CASE REPORT

Strangulated inguinal hernia accompanied by paratesticular leiomyosarcoma

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ABSTRACT

Sarcomas that arise from the spermatic cord constitute 2.1% of soft tissue sarcomas and are observed at a rate of 1% to 2% in the genitourinary system. A 74-year-old patient presented at the emergency department with complaints of groin pain and swelling persisting for 3 days. On physical examination, a firm mass that was approximately 5x5 cm in size and could not be reduced was observed at the right inguinal area as well as a right inguinal hernia. Computed tomography revealed a large hernia sac in the right inguinal area and a mass that was 77x55 mm in size within the hernial sac. A radical orchiectomy and hernia repair were performed. A diagnosis of leiomyosarcoma was made based on the pathological evaluation of the mass. At the 7-month postoperative follow-up, no local relapse or distant metastasis was found. To conclude, although paratesticular leiomyosarcoma is rare, it should be kept in mind as one of the possible diagnoses for older patients presenting with an inguinal mass.

Keywords: Inguinal hernia; leiomyosarcoma; paratesticular tumor.

INTRODUCTION

Sarcomas constitute less than 1% of all malignant tumors, and less than 3% to 5% of these arise from the genitourinary system, particularly from the paratesticular region. The paratesticular region consists of the spermatic cord, testicular tunics, epididymis, and the vestigial remnants. Tumors in this region contain multiple structures that are of embryologically different origins and show different characteristics, and while 70% of them are benign, 30% of these tumors are malignant in nature. Paratesticular leiomyosarcomas account for 1% to 2% of the tumors that stem from the genitourinary system.[1] Tumors that arise from the spermatic cord structure may be observed with inguinal hernia and this condition is often detected during surgery.[2]

A case of strangulated indirect inguinal hernia and a mass originating from the spermatic cord that required radical orchiectomy is presented.

CASE REPORT

A 74-year-old patient presented at the emergency department with the complaints of groin pain that had been persisting for 3 months, and a solid swelling in the groin present for the last 3 days. There was no urinary system symptom or history of trauma, but the physical examination revealed a mass extending from the right inguinal canal to the scrotum and a right indirect hernia. The lump, which was thought to be strangulated, could not be reduced. Routine biochemical and hematological test results were within the normal range. Lower abdominal and pelvic computed tomography (CT) scans with contrast were performed and revealed a large right-side inguinal hernia that did not contain bowel loops and a structure that occupied 77x50 mm of space within the sac (Fig. 1a). Concurrent ultrasonography (US) revealed a mass located at the mid inguinal canal that was hypoechoic with lobulated borders, approximately 60x65 mm in size, along with a 2 cm opening observed

Cite this article as: Ağca B, İşcan Y, Aydın T, Şahin A, Memişoğlu K. Strangulated inguinal hernia accompanied by paratesticular leiomyosarcoma. Ulus Travma Acil Cerrahi Derg 2019;25:307-310.

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Ulus Travma Acil Cerrahi Derg 2019;25(3):307-310 DOI: 10.5505/tjtes.2018.68709 Submitted: 11.01.2018 Accepted: 24.10.2018 Online: 25.10.2018

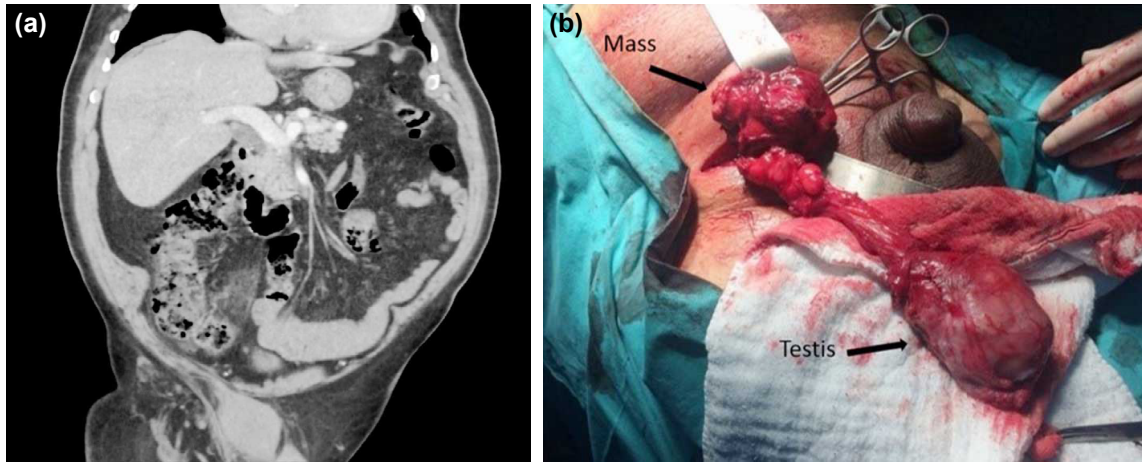

Figure 1. (a) Hernia in right inguinal region and a mass of 77x50 mm within the hernia sac. **(b)** Sarcoma located in right spermatic cord and enlarged testis.

in the transverse fascia. It was found that intestinal loops had herniated through this opening. Surgery was performed with a preliminary diagnosis of strangulated hernia. Bassini's incision was performed. Upon inguinal exploration, a right-sided strangulated indirect inguinal hernia and a mass lesion that was thought to have arisen from the spermatic cord was observed adjacent to the hernia sac (Fig. 1b). It was observed that the right spermatic cord was thickened and the right testis was approximately more than 3 times larger than the left testis. A perioperative urology consultation was conducted. The mass was observed to have originated from the paratesticular region and extended to the right scrotum, with cord ligation at the inguinal ring level. A radical orchiectomy was performed. The small bowel loops inside the hernia were seen to be healthy and were reduced back to the abdomen, and an indirect hernia repair was performed using the tension-free mesh repair method. The patient was discharged uneventfully on the second postoperative day.

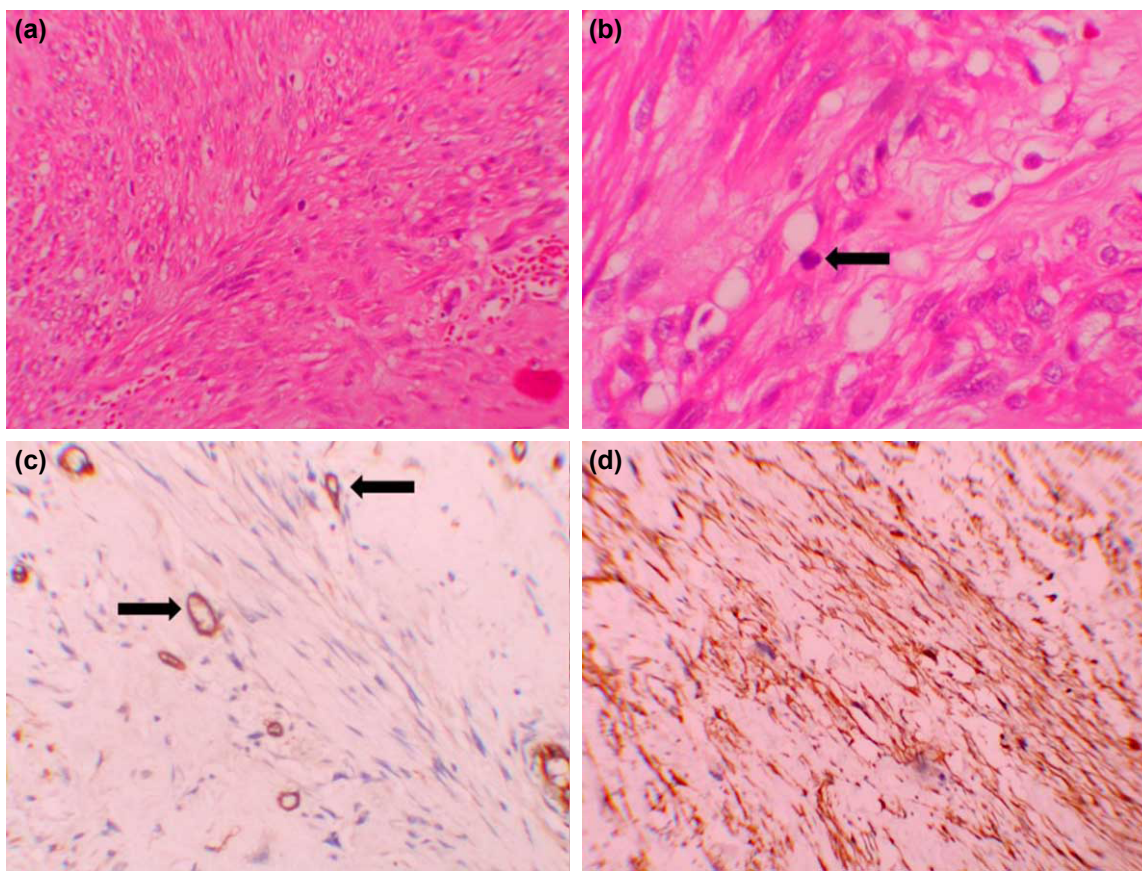

Figure 2. (a) Spindle-cell tumor proliferation with smooth bundles, **(b)** mitotic figures observed in some goblet cells, **(c)** no CD34 staining outside the vessels, **(d)** desmin positivity.

The pathological examination of the specimen determined paratesticular leiomyosarcoma that had originated from the spermatic cord. The radical orchiectomy material macroscopically comprised a specimen that was 8x6x6 cm in size with 11x4 cm of spermatic cord on it. In the specimen section, a regularly structured testis with an intact capsule, which was 6x5 cm in size, was detected. In the spermatic cord, a tumoral mass with irregular borders, which was approximately 2 cm from the surgical margin and 6x7 cm in size, was observed. Within the serial sections that were obtained, a tumoral proliferation of fusiform mesenchymal cells was determined. Cellular atypia, atypical mitotic figures, and increased focal cellularity were observed in the tumoral proliferation. In addition, during the immunohistochemical evaluation, vimentin/SMA/HHF-35 was found to be strongly and diffusely positive, CD34 was positive in vascular structures, CD 117 was weakly positive, and Ki-67 proliferation was low. When morphological and immunohistochemical features were evaluated together, a diagnosis of leiomyosarcoma was made (Fig. 2a-d).

No residual mass, lymph nodes, or distant metastasis were found in radiological evaluations aimed at detecting metastasis after the pathological diagnosis. The case was discussed at the surgical oncology meeting and a decision was made to pursue conservative follow-up. At the 7-month postoperative follow-up, no local recurrence or systemic disease was observed.

DISCUSSION

Paratesticular leiomyosarcoma is a malignancy that arises from the spermatic cord, scrotum, or epididymis, and constitutes approximately 10% of all sarcomas.[3] Sarcomas account for 90% of the paratesticular tumors with malignant characteristics and are mostly of mesenchymal origin.[1,4] Spermatic cord leiomyosarcomas differ from scrotal leiomyomas, which are encountered more frequently in the 6th and 7th decades of life, as in our case.[5] Patients mainly present to polyclinics with a mass that may be painful or painless.[6] Our patient presented at the polyclinic with complaints of groin pain, which had been present for 3 months and a swelling that he had noticed 3 days previously. In such cases, tumor markers and imaging tests should be ordered in addition to performing a physical examination. Since a definitive diagnosis can be made with pathological evaluation, a differential diagnosis from lipomas of the cord and inguinal hernia should be made carefully.[2] In a study conducted by Cardenosa et al.,[7] it was suggested that a definitive preoperative diagnosis is quite hard to reach. In our case, although the hernia had been detected prior to surgery, a conclusive diagnosis was made with the pathological evaluation of the mass encountered in the spermatic cord.

Sarcomas are usually spotted on US as solid and extensive, hyperechoic, extratesticular masses. However, the sonographic appearance of these tumors is variable and not specific. CT and/or magnetic resonance imaging is more specific in terms of diagnosis.[8] Even so, with the evaluation of a CT scan in our case, a clear diagnosis regarding the content of the mass could not be made and US only displayed the hernia sac and the position of the bowel loops. All of these findings show that the role of preoperative methods is limited when reaching a definitive diagnosis, and that benign leiomyoma; fibrous mesothelioma; various benign fibrous tumors; pseudotumors, including fibromatous, inguinoscrotal hernia; testicular malignancy; and other rare tumors are also possibilities that must be kept in mind in the differential diagnosis.[9]

In a study of 24 cases with primary paratesticular leiomyosarcoma, the tumor location was reported as follows: 11 in the testicular tunica, 10 in the spermatic cord, 1 in the scrotal subcutis, 1 in the dartos muscle, and 1 in the epididymis.[9] It was also reported in another study that the spermatic cord was the most frequent location, at a rate of 90%.[4] In our case, the tumor location was the spermatic cord.

Smooth muscle actin, muscle-specific actin, and desmin expression have been observed in most of leiomyosarcomas in the pathological evaluation, while in some cases, CD117, myogenin, Ki-67, S-100 protein, and cytokeratin expression have been reported in immunohistochemical staining as well.[4] Smooth muscle vimentin, CD34, CD117 and Ki-67 were found to be positive in our case.

Since paratesticular sarcomas are seen rarely, there is no clinical consensus regarding the treatment. Despite this fact, radical orchiectomy and high ligation of the spermatic cord is the approach most often adopted. The tumor's tendency to expand to the surrounding tissues is a factor that complicates the complete excision of the tumor.[4] In our case, with radical orchiectomy, the tumor was excised completely with a 2-cm surgical margin.

Post orchiectomy, the local recurrence rate in the groin and the scrotal area has been reported to be 27%.[10,11] Local adjuvant radiotherapy and/or surgical intervention after orchiectomy appears to reduce the risk of local recurrence.[4]

There is still no clear evidence with regard to adjuvant chemotherapy in the treatment of paratesticular leiomyosarcoma. Nonetheless, doxorubicin-based adjuvant chemotherapy has been shown to be beneficial for local and distant metastases.[12]

In conclusion, although paratesticular leiomyosarcomas are seen quite rarely in surgical practice, it is a differential diagnostic possibility that must be kept in mind, particularly in male patients in the 6th and the 7th decades, when a mass is detected in the groin area and strangulated inguinal hernia is observed.

Conflict of interest: None declared.

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OLGU SUNUMU - *ÖZET*

Strangüle inguinal herniye eşlik eden paratestiküler leiomiyosarkom

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Spermatik kord kaynaklı sarkomlar yumuşak doku sarkomlarının %2.1 ve genitoüriner sistemde ise %1–2 oranlarında görülen nadir malignitedir. Yetmiş dört yaşında erkek hasta son üç gündür süren kasık ağrısı ve şişlik şikâyeti ile acil polikliniğimize başvurdu. Fizik muayenede sağ inguinal bölgede yaklaşık 5x5 cm boyutlarında sert redükte edilemeyen kitle ve sağ inguinal herni tespit edildi. Bilgisayarlı tomografide sağ inguinal bölgede büyük bir herni kesesi ve içerisinde 77x55 mm ölçülerinde kitle saptanan hastaya radikal orşiektomi ve herni onarımı yapıldı. Kitlenin patolojik değerlendirmesinde paratestiküler leiomyosarkom tanısı konuldu. Ameliyat sonrası yedinci ayda lokal nüks veya uzak metastaz izlenmedi. Sonuç olarak paratestiküler leiomyosarkomlar nadir görülmelerine rağmen özellikle inguinal kitle ile başvuran ileri yaştaki hasta grubunda akılda tutulması gereken bir durumdur.

Anahtar sözcükler: Kasık fıtığı; leiomyosarkom; paratestiküler tümör.

Ulus Travma Acil Cerrahi Derg 2019;25(3):307-310 doi: 10.5505/tjtes.2018.68709