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Case Report

Malignant triton tumor of the prostate: a case report

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Abstract: Malignant triton tumor (MTT) is a malignant peripheral nerve sheath tumor (MPNST) with rhabdomyoblastic differentiation. It is very rare and has an aggressive clinical course and poor prognosis. A 27-year-old male patient presented with complaints of difficulty in urination for 3 months. Computerized tomography (CT) imaging demonstrated a huge irregular mass localized in the prostate, with invasion into the bladder, seminal vesicle, cavernous bodies and rectum. In addition, generalized lymphadenopathy was shown at the pelvic and inguinal regions. After ultrasound-guided transrectal core biopsies of the prostate, the histopathological finding of rhabdomyoblasts among malignant Schwann cells in a tumor arising from a peripheral nerve supported by immunostaining (IHS) with vimentin, desmin, MyoD1 and S-100 confirmed the diagnosis. The patient underwent urinary diversion, palliative radiotherapy and interstitial chemotherapy and received traditional Chinese medicine. This patient appears to be the second reported case of the localization of an MTT in the prostate. Early diagnosis, the extent of surgical excision, and comprehensive treatment to improve the survival rate and the quality of life for patients with MTT are very important.

Keywords: Malignant peripheral nerve sheath tumor, malignant triton tumor, S-100 protein, vimentin protein, desmin protein

Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a rare soft tissue neoplasm that may consist of tissues such as bone, cartilage, glandular epithelium, adipose tissue, or even squamous cells [1]. Tumors with rhabdomyoblastic differentiation and malignant Schwann cells are referred to as malignant triton tumor (MTT), which constitutes approximately 5% of all MPNSTs [2, 3]. MTT is to date very rarely reported in the English literature [4] and originates predominantly in the head, neck and trunk regions [5]. To the best of our knowledge, there have been only two reports showing an MPNST localized in the prostate [6, 7]. This report regards an MTT localized in the prostate in a 27-year-old man.

Case report

In Affiliated Hospital of Guizhou Medical University, a 27-year-old male patient presented with complaints of difficulty in urination for 3 months starting in March 2013. His medical and family histories were unremarkable and negative for prior radiation exposure and neurofibromatosis type 1 (NF-1), respectively. On physical examination, there were no signs of neurofibromatosis disease, such as subcutaneous nodules or cafe au lait spots.

Computerized tomography (CT) imaging revealed a large irregular mass (10.5×7.5×5.0 cm) localized in the prostate and invading the bladder, seminal vesicle, cavernous bodies and rectum. In addition, generalized lymphadenopathy was observed in the pelvic and inguinal region (**Figure 1A**).

Ultrasound-guided transrectal core biopsies of the prostate were performed. Microscopic examination showed a partially encapsulated cellular mass composed of interlacing fascicles of hyperchromatic serpentine spindle cells. Large pleomorphic cells with abundant eosinophilic cytoplasm and rounded eccentric nuclei with large nucleoli were present (**Figure 1B**, **1C**). The tumor cells were stained with antibodies against S-100 protein, vimentin, desmin, MyoD1,

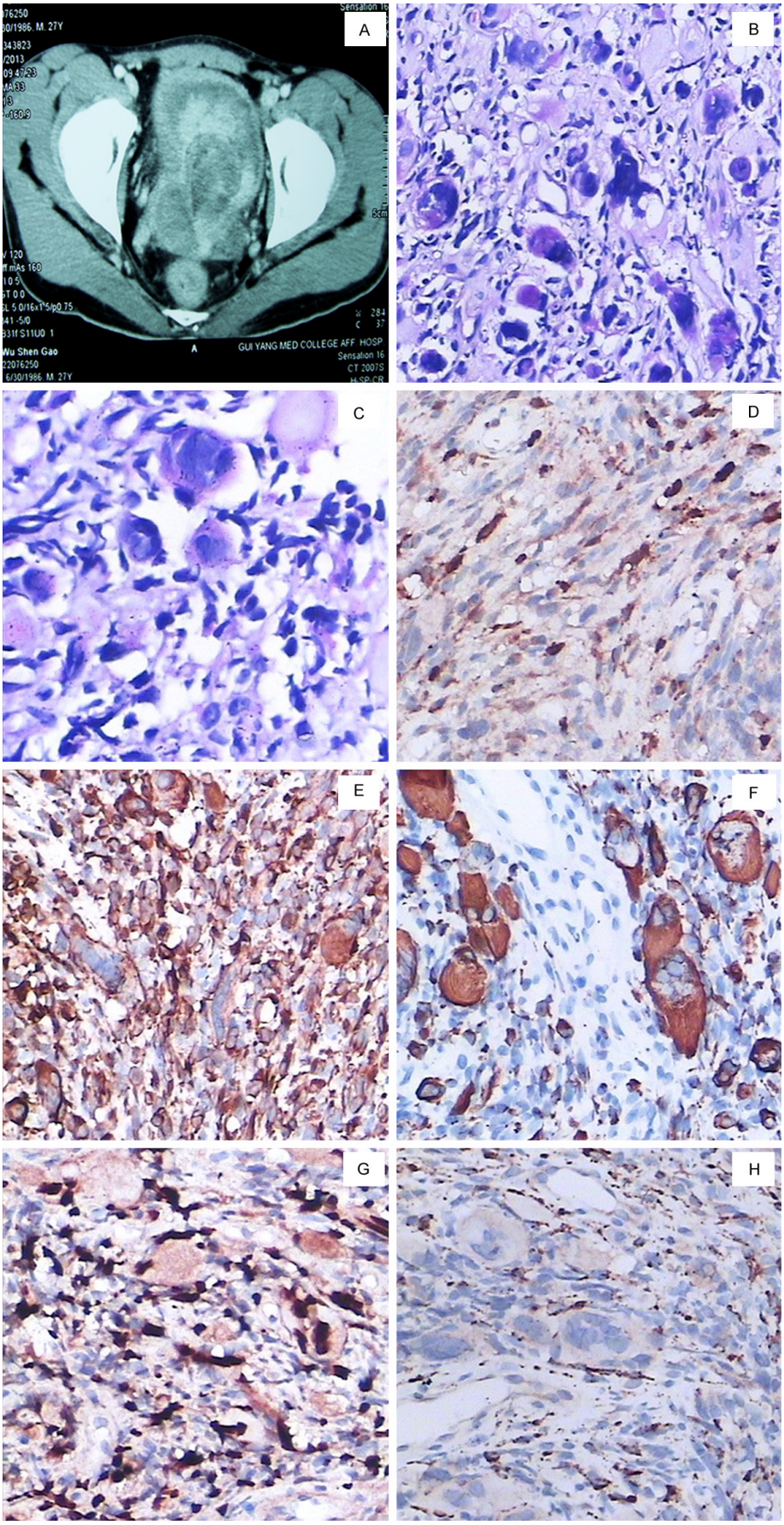
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Figure 1. A. Computed tomography (CT) scan of the pelvis at initial presentation. CT imaging demonstrated a huge irregular mass localized in the prostate, with invasion into the bladder, seminal vesicle, cavernous bodies and rectum. B and C. Histopathology of the resected specimen: ×200 and ×400. A partially encapsulated cellular mass composed of interlacing fascicles of hyperchromatic serpentine spindle cells. Large pleomorphic cells with abundant eosinophilic cytoplasm and rounded eccentric nuclei with large nucleoli were present. D. S-100 protein positivity (×200). E. Vimentin positivity (×200). F. Desmin positivity (×200). G. MyoD1 positivity (×200). H. CD57 positivity (×200).

Table 1. Summary of previous similar case reports of MTT

Location	Treatment
Medial side of the left arm [15]	Surgical excision
Left neck [18]	Wide surgical excision
The anterior mediastinum [21]	Palliative chemoradiotherapy and interstitial chemotherapy and received Traditional Chinese medicine

CD57, GFAP, CD68, SMA, DOG-1, P63, CK, CKH, CD34, AMACR and CD117. The cells exhibited a positive reaction with S-100, vimentin, desmin, and MyoD1, and a few cells had a positive reaction with CD57 (**Figure 1D-H**); however, the cells had a negative reaction with GFAP, CD68, SMA, DOG-1, P63, CK, CKH, CD34, AMACR and CD117.

After extensive discussion with urologists of the Affiliated Hospital of Guiyang Medical College and discussing the illness with the patient and his family members, radical prostatectomy was ruled out. The patient underwent urinary diversion, palliative radiotherapy and interstitial chemotherapy and received traditional Chinese medicine. The patient has survived with the disease for more than 1 year.

Discussion

MTT is a subgroup of MPNST that displays rhabdomyosarcomatous differentiation and follows a particularly aggressive course [8]. The original histological criteria for establishing the diagnosis of MTT were proposed by Woodruff in 1973 [3]. While it is a rare tumor, the head and neck region is one of the most frequent sites of involvement of MTT [5]. To the best of our knowledge, MTT localized in the prostate has rarely been reported in the English literature: this report appears to be the second case [9].

The diagnosis of MTT requires cytological analysis. The rates of association reported in the literature between malignant triton tumors and NF-1 range from 23% to 69% [10]. In this case, testing for NF-1 was not performed because the patient did not have any clinical features suggestive of NF-1. Previous research has shown the potential of schwannoma cells to exhibit myogenic differentiation [11]. The diagnosis is based on microscopic examination along with immunohistochemical staining (IHS). Nerve sheath differentiation is confirmed by S-100 protein and CD57 positivity [12, 13], whereas rhabdomyoblastic differentiation is confirmed by positivity to myoglobin, desmin, muscle-specific actin, sarcomeric actin, myoD and myogenin [14]. In this case, the cells were positive for S-100, vimentin, desmin, and MyoD1, and a few cells were positive for CD57, which indicated nerve sheath and rhabdomyoblastic components.

The prognosis of MTT is generally poor; its 5-year survival rate is only 5% to 15%, in contrast to MPNST, which is 50% to 60% [15]. Previous studies have shown that the prognosis of MTT depends on the location (better for the head, neck and extremities; worse for the trunk, buttocks, retroperitoneal and central nervous system), degree of differentiation (better for well-differentiated tumors, worse for undifferentiated ones), completeness of surgical margins, size of the tumor, performance status and comorbidities, and Ki67 labeling index [16-19]. Moreover, other prognostic factors should be confirmed for MTT in further research. No standard or ideal treatment for MTT has been established. Various recommendations have been proposed in previous studies, including radical excision, radical excision followed by radiotherapy and chemotherapy, excision and radiotherapy and traditional Chinese medicine [10, 20-22]. A summary of previous similar case reports of MTT in the past 3 years is shown in **Table 1**.

In summary, early diagnosis, extent of surgical excision, and comprehensive treatment to

improve the survival rate and quality of life for patients with MTT are very important, which will require more intense elucidation of this rare disease by further studies. More investigations are warranted to define the optimal treatment.

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Disclosure of conflict of interest

None.

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