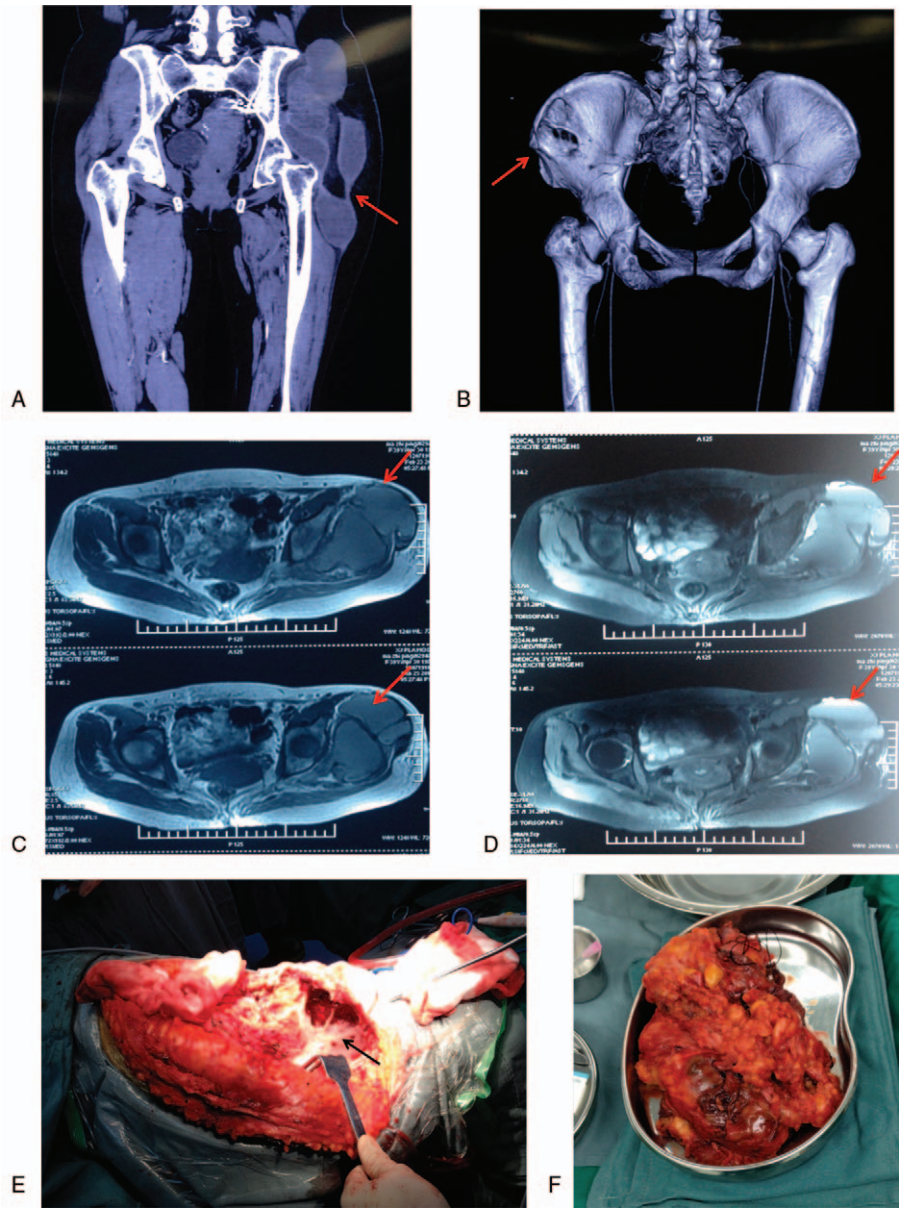

Figure 2. A–D, X-ray, 3-D CT, and MRI of the left hip showing a mass with division and bone destruction in a 39-year-old female patient. E, Photographs of the intraoperative situs. F, Photographs of the resected malignant triton tumor with partial iliac bone. CT = chemotherapy, MRI = magnetic resonance imaging.

Management strategies toward MTTs are difficult to established because of its rarity.[7] To date, the mainstay of treatment is complete surgical resection, meanwhile adjuvant radiotherapy is used frequently to strengthen the control of local disease.[8] In our study, all the patients except 1 (the patient give up treatment) underwent the complementary operation with wide excision, and the 3 case each received at a total dose of 60/100/40 Gy radiotherapy, but 2 of them have died post the operation shortly. Chemotherapeutic regimen maybe used, but the result is unclear.

In conclusion, MTTs are uncommon, aggressive tumors with both muscle and nerve components. The high probability of developing local recurrence together with distant metastases leads to poor prognosis. The only therapeutic method with curative purpose at the moment is radical surgical excision. Additional radiation or chemotherapy shows little effect.

Author contributions

Conceptualization: Yong Liu.
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Supervision: Dachun Zhao, Huanwen Wu.
Writing – original draft: Yanyan Bian.
Writing – review & editing: Yanyan Bian, Xiang Yongbo.

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