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Case report

Testicular leiomyosarcoma: A case report and literature review

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ABSTRACT

Introduction and importance: Leiomyosarcoma is a malignant mesenchymal tumor derived from the smooth muscle, it represents approximately 7% of all soft tissue sarcomas.
Male genitourinary leiomyosarcomas are rare (Abdullazade et al., 2013 [1]).
Primary testicular leiomyosarcoma is an exceptional entity with only 30 cases reported in the literature (Giridhar et al., 2011). Due to its rarity, additional studies are necessary to better define the optimal therapeutic management.
Case presentation: We report a case of a 42-years-old male diagnosed in the urology department A of the University Hospital Ibn Sina in Rabat who complains of testicular swelling.
The anatomopathological examination and immunohistochemical study revealed a leiomyosarcoma therefore, a radical inguinal orchiectomy with a primary ligation of the spermatic cord was performed for diagnostic and therapeutic purposes.
The assessment of extension did not reveal any lymph node location or secondary appearance thus the decision of the multidisciplinary meeting opted for regular cancer check-ups without adjuvant treatment.
Discussion: The actual etiology of testicular leiomyosarcoma is still unknown added to its clinical presentation and radiological results that are non-specific.
Conclusion: Leiomyosarcoma of the testis is a very rare tumor and its clinical and radiological presentation remains similar to other testicular malignancies.

1. Introduction

Leiomyosarcoma of the testis is a rare tumor [3], a review of the literature allowed us to identify 30 cases worldwide, the first one was reported by Yachia.D and Auslaender in 1989 [4]. The latest was reported by Siraj and al in 2018 [5].

Its clinical aspects doesn't differ from other testicular malignancies, generally a painless testicular mass is the common clinical manifestation [3,2]. Scrotal Ultrasound usually shows a well-delineated hypoechoic mass with or without calcifications. Computed tomography is necessary to detect metastases.

The diagnosis is confirmed after histological and immunohistochemical studies [3].

Radical inguinal orchiectomy is the treatment of choice, that should be followed by monitoring [6,7].

2. Case presentation

This work has been drawn up according to the SCARE (Surgical Case Report) criteria [20].We present the case of a 42-years-old male patient, with no significant medical history, consulting for a progressive testicular swelling.

The patient does not report any notion of drug intake, nor similar cases in the family.

Physical examination showed a firm, painless 3 cm node of the right testis, with a palpable epididymo-testicular groove and no inguinal lymph nodes.

Tumor markers tests were normal except for LDH high levels. The testicular Doppler ultrasound revealed a large testis with a heterogeneous lobulated tissular node, highly vascularized, sharply circumscribed measuring 30 mm × 19.7 mm (Fig. 1). (See Tables 1 and 2.)

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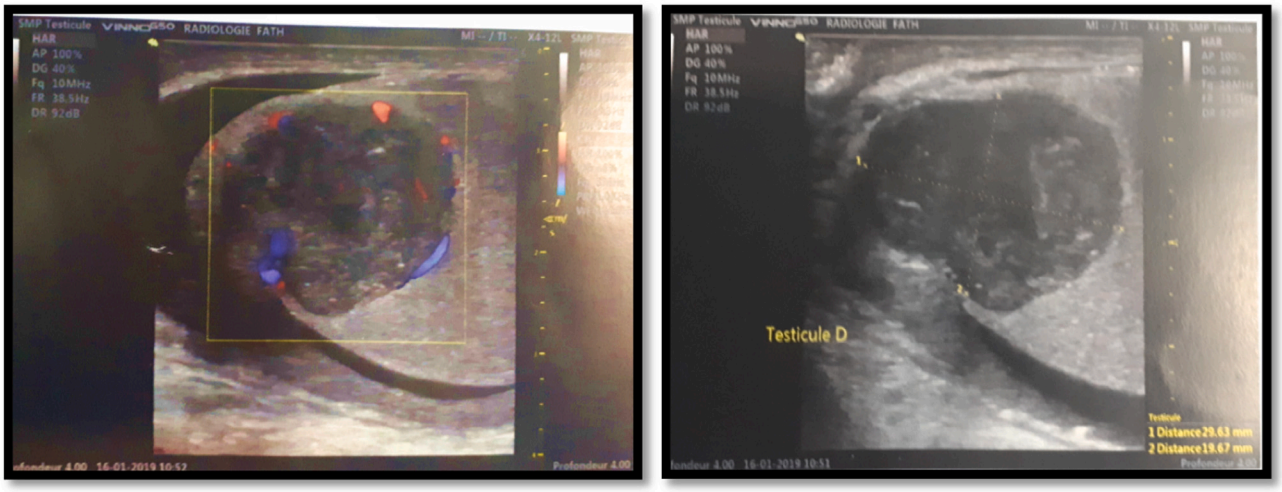

Fig. 1. Testicular doppler ultrasound showing a heterogenous tissular node.

Table 1
A worldwide literature review for clinical, biological and anatomopathological results of intratesticular Leiomyosarcomas.

Author	Year of publica-tion	Country	Age	Affected side	Tumor markers	Tumor size (cm)	Tumor stage
Yachia	1989	Israël	55	Right	Normal	4,5	I
Pellice	1994	France	37	Left	Normal	–	I
Weshecka	1996	USA	47	Right	Normal	4,8	I
			40	Right	Normal	4	I
Frohner	1999	Germany	32	Right	Normal	1,7	I
Hachi	2002	Morocco	70	Left	Normal	Unknown	I
Ali	2002	Kuwait	65	Right	Normal	12	I
Sattary	2003	england	27	Left	Normal	4,5	I
Singh	2004	India	26	Left	Normal	2,6	I
Wakhlu	2004	India	8 months	Left	Normal	23	I
Takizawa	2005	Japan	76	Left	Normal	7,4	I
Canales	2005	USA	30	Right	Unknown	4	I
Borges	2007	Portugal	19	Left	Normal	7	I
Fadl-Elmula	2007	Sudan	20	Left	Normal	20	II
Raspollini	2009	Italy	77	Left	Normal	4	I
Kumar	2009	India	65	Right	Normal	8,5	II
Yoshimine	2009	Japan	73	Left	High BHCG levels	20	III
Labanaris	2010	Germany	73	Right	Normal	3,5	I
Tobe	2010	Japan	71	Right	Normal	Unknown	I
Bakhshi	2011	India	60	Right	High LDH levels	10	I
Mohd	2011	India	45	Right	Normal	3	II
Gridhar	2011	India	55	Left	Normal	7	II
Komeya	2012	India	70	Left	Normal	4,5	I
Abdullazad	2013	Tukey	49	Left	Normal	3,5	I
Bostanci	2013	USA	68	Left	Normal	10	I
Damle	2013	India	68	Right	Normal	19	III
Hmida	2014	Tunisia	78	Right	High LDH levels	9	II
Rana	2017	India	50	Left	Normal	8,5	I
Rajagopal	2017	India	70	Right	Normal	4	II
Siraj	2018	Turkey	27	Left	Normal	10	II
Our case	2019	Morroco	42	Right	Normal	2,7	I

A radical right inguinal orchiectomy under spinal anesthesia was performed by our urology team; the postoperative follow up showed no complications.

Histological study confirmed the diagnosis of FNCLLC grade I right intratesticular leimyosarcoma by defining microscopically a proliferation of spindle-shaped cells in the eosinophilic cytoplasm having elongated nucleus with clumped chromatin, weakly nucleolated, with absence of tumor necrosis (Fig. 2).

Tumor cells expressed antibodies: anti-smooth muscle (AML), and caldesmone, while PS100 and CD34 were negative. The thoraco-abdominal-pelvic scan did not show any suspicious lymph nodes or distant metastasis.

The case was discussed within a multidisciplinary concertation meeting and the decision was based on a regular oncological follow up without adjuvant treatment. There were no recurrences or metastasis within a one year follow-up.

3. Discussion

Scrotal Leiomyosarcomas are divided into 2 categories: paratesticular and intratesticular [8].

Paratesticular leiomyosarcomas are relatively common and around 100 cases have been reported [9].

Primary leiomyosarcoma of the testis is a rare entity with few cases reported in the literature [2].

The actual etiology of testicular leiomyosarcomas is already unknown, but hormonal stimulation has been suggested to play a role in the carcinogenesis of leiomyosarcoma [10].

Table 2
Comparison of the surveillance and adjuvant treatment results for different case reported in the literature.

Author	Year	Duration of follow up	Results	Adjuvant treatment
Yachia	1989	24 months	No recurrence - No metastasis	None
Pellice	1994	24 months	No recurrence- No metastasis	None
Weshecka	1996	6 months	No recurrence- No metastasis	None
		6 months	No recurrence - No metastasis	None
Frohner	1999	6 yearsand a Half	No recurrence - No metastasis	DRPGL
Hachi	2002	14 mois	Metastasis: lung Death	None
Ali	2002	12 months	No recurrence – No metastasis	None
Sattary	2003	30 months	No recurrence - No metastasis	None
Singh	2004	Unknown	No recurrence – No metastasis	None
Wakhlu	2004	12 months	No recurrence - No metastasis	Chemotherapy
Takizawa	2005	12 months	No recurrence – No metastasis	None
Canales	2005	6 months	No recurrence - No metastasis	None
Borges	2007	16 months	Retroperitoneal metastasis	Chemotherapy + radiotherapy
Fadl-Elmula	2007	11 months	Cerebral metastasis + local recurrence + para aortic lymph nodes	Chemotherapy + radiotherapy + suregery for local recurrence
Raspollini	2009	12 months	No recurrence – No metastasis	None
Kumar	2009	6 months	No recurrence - No metastasis	None
Yoshimine	2009	9 months	Multiple metastasis: lungs- lymphatic- spleen- muscles- spinal	Chemotherapy (CYVADIC)
Labanaris	2010	28 months	No recurrence - No metastasis	None
Tobe	2010	7 months	No recurrence- No metastasis	None
Bakhshi	2011	12 months	No recurrence - No metastasis	Radiotherapy
Mohd	2011	Unknown	Ganglionar metastasis	Chemotherapy
Gridhar	2011	8 months	Bone metastasis and diffuse soft tissue	Chemotherapy
Komeya	2012	34 months	Retroperitoneal metastasis	DRPGL
Abdullazade	2013	24 months	No recurrence – No metastasis	None
Bostanci	2013	12 months	No recurrence – No metastasis	None
Damle	2013	6 months	No recurrence – No metastasis	None
Hmida	2014	24 months	No recurrence – No metastasis	None
Rana	2017	4 months	No recurrence – No metastasis	None
Rajagopal	2017	Unknown	Local recurrence No metastasis	Radiotherapy + wide local excision
Siraj	2018	24 months	No recurrence - No metastasis	None
Our case	2019	12 months	No recurrence – No metastasis	None

After literature reviews and case studies, it has been associated to radiotherapy, long-term use of anabolic steroids, and chronic inflammation [10,11,12].

The clinical presentation and the radiological results of this tumor are non-specific thus it seems not different from other malignant testicular tumors [13,3].

Scrotal ultrasound is currently used as a reference for morphological exploration of bursa [14]. Some authors find that MRI can add an interesting contribution to the diagnosis of testicular cancers [15,16].

Tumor markers are generally within the normal range of leiomyosarcomas [17].

The exact diagnosis is based on histological studies [18].

Histologically, leiomyosarcoma presents as spindle-shaped smooth muscle cells with typical nucleus. In the presence of significant nuclear atypia, a mitotic index $\geq$ 10/10 high magnification fields and coagulant necrosis with nuclear debris are criteria for the Diagnosis of uterine spindle cell leiomyosarcoma, the same diagnostic criteria can be also used for the diagnosis of intratesticular leiomyosarcoma, with a subsequent immunohistochemical profile to support the diagnosis of a primary intratesticular smooth muscle tumor [3,7].

The extension assessment is based on Thoraco-abdominal-pelvic CT, to look for suspicious lymph node or distant lesions, especially in the lungs, that may be a sign of a poor prognosis.

Radical orchiectomy is the main treatment [2]. Chemotherapy is advised for advanced stage tumors because radiotherapy is not effective [19].

According to the data in the literature, adjuvant treatment (chemotherapy and radiotherapy) is not justified, hence the interest of reporting and monitoring these rare cases, thus overtime we will be able to clarify the disease process and guide the correct management of patients in the future.

Semi-annual pelvic CT scan [13] and palpation of the scrotum and inguinal region have been considered essential surveillance forms.

4. Conclusion

Leiomyosarcoma of the testis is a very rare tumor and its clinical and radiological presentation remains similar to other testicular malignancies. Inguinal orchiectomy is the standard initial treatment but it is difficult to define optimal management due to the lack of research and recent recommendations.

Informed consent

An informed consent was obtained from the patient for publication of his case, along with appropriate images. No information that would enable his identification has been provided.

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Ethical approval

The study is exempt from ethical approval in our institution.

Author contribution

Hussein Abdallah carried out the data collection, analyzing the data and drafted the manuscript. All Authors have read and approved the manuscript.

Guarantor

Hussein Abdallah.

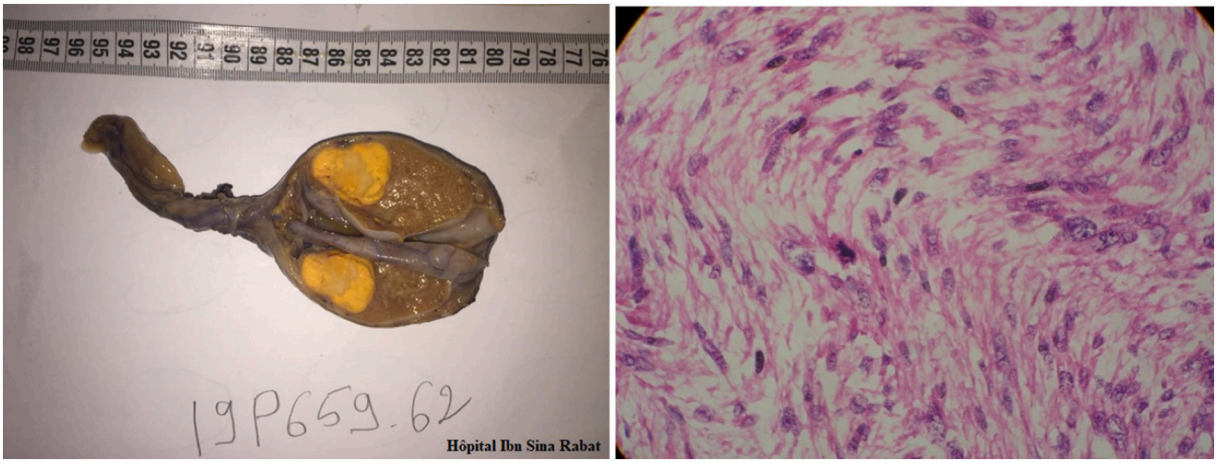

Fig. 2. Macroscopic and microscopic aspect of the right testis.

Declaration of competing interest

The authors declare that they have no conflict of interest.

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