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.)
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 leiomyosarcoma 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: para-testicular 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 1
A worldwide literature review for clinical, biological and anatomopathological results of intratesticular Leiomyosarcomas.

| Author     | Year of publication | Country   | Age  | Affected side | Tumor markers | Tumor size (cm) | Tumor stage |
|------------|---------------------|-----------|------|---------------|---------------|----------------|-------------|
| Yachia     | 1989    | Israel   | 55   | Right        | Normal        | 4,5            | I           |
| Pellice    | 1994    | France   | 37   | Left         | Normal        | –              | I           |
| Wecheeka   | 1996    | USA      | 47   | Right        | Normal        | 4,8            | I           |
| Frohner    | 1999    | Germany  | 52   | Right        | Normal        | 4              | 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           |
| Wakhu      | 2004    | India    | 8 month | 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           |
| Fadi-Elmula| 2007   | Sudan    | 20   | Left         | Normal        | 20             | II          |
| Raspolini  | 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        | Unknown       | 1              | 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           |
| Abdullahad | 2013    | Turkey   | 49   | Left         | Normal        | 3,5            | I           |
| Botraneci  | 2013    | USA      | 68   | Left         | Normal        | 10             | I           |
| Damle      | 2013    | India    | 68   | Right        | Normal        | 19             | III         |
| Himida     | 2014    | Tunisia  | 78   | Right        | High LDH levels | 9     | II         |
| Rana       | 2017    | India    | 50   | Left         | Normal        | 8,5            | I           |
| Rajaogal   | 2017    | India    | 70   | Right        | Normal        | 4              | II          |
| Siraj      | 2018    | Turkey   | 27   | Left         | Normal        | 10             | II          |
| Our case   | 2019    | Morocco  | 42   | Right        | Normal        | 2,7            | I           |

Fig. 1. Testicular doppler ultrasound showing a heterogeneous tissue node.


| 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               |
| Weschecka   | 1996 | 6 months               | No recurrence - No metastasis| None               |
| 6 months    | No recurrence - No metastasis| None               |
| Frohner     | 1996 | 6 years and 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 |
| Fadi-Elmula | 2007 | 11 months              | Cerebral metastasis + local recurrence + para aortic lymph nodes| Chemotherapy + radiotherapy + surgery for local recurrence |
| Raspolini   | 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 (GYVADIC) |
| Labanaris   | 2010 | 28 months              | No recurrence - No metastasis| None               |
| Tobe        | 2010 | 7 months               | No recurrence- No metastasis | None               |
| Bakshhi     | 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 |
| Kongeya     | 2012 | 34 months              | Retroperitoneal metastasis   | DRPGL              |
| Abdallazade | 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               |
| Hamda       | 2014 | 24 months              | No recurrence – No metastasis| None               |
| Rana        | 2017 | 4 months               | No recurrence – No metastasis| Radiotherapy + wide local excision |
| Rajagopal   | 2017 | Unknown                | Local recurrence No metastasis| None               |
| 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 [13,15].

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 [19,20].

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 orchectomy 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.
Declaration of competing interest

The authors declare that they have no conflict of interest.

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