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LETTER TO THE EDITOR

A rare case: paratesticular leiomyosarcoma

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Dear Editor,

We would like to present a rare case of primary paratesticular leiomyosarcoma. Primary leiomyosarcoma of testis is a rare entity with few cases reported in literature. Most cases reported in the literature indicate that this may be an indolent tumor with a potential for cure if treated early. A 53-year-old man presented with a right scrotal swelling. A computarized tomography (CT) scan showed a heterogeneous enhancing 12×9 cm right scrotal mass. Radical orchiectomy with high ligation of the spermatic cord was performed. The mass was characterized as a grade 2 paratesticular leiomyosarcoma. Intensive follow-up including repeated CT scans was performed. No metastasis was found.

Sarcomas of the genitourinary tract account for <5% of all sarcomas and <2% of all urological tumors. Approximately 100 paratesticular leiomyosarcomas have been reported in the literature. Leiomyosarcoma is a malignant soft tissue tumor that can arise from any tissue containing smooth muscle. However, leiomyosarcoma arising in paratesticular tissue is extremely rare and approximately only 10% of all paratesticular sarcomas are leiomyosarcomas.[1] There is a lack of data on the natural history, histological criteria for diagnosis and recommendations for treatment because of rarity of this disease. The primary treatment for this tumor is radical orchiectomy with high cord ligation; however, the optimal local and systemic treatment remains controversial. A 53-year-old male patient with a 3-year history of a huge right scrotal mass, which had shown rapid growth for more than 3 months, was first diagnosed as orchitis and referred to emergency room in our hospital. Results of the physical examination revealed a right paratesticular mass measuring approximately 12×9 cm in diameter; the mass was firm and non-tender. The serum lactate dehydrogenase level was 280 IU l^{-1}, the α-fetoprotein level was 4.1 ng ml^{-1}, and the β-human chorionic gonadotropin level was 1.2 mIU ml^{-1}, which were all within the normal ranges. Findings on CT scan showed a huge heterogeneous enhancing right scrotal mass (12×9 cm). Right radical orchiectomy was performed and the macroscopic pathological features showed a grossly intact testis with a well-defined, huge, whitish solid mass adjacent to the testis. The tumor was 5.5 cm in diameter. Necrosis rate of approximately 30%–40% and foci of squamous metaplasia and atypical mitosis in the tumor is remarkable. Tunica albuginea and epididymis were invaded with tumor spread. Spermatic cord surgical margin was negative. Immunohistochemical staining showed a positive result for smooth muscle actin and desmin; however, immunostaining for CD117, S-100 protein was negative. The mass was characterized as a leiomyosarcoma, grade 2 (according to National Federation of French Cancer Centers and National Cancer Institute system). The patient did not receive any adjuvant therapy. Four months after the operation, there has been no recurrence. Leiomyosarcoma is a soft tissue tumor arising from smooth muscle cells of a mesenchymal origin. Paratesticular leiomyosarcoma originates from the spermatic cord, the scrotum, or the epididymis. The most common type is the spermatic cord type, which arises from undifferentiated mesenchymal cells of the cremasteric muscle and the vas deferens. The epididymal and scrotal types are less frequent and they originate from the smooth muscle surrounding the basement membrane of the epididymal canal and dartos layer, respectively.[2] Most paratesticular leiomyosarcomas present as painless, slow-growing scrotal tumors in men of middle or older age. Occurrence of this disease is rare; therefore, the mode of spread is important. The most common means of spread is lymphatic, followed by hematogenous, and, last, by local extension. The route of lymphatic dissemination may involve the external iliac, hypogastric, common iliac and para-aortic nodes. The lung is the primary site of blood-borne metastases.[3] Definitive diagnosis of leiomyosarcoma requires histologic examination of a resected specimen for features of smooth muscle differentiation and malignancy. On immunohistochemical staining, expression of smooth muscle actin, muscle-specific actin and desmin is observed in most leiomyosarcomas, while expression of CD117, myogenin, Ki-67, S-100 protein and cytokeratin has also been reported in some cases.[4] The present case showed a positive result for smooth muscle actin and desmin. Paratesticular leiomyosarcoma presents as a discrete nodular mass, which is completely separate from the testicle.[5] There is relatively little evidence-based consensus on the management of paratesticular leiomyosarcoma. Radical orchiectomy is the accepted primary treatment,[6] but there remains the problem of local recurrence. Complete surgical excision can achieve tumor free margins, but wide resection can be difficult to obtain in the paratesticular region. With local excision and surveillance, the survival rate has been reported to be around 50%–80% and there is microscopic residual disease in 27% of cases that underwent repeat excision.[7] Other researchers have reported that one-third of patients were harboring occult local residual disease following local excision or orchiectomy.[8] Given these reported rates of recurrence, there is a clear need for long-term follow-up. Regular chest X-rays and abdominopelvic CT scans are recommended. Leiomyosarcoma is an important differential diagnosis for paratesticular masses in the male. The primary management is radical inguinal orchiectomy with high ligation of the spermatic cord and subsequent long-term follow-up.

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AUTHOR CONTRIBUTIONS

GU carried out the pathologic and immunohistochemical studies. OC and GU made analysis and interpretation of data. OC gave final approval version to be published.

COMPETING FINANCIAL INTERESTS

Both authors declare that there are no competing financial interests.

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