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Leiomyosarcoma of the spermatic cord: case report and literature review

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ABSTRACT
Leiomyosarcoma of the spermatic cord is a rare entity. It can arise from any mesenchymal cell of its structure. It occurs mainly in elderly patients. It presents as a firm and painless intrascrotal mass. We present a case of a 30-year-old man with a 3-year history of a painless left-sided scrotal mass. Ultrasound examination showed a scrotal mass. Further work-up revealed no distant metastasis before surgery. A left radical 1-block orchidectomy with epididymis and spermatic cord was performed. The final histopathologic examination showed a pattern of leiomyosarcoma of the spermatic cord. Considering the result of the histopathological examination, the Oncological Council did not recommend any additional surgery, but only watchful waiting. Almost 4 years of follow-up have passed without additional treatment. Primary scrotal leiomyosarcoma is an exceptional entity with only 100 cases reported in the literature. Due to its rarity, additional studies are needed to better define the optimal therapeutic management. A literature review and treatment strategy are discussed.
Keywords: spermatic cord; sarcomas; epidemiology; treatment.

INTRODUCTION

Scrotal leiomyosarcomas are divided into 2 categories: paratesticular and intratesticular [1]. Leiomyosarcoma of the spermatic cord is an extremely rare tumor in men [2]. It occurs mainly in elderly patients, but also has been reported in younger persons. Its incidence is estimated to be less than 1% of all urological neoplasms [3, 4]. We believe that nearly 100 cases of leiomyosarcoma of the spermatic cord have been reported since its first description by Patel in 1907 [5, 6].

It may arise from mesenchymal cells of the spermatic cord. The most common histopathologic subtypes are liposarcoma (20–32%), leiomyosarcoma (19–32%), rhabdomyosarcoma (11–24%) and fibrosarcoma (less than 10%) [7, 8]. On average, these tumors are relatively small, usually no larger than 2 cm [7, 9]. Spread of leiomyosarcoma of the spermatic cord is thought to be primarily by the hematogenous route. The first case of paratesticular sarcoma was reported by Lesauvage in 1845 [3], but the first isolated report of sarcoma of the spermatic cord was given much later, in 1907 [6]. Mostly, these tumors are asymptomatic, benign in nature and present as a small mass in the genital area [10]. For diagnosis, radiologic methods such as ultrasound, computed tomography (CT) or magnetic resonance imaging (MRI) are necessary, but the diagnosis of spermatic cord leiomyosarcoma is difficult and practically undiagnosable preoperatively. Therefore, a secondary complementary resection is often necessary when the histopathologic diagnosis of leiomyosarcoma is made after the initial surgery [11].

As with many other soft tissue sarcomas, the prognosis for patients with leiomyosarcoma of the spermatic cord is poor. Some authors have suggested a probable 5-year survival rate of 10–15% [12, 13].

CASE REPORT

A 30-year-old patient, was admitted to our department with a small tangerine-sized mass on the left side of his scrotum. Physical examination revealed a round, painless, smooth and firm mass located close to the upper pole of the left testis. The patient followed its development for 3 years; initially, it was very small. The patient discovered it accidentally, he did not feel any pain from his testis or other structures of the genitourinary system. The remaining scrotal organs were normal on physical examination. The regional inguinal lymph nodes were not palpable. The patient had no history of any trauma or past medical history. Ultrasound examination revealed a normal left testis (Fig. 1) and a well-defined hypoechogenic mass of approx. 6.5 x 5.0 x 4.0 cm near the upper pole of the left testis (Fig. 2). Minimal internal vascularity was demonstrated by color Doppler. Laboratory tests including alpha-fetoprotein, beta choriongonadotropin and lactic acid dehydrogenase were all within normal limits. A radical right inguinal orchidectomy under spinal anesthesia was performed by our urologic team. During surgery, a round mass was found near the upper pole of the testis and was completely resected in 1 block with the testis, epididymis, and spermatic cord (Fig. 3). The inguinal lymph nodes were of normal size and not removed. Histopathological results – No. 103002205 from the Pathomorphologic Department Diagnostic Consillo, Poznań – macroscopically: in 1 block the left testis with epididymis and spermatic cord with tumor were delivered. The tumor was located around the upper pole of the testis and measured 6.5 x 5 x 4 cm. The left testicle was about 4 x 3 x 2.5 cm, and the spermatic cord was about 8 cm long. The cut surfaces of the tumor showed

solid greyish-white nodules. Microscopically, the cells were arranged in roots and tubules separated by connective tissue, all cells were lined by flattened epithelium with cytoplasmic vacuoles. Fusiform neoplasm with focal pronounced nuclear atypia with approx. 15 mitoses/10 HPF. Foci of necrosis were demonstrated. The intermingled stroma showed abundant fibrous and fatty tissue with smooth muscle. Immunohistochemical positivity for mesothelial markers confirms the mesothelial origin of the tumor, which is most consistent with the diagnosis of leiomyosarcoma of the seminal cord. The following immunophenotypic alterations were observed: smooth muscle actin (+), vimentin (+), pan-cytokeratin (+), cluster of differentiation 17 (–), cluster of differentiation 34 (–), beta-catenin (–), and Ki67 – about 80%. The tumor was limited and completely removed. The epididymis and testis were without neoplastic infiltration. Tumor nodules metastas classification was pT2R0. Figure 3 shows the histopathological features of neoplastic leiomyosarcoma cells. Taking into account the results of histopathologic examination, the Oncological Council did not recommend additional surgery, but only watchful waiting. Four-year post-operative follow-up showed no recurrence.

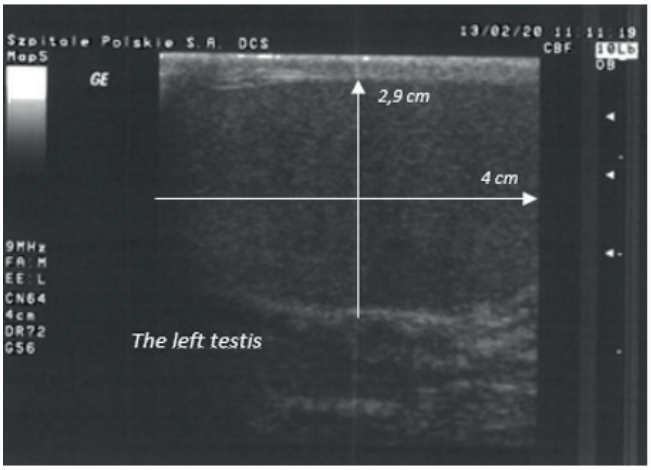

FIGURE 1. Ultrasound showed a normal left testis with normal echogenicity

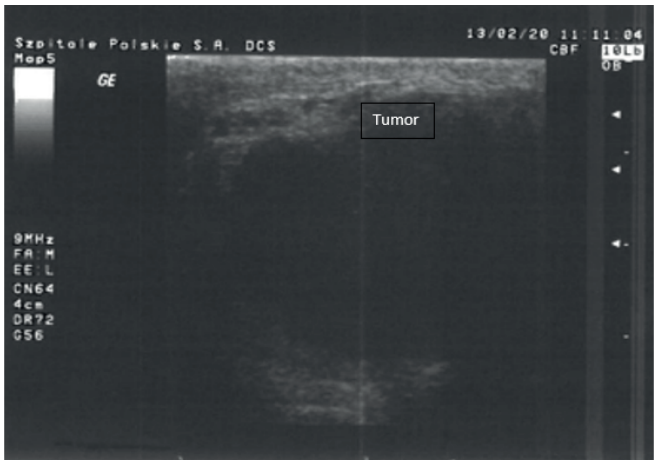

FIGURE 2. Ultrasound showed the tumor of irregular shape

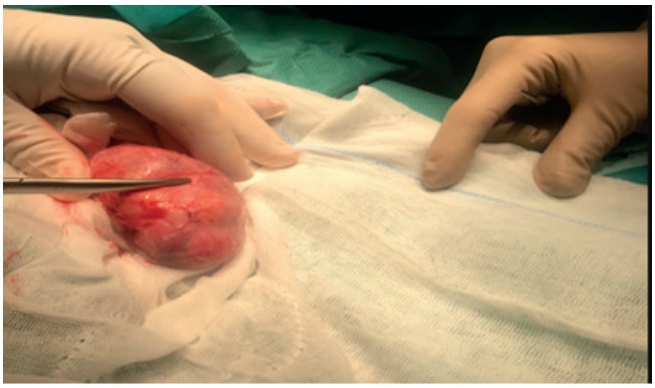

FIGURE 3. Intraoperative view of the tumor

DISCUSSION

Leiomyosarcoma of the spermatic cord is a rare disease and occurs mainly in elderly patients [4, 7]. Its incidence is estimated to be less than 1% of all urological neoplasms [3]. As of 2019, only 110 cases of leiomyosarcoma of the spermatic cord were described worldwide. It can arise from any mesenchymal cell of the spermatic cord [5]. The most common histopathologic subtypes are liposarcoma (20–32%), leiomyosarcoma (19–32%), rhabdomyosarcoma (11–24%) and fibrosarcoma (less than 10%) [8, 12]. On average, these tumors are relatively small, usually no larger than 2 cm [9]. The first case of paratesticular sarcoma was reported by Lesauvage in 1845 [3]. Mostly, these tumors are asymptomatic, benign in nature and present as a small mass in the genital area [12]. For diagnosis, radiologic methods such as ultrasound, CT or MRI are necessary but the diagnosis of spermatic cord leiomyosarcoma is difficult, and practically undiagnosable preoperatively [13]. Therefore, secondary complementary resection is often necessary when the histopathologic diagnosis of leiomyosarcoma is revealed after the first surgery [11, 13].

No treatment protocol has been established for paratesticular leiomyosarcoma due to the rarity of the disease [7]. The standard treatment is radical orchidectomy with high ligation of the spermatic cord [13]. The benefit of adjuvant chemotherapy, radiotherapy, and retroperitoneal lymphadenectomy is not well understood and may vary according to surgeon preferences [12, 13].

CONCLUSIONS

The purpose of this presentation was to show that leiomyosarcoma can affect young people and that its size can sometimes exceed the size of a normal testicle. Although the recommendation of watchful waiting issued by the Oncological Council after surgery may seem controversial, it seems correct so far, even though the urologists opted for the diagnostic removal of the regional inguinal nodes for histopathologic examination. The rarity of leiomyosarcoma poses a challenge to urologists

in its diagnosis and treatment. And therefore further reports are needed to increase the group which will allow to better define optimal management strategies.

Informed consent

Informed consent was obtained from the patient for the publication of his case along with appropriate images. No information was provided that would allow his identification.

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