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LETTER TO THE EDITOR

Male Health

Clinical features in a man with primary synovial sarcoma of the spermatic cord

Gong-Cheng Wang[1,*], Xue-Qin Li[2,*], Yun-Yan Wang[1], Zhong-Ying Guo[3], Pei-Jin Hou[1]

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Dear Editor,

Synovial sarcoma (SS) is a rare malignant neoplasm in young adults.[1] It commonly arises around the major joints or tendon sheaths. Furthermore, it has been described in numerous locations unrelated to joint structures, such as the heart, kidney, prostate, *etc*., which may lead to a diagnosis pitfall. Here, we present an extremely rare case of primary SS originating in the spermatic cord.

A 53-year-old man presented at our hospital with a complaint of asymptomatic right groin mass for three months. On physical examination, an irreducible firm mass was detected in the right inguinal region. Blood testing, including the tumor marker, showed no abnormality. Ultrasonography and computed tomography (CT) demonstrated an approximately 6 cm × 3 cm × 2 cm well-defined mass with heterogeneous density, including myxoid and fat component, extending from the right spermatic cord to the right inguinal subcutaneous layer **(Figure 1a)**. There was no radiographic evidence of any regional lymph node involvement or invasion involving a contiguous structure on CT scan. From these findings, we diagnosed his disease as primary malignant tumor of the spermatic cord without metastasis. After obtained informed consent from the patient, radical orchiectomy was carried out. During surgery, a 6 cm × 3 cm × 2 cm solid mass was discovered which was adherent to the spermatic cord, but simultaneously mobile and free from attachments with other soft tissue elements. The resected tumor was elongated and well circumscribed, measuring 6 cm × 3 cm × 2 cm in size **(Figure 1b)**. On cut section, the tumor showed a whitish-yellow appearance with focal hemorrhage and without calcification **(Figure 1c)**. Histologically, the tumor showed the monotonous proliferation of spindle-shaped cells **(Figure 1d)**. These cells were relatively small and uniform in size with fusiform or ovoid nuclei and a small or inconspicuous nucleolus. In some areas, coagulation necrosis was noted, and numerous mitotic figures were seen. Immunohistochemical study showed *Ki67* was partially positive in tumor cells **(Figure 1e)**, whereas *MyoD1*, *CD34*, *Desmin*, *S-100*, *WT1*, *SMA*, *CK5/6*, *EMA*, and *D2-40* were immunonegative. Furthermore, by fluorescence *in situ* hybridization (FISH) detection, rearrangement of the *SYT* gene (18q11.2) was detected **(Figure 1f)**. Fifteen days after the radical orchiectomy, the patient was referred to the oncology department and received adjuvant chemotherapy with epirubicin hydrochloride (60 mg m^{-2}) and cisplatin (30 mg m^{-2}). Besides that, the patient also got adjuvant radiation therapy (54 Gy). The patient has not shown any evidence of recurrence for 36 months follow-up.

The incidence of spermatic cord tumors is approximately 0.3 cases per million, and sarcoma is the most common malignance tumors.[2] SS is a rare tumor, with approximately 800 new cases diagnosed in the United States per year.[3] It accounts for 6%–9%[1] of all soft sarcomas and < 1% of all malignancies.[3] It is most prevalent in adolescents and young adults aged between 15 and 40 years, and it occurs equally in males and females with no predilection for either sex.[1] However, primary SS originating in the spermatic cord is extremely rare. Considering it may slow growth, may have a benign appearance on imaging studies, may vary in size, and may have pain similar to that associated with trauma, SS is thought to be the most commonly misdiagnosed soft tissue malignancy.[4]

Imaging examination is the first step for diagnosing SS.[5] SS appears differently in different images. Most of the time, they are well defined and appear to be soft tissue masses. However, 30% of soft tissue masses demonstrate calcification.[1] The degree of calcification is relevant to the grade of malignancy: more calcification signifies less malignancy. In our case, there was no calcification in the tumor; it might indicate that the malignant degree of the case was higher.

Synovial sarcoma may be classified into three subtypes: biphasic, monophasic, and poorly differentiated. SS expresses many immunohistochemical markers, which are helpful in the diagnosis of SS and differential diagnosis with other soft tissue sarcomas such as fibrosarcoma and malignant peripheral nerve sheath tumor. In our case, the tumor was monophasic fibrous type, and it was partially positive for *Ki67*. Although positive immunostaining for *keratin* was seen in nearly all biphasic type and in many of the monophasic fibrous type, it was also reported that balanced translocation between chromosomes X and 18, t (X; 18) (p11.2; q11.2) was usually the only abnormality, and occurred in virtually all (>90%) variants of SS.[6] In our case, we made a definite diagnosis of SS by detecting *SYT–SSX* fusion using FISH analysis.

The detection of *SYT-SSX* fusion has been established clinically as a molecular diagnostic test for this tumor; therefore, this translocation is considered the driving oncogenic event in the development of SS.[6] About two-thirds of cases contain an *SYT–SSX1* fusion and another third contain an *SYT–SXX2* fusion. However, the *SYT–SXX4* fusion

[1]Department of Urology, Huai'an First People's Hospital, Nanjing Medical University, Huai'an, China; [2]Department of Gerontology, Huai'an First People's Hospital, Nanjing Medical University, Huai'an, China; [3]Department of Pathology, Huai'an First People's Hospital, Nanjing Medical University, Huai'an, China.
*These authors contributed equally to this work.
Correspondence: Dr. PJ Hou (doctorhoupj@163.com)
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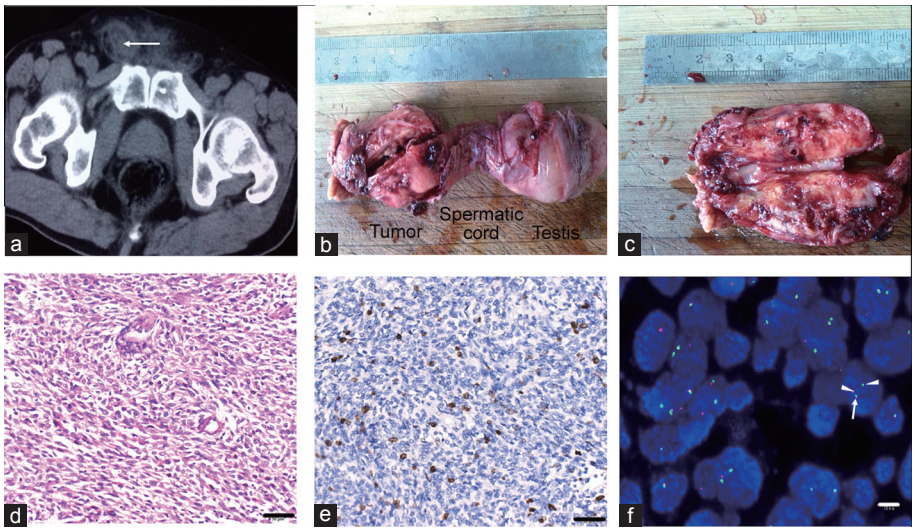

Figure 1: Characterization of the specimen. (**a**) Computed tomography findings. An approximately 6 cm × 3 cm × 2 cm well-defined mass with heterogeneous density, including myxoid and fat component was found, it was extending from the right spermatic cord to the right inguinal subcutaneous layer. (**b** and **c**) Gross findings. The resected specimen forms a well-encapsulated solid tumor originating from the spermatic cord (6 cm × 3 cm × 2 cm). On cut section, the tumor showed a whitish-yellow appearance with focal hemorrhage and without calcification. (**d**) H and E staining. Monophasic tumor with spindle cells in fascicular growth pattern is seen (light microscope, scale bar = 50 μm). (**e**) Immunohistochemistry. Tumor cells are partially positive for *Ki67* (light microscope, scale bar = 50 μm). (**f**) Tumor cells comprise a normal fusion signal (consisting of a 650-kb spectrum red probe that lies distal from the *SYT* gene and a 1040-kb spectrum green probe that lies proximal to the *SYT* gene, arrow) and a pair of split signals (arrowheads), indicating a rearrangement involving the *SYT* gene (fluorescence *in situ* hybridization, scale bar = 10 μm).

is rare. In approximately 10% of patients, the *SYT–SXX1* fusion and the *SYT–SXX2* fusion exist in the same sarcoma.[7] Some authors have observed an association between the type of *SYT–SSX* fusion and histological glandular differentiation, biphasic histology occurs in 38.6% of *SYT–SSX1* tumors but only 3.3% of *SYT–SSX2* tumors. Furthermore, *SYT–SSX2* tumors did better than those with *SYT–SSX1* for overall survival.[8]

The best treatment for SS is complete surgical resection with negative margins. Primary SS of the spermatic cord is typically recommended radical orchiectomy. However, there was an approximately 50% rate of local-regional recurrence even after such definitive surgery.[9] The factors determining high risk include size > 5 cm, deep-seated, inadequate surgical resection, and local recurrence.[10] Although adjuvant chemotherapy and radiation therapy for SS are still controversial, they should be considered particularly in high-risk patients.[9,11] In our case, the patient received adjuvant chemotherapy and radiotherapy after radical orchiectomy because he was in patients at high risk of recurrence.

In conclusion, primary SS of the spermatic cord is very rare and difficult to diagnosis and treat. It should be considered in the differential diagnosis in patients with an inguinal mass in order to perform the proper therapeutic strategy. Molecular analysis may contribute to the final diagnosis occurring in this unusual location.

AUTHOR CONTRIBUTIONS

GCW and XQL carried out the experiments, collected clinical data and drafted the manuscript. YYW and ZYG helped to collect some clinical data. PJH conceived of the study, and participated in its design and coordination and helped to draft the manuscript. All authors read and approved the final manuscript.

COMPETING INTERESTS

The authors declare that they have no competing interests.

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