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Case Report

Leiomyosarcoma of the Epididymis: Case Report of Uncommon Location and Review of the Literature

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Submitted: 28 August 2018

Accepted: 27 October 2018

Published: 29 October 2018

ISSN: 2373-9819

OPEN ACCESS

Keywords

- Cancer
- Epididymis
- Leiomyosarcoma
- Testis
- Immunohistochemistry

Abstract

Leiomyosarcoma of the testis is extremely rare. We present the case of a 42-year-old male with a firm right para-testicular mass who underwent a radical wide margin orchiectomy. Histological examination revealed a high-grade leiomyosarcoma.

ABBREVIATIONS

SMM: Smooth Muscle Myosin

INTRODUCTION

Leiomyosarcoma is a soft-tissue malignant smooth muscle cell neoplasm of mesenchymal origin [1,2]. It accounts for 5%–10% of soft tissue sarcomas that originate from the spermatic cord; the scrotum or the epididymis. The less frequent epididymal form originates from the smooth muscle surrounding the basement membrane of the epididymal tubule and/or vasculature [3].

CASE PRESENTATION

A 42-year-old male presented with a right testicular mass. Scrotal ultrasonography showed a solid; heterogeneous extra-testicular mass arising from the head of the right epididymis. A radical wide margin right orchiectomy was performed that included the testicle; epididymis and spermatic cord (Figure 1 A).The outer surface was boss elated with areas of congestion. Microscopically; the mass measured 3.8 cm and showed a markedly malignant mitotically active high grade spindle cell neoplasm with fascicular architecture (Figure-1 B-C). Mitotic figures were numerous measured up to >20 mitotic figures per 10 high power field [4]. Prominent cellular atypia was present; with tumor-type giant cells visible (Figure-1 D). Rare foci of necrosis were noted. The French Federation of Cancer Centers Sarcoma Group grading system score (Table 1) was 7 out of 9 (sum of 3 for degree of differentiation; 1 for minimal necrosis; and 3 for degree of mitotic activity) which corresponds to an overall Grade 3 out of 3 (Figure-1 D). Immunohistochemistry studies were reported as follows; positive for Smooth Muscle Myosin (SMM); desmin; and negative for s100.

High proliferative activity with 20% nuclear staining with Ki-67 was also present. The combined histologic and immunochemical findings were diagnostic of a primary high grade leiomyosarcoma of the epididymis. All surgical margins were free of residual leiomyosarcoma at least greater than 1cm away. A smaller lipoma of the cord measuring 3.2cm was also present.

The patient was followed with ongoing chest x-rays every 6 months and CT imaging of the abdomen and pelvis every 12 months for 3 years; with no evidence of recurrent disease. The patient was eventually lost for follow up.

DISCUSSION

It is estimated that in 2016 approximately 6; 890 new cases of soft-tissue sarcoma were identified in males in the United States. Sarcomas constitute less than 1% of all cancer cases annually [1]. From that 1%; those of the genitourinary tract account for less than 5% and only account for 2% of urological malignancies [1]. Most masses encountered within the scrotal sac are neoplastic within the testis; but a subset (2-3%) of these tumors is extra-testicular and usually arises from para-testicular tissue [5]. Leiomyosarcoma is the most commonly reported histologic type of para-testicular sarcoma with the peak incidence reported to occur in the sixth and seventh decades of life [6]. Paratesticular leiomyosarcoma is am extremely rare neoplasia; it has been reported in less than 100 cases so far [7]. According to its prevalence; this neoplasm is originated from testicular tunica (48%); spermatic cord (48%); epididymis (2%) and dartos muscle [8]. Given these statistics; leiomyosarcoma of the epididymis in a 42-year-old male is highly unusual [9].

Leiomyosarcoma is a soft-tissue malignant neoplasm of

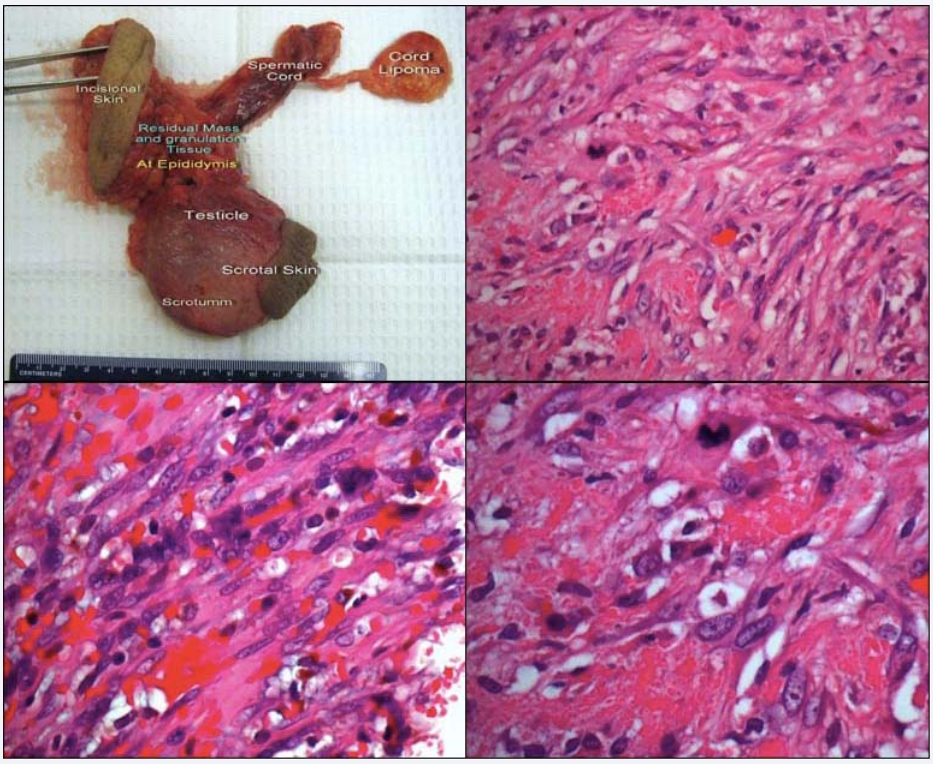

Figure 1 Pathological analysis of the mass grossly and microscopic examination
A: Gross examination of labelled specimen post wide excision including; incisional skin, residual mass and granulation tissue at epididymis, testicle, scrotum, scrotal skin, spermatic cord, and cord lipoma
B: Biopsy from the epididymis mass stained with H&E showing bundles atypical malignant spindle cells; X40
C: High power biopsy stained with H&E showing fascicular architecture, nuclear atypia, polychromasia, and presence of mitotic spindles; X60
D: Wide Excision; histological image of the epididymal leiomyosarcoma showing atypical spindle cells with visible tumor-type giant cells, prominent cellular atypia and abnormal mitosis; X100

smooth muscle cells of mesenchymal origin [2]. Clinically leiomyosarcoma presents as a painless; firm; para-testicular intra-scrotal mass; that is easily mobile within the scrotum and sometimes associated with a small hydrocele [10] (Figures-1 A). There are no reported predisposing factors leading to epididymal leiomyosarcoma [11].

Epididymal leiomyosarcomas metastasize via lymphatic drainage to the retroperitoneal nodes and hematogenously to the lungs [10]. Like other sarcomas; it tends to invade local tissues such as the scrotum; inguinal canal or pelvis via the vas deferens [3]. Definitive diagnosis of leiomyosarcoma requires histologic examination of a resected specimen [9]. The classic histologic features present pleomorphic spindle cells with fascicular architecture at low power (Figure-1 D). At high power; eosinophilic cytoplasm is present containing delicate longitudinal fibrils and blunt-ended nuclei8. Smooth muscle differentiation is identified by an immunohistochemical profile that stains positively for smooth muscle actin; muscle-specific actin and desmin [12]. Data from several systematic studies suggests that desmin is the most sensitive and specific marker for muscle differentiation [13].

High mitotic activity is an important criterion for differentiating leiomyosarcoma from leiomyoma. Necrosis may be seen in high-grade tumors and the threshold number of mitotic figures per high power field that characterizes malignant growth may vary depending on the location from which the tumor arises (Figure-1 B,C). While low-grade tumors tend to be slow growing; high-grade tumors are aggressive and associate with poor clinical outcome [9]. Once the diagnosis of leiomyosarcoma has been established by surgery; clinical staging is necessary [3]. The natural course of leiomyosarcoma depends on site; size; grade and evidence of nodal or distant metastasis [4]. Like any soft tissue sarcoma; the influence of prognostic factors changes over the natural history and the time to recurrence exerts significant influence over complete resection rates [14]. Because of the rarity of epidydimal leiomyosarcoma; the true biological behavior and role of adjuvant chemotherapy and radiotherapy are not well defined and should be treated according to leiomyosarcoma arising out of other common sites [15]. Leiomyosarcoma of the epididymis is an important differential diagnosis for adult men presenting with para-testicular masses. The clinical presentation does not seem to differ from that of other testicular malignancies; however; Immunohistological studies should be included to rule out for leiomyosarcoma on small biopsies. Due to the small number of reported cases the clinical and biological behavior of these tumors are very hard to predict; however; orchiectomy with high ligation of the spermatic cord followed by surveillance appears to be the most effective treatment [16]. To our knowledge; only 22 cases

Table 1: French federation of cancer center sarcoma group grading system.	
Tumor differentiation	
Score 1: Sarcomas that closely resemble normal adult mesenchymal tissues. Score 2: Sarcomas for which histologic typing is certain. Score 3: Embryonal and undifferentiated sarcomas, synovial sarcoma, and sarcomas of uncertain differentiation.	
Mitotic Count	
Score 1: 0-9 mitoses/10 hpf Score 2: 10-19 mitoses/10 hpf Score 3: ≥ 20 mitoses/10 hpf	
Tumor necrosis	
Score 0: no necrosis Score 1: < 50% tumor necrosis Score 3: ≥ 50% tumor necrosis	
Histologic grade (tumor differentiation + mitotic count + tumor necrosis)	
Grade 1 (low grade) Grade 2 (intermediate grade) Grade 3 (high-grade)	Total score: 2 or 3 Total score: 4 or 5 Total score: 6,7 or 8
Abbreviations: HPF: High-Power Field	

Table 2: Literature Review.

Authors year of publication	Year of Publication	AGE of Presentation	Side/Size	Complaints	Metastasis at Presentation	Surgical Treatment	ADJUVANT-TREATMENT	Follow Up	Status At last Follow Up
Kwae et al and bergman	1949	50	Left 13 cm	Swelling	None	Excisionof tumor +ilioinguinal dissection	Not Mentioned	Not Disease mentioned free2 mentioned free2	
Sherwin And Bergman	1952	66	Right 3.5 cm	Swelling	None	Orchiectomy + excision of inguinal lymphnodes	None	14 Months	Metastasis To right humerus
Abelland Holtz	1969	66,67		None	None	None	None	11 Months	Died at11 monthsnorecurrence
Yadav et al	1969	28	Right 6 cm	Swelling	None	Orchiectomy	Not Reported	Not Reported	Not Reported
Rushwor Th et al	1971	53	Left 3.5 cm	Swelling	None	High inguinalorchiectomy	None	5 Months	Disease Free at 5 months
Davides Kc et al	1975	58	Right 5 cm	Swelling	None	High inguinalorchiectomy	None	2 Years	Disease Free at 2 Years
Deluise Et Al	1976	74	None	None	None	None	None	3 Years	Died at 3 Years no recurrence
Farrell And Donnelly	1980	6,55	Left 6 cm, Right 8 cm	Swelling	None	High inguinalorchiectomy	None	21 years, 6 months	Disease Free at 6 months
Helm And Al-Tikriti	1986	67	Right 8 cm	Swelling	None	Orchiectomy ligation atexternalring	Not Reported	6 months	Disease Free at 6 months
Demetrio U D et al	1994	67	Left	Swelling	None	Radical orchiectomy	None	2 Years	Disease Free at 2 Years
Planzb et al	1998	85	Right 7 cm	Swelling	None	orchiectomy orchiectomy	None	6 years	Recurrence At root of Penis at 6years
Fisher et al	2001	52	Not Reported	Not Reported	Not Reported	Not Reported	Not Reported	Not reported	Not reported
Bressenota et al	2009	78	None	Lump	None	High Orchiectomy	None	2 Years	Dead at2years dueto nonrelatedcause
Mechri et al	2009	78	Left 6-7 cm	Lump	None	Inguinal Noneorchiectomy	None	3 Years	Diseasefreeat3years
Yuen et al	2011	58,75	Left 2.5 cm, Right 3.8 cm	Lump	None	Radical noneorchiectomy	None	Every 6 months for 2.5 years,	Diseasefreeafter2.5years,diseasefreeafter5months

Tazi et al	2012	70	Left 25 cm	Mass	Inguinal-node	Mass Chemothera-pyexcision	NOne	1 Month	Died at1month
Muduly et al	2012	35	Left 3 cm	Lump	None	High Noneinguinal-orchiectomy	None	2 Years	Diseasefreeaf-ter2years
Humphrey	2013	57	3.6 cm	Lump	None	Radical Noneor-chiectomy	NOne	Not Re-ported	Not reported
Schmidt/ Rani-wsky	None	12	3.8 cm	Lump	None	Radical orchiec-tomy	None	3 Years	Lost to follow up

of Leiomyosarcoma of the Epididymis were reported in the English literature (Table 2). Among the 22 patients treated for leiomyosarcoma of the epididymis; only 1 had reported adjuvant chemotherapy due to his advanced disease. All patients received an orchiectomy and only 1 had recurrence at the root of the penis with most being disease frees at 6 months. Based on this data; orchiectomy in the setting of local leiomyosarcoma of the epididymis is associated with a low rate of recurrence at 6 months confirming its use as the primary modality of treatment.

It is our hope that this report raises the awareness of clinicians and pathologists to include Leiomyosarcoma of the Epididymis in the differential diagnosis of a presentation with a para-testicular mass and that continued investigation drives further development of efficacious and safe treatments for improving patient outcomes.

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Cite this article

Schmidt M, Raniwsky A, Sebald A, Balakrishnan E, Aziz M (2018) Leiomyosarcoma of the Epididymis: Case Report of Uncommon Location and Review of the Literature. JSM Clin Case Rep 6(4): 1159.