https://doi.org/10.31871/WJIR.8.4.40
World Journal of Innovative Research (WJIR)
ISSN: 2454-8236, Volume-8, Issue-4, April 2020 Pages 13-15

A Rare Case of Paratesticular Leiomyosarcoma

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***Abstract*— Sarcomas of the genitourinary tract are uncommon and represent only 1–2% of all urological malignancies. Paratesticular tumors, which include those arising from testicular tunics, epididymis and spermatic cord are remarkably rare entities. Paratesticular leiomyosarcoma (LMS) is a malignant mesenchymal tumor with smooth muscle differentiation which is difficult to diagnose preoperatively.**

The definitive diagnosis requires a histologic examination of a resected specimen to observe morphological and immunohistochemical differentiation. The standard primary treatment for this tumor is radical orchiectomy with high cord ligation. However, a consensus on the optimal treatment has not yet been reached due to the paucity of cases. Close follow-up is necessary to prevent recurrence and distant metastases.

Here we report a case of a 64 year-old man who presented low grade paratesticular leiomyosarcoma with hepatic metastases.

***Index Terms*— Chemotherapy, Metastases, Orchidectomy, Paratesticular leiomyosarcoma**

I. INTRODUCTION

Sarcomas of the genitourinary tract are uncommon and represent only 1–2% of all urological malignancies [1]. Paratesticular tumors, which include those arising from testicular tunica, epididymis and spermatic cord are less common and their incidence is difficult to estimate [2]. Paratesticular leimyosarcoma is a malignant mesenchymal tumor with smooth muscle differentiation which is extremely rare and most of the available information derives from small series or case reports.

There is a lack of data on the natural history, histological criteria for diagnosis and recommendations for treatment because of rarity of this disease.

We present a case of 64 year-old man who presented low grade paratesticular leiomyosarcoma with hepatic metastases.

II. CASE REPORT

A 64 year-old male patient presented with 3 years history for a right paratesticular mass growing up slowly without any complains of pain, however had recently been increasing in size . A scrotal ultrasound was performed which revealed a mass on the path of the right cord in favor of a tumor lesion confirmed by pelvic MRI. Tumor markers (β-human chorionic gonadotropin, alpha-fetoprotein and lactate dehydrogenase) were all within the normal ranges. Right radical orchidectomy was underwent where a mass measuring 4,5 cm was excised . Histological features were consistent with a diagnosis of paratesticular leiomyosarcoma, grade 1, according to FNCLCC. Immunohistochemistry showed that tumor cells were positive for desmin, smooth muscle actin and h-caldesmon. A subsequent staging CT of the chest, abdomen and pelvis revealed hepatic lesions suggestive of metastases, confirmed by hepatic MRI (Figure 1).

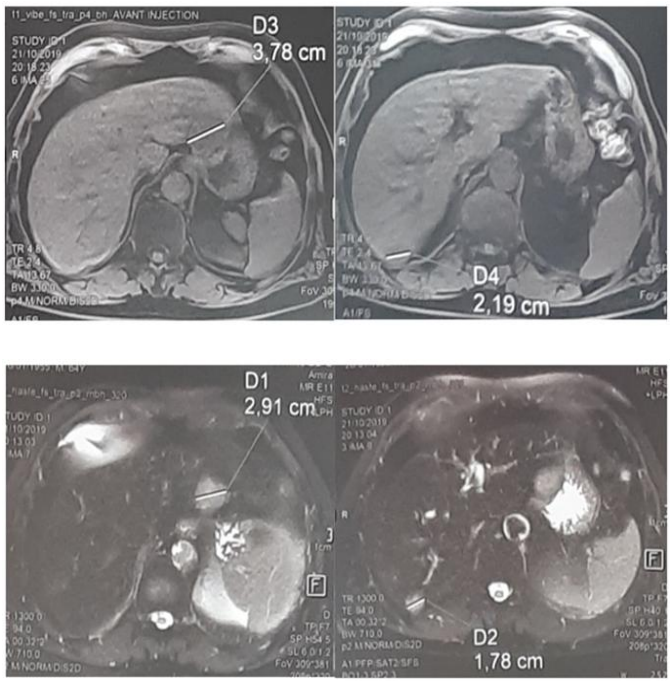

Figure 1: hepatic MRI showing metastases to the liver

A biopsy of these secondary lesions to the liver was carry out and the pathological examination confirmed metastases of leiomyosarcoma, similar to the primary tumor. Thus, six cycles of chemotherapy with adriamycin and dacarbazine were performed on the patient; the disease stabilised and the patient is actually under surveillance.

III. DISCUSSION

Paratesticular leiomyosarcoma is a rare entity which originates from the spermatic cord, the scrotum, or the

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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 [3].

Like other sarcomas, leiomyosarcoma tends to infiltrate local tissues. 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. Haematogenous metastases are primarily pulmonary. The vas deferens can act as a conduit for local spread, which may involve the scrotum, inguinal canal or pelvis [4].

Leiomyosarcoma may be divided anatomically into deep soft-tissue, cutaneous-subcutaneous, and vascular. The American Joint Committee on Cancer (AJCC) classifies spermatic cord leiomyosarcoma as deep tissue [5]. The presence of mitotic activity, percentage of necrosis, and severity of nuclear pleomorphism are all evaluated to grade the disease [6].

Peak incidence is in the sixth and seventh decade [7]. Typical clinical presentation is of a painless, firm, slow-growing, intrascrotal mass with palpation usually revealing the mass to be well defined, lobulated, mobile, and sometimes associated with a small hydrocele [8]. Investigation should begin with ultrasound scanning (USS), which is the primary choice of imaging for any scrotal abnormality with a sensitivity of 95–100% for distinguishing between intra- and extra-testicular lesions [9]. CT and magnetic resonance imaging are useful to delimitate the tumor extension. PET-CT is useful to see the nodal involvement [10].

Definitive diagnosis of leiomyosarcoma requires histologic examination of a resected specimen for features of smooth muscle differentiation and malignancy. Typical histological findings include perpendicularly organised spindle cells with fascicular arrangement at low power and eosinophilic cytoplasm containing longitudinal fibrils and hyperchromatic blunt-ended nuclei at high power [11]. 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 [12].

Due to the rarity of paratesticular LMS, no treatment protocol exists. The standard primary treatment is radical orchidectomy with high ligation of the spermatic cord [13]. Nevertheless, due to anatomical constraints, wide circumferential resection margins are rarely achieved; thus aggressive surgical strategies are advocated involving wide en bloc excisions of all potentially contaminated surrounding soft tissues aiming to obtain negative margin status as well as performing wide inguinal re-resection of soft tissue and scar excision in patients found to have inadequately resected disease [14]. If scrotal skin is involved, hemiscrotectomy is indicated [15]. The role of lymph node dissection is not already proved [16].

As sarcomas of all grades have a tendency to infiltrate local tissues, adequate initial surgical resection can be difficult and regional recurrence is a major problem, with scrotal recurrence rates as high as 25–37% [17]. There is some evidence supporting the use of adjuvant radiotherapy for paratesticular sarcomas to reduce rates of recurrence. [18; 19]. In a series of 21 cases, Catton and colleagues noted a 5-year disease-free survival of 58% with surgery alone and 100% with the addition of adjuvant radiotherapy [20]. A study from Massachusetts confirmed these results in a series of 18 patients with five of nine patients (56%) treated with surgery alone developing locoregional failure, whilst there were no cases of locoregional recurrence amongst the nine patients treated with both surgery and radiation [18].However, adjuvant therapy has not been well established and remains controversial.

There is currently no clear role for adjuvant chemotherapy in the treatment of paratesticular leiomyosarcoma. In 1997 the Sarcoma Meta-analysis Collaboration showed significant relapse free survival rates (median follow-up 9.4 years) with adjuvant doxorubicin in soft-tissue sarcomas (n=1568) [21]. However Woll et al. in the largest Phase III randomized control trial to date failed to show an improvement with chemotherapy (n=351) [22]. Conventional adjuvant systemic chemotherapy has no exact efficacy for spermatic cord leiomyosarcoma. At present, the role of chemotherapy remains controversial and restricted to the presence of metastatic disease [14].

All patients with paratesticular leimyosarcoma require a strong follow-up for detect a recurrence, because recurrences are common. Multivariate analysis revealed that tumor grade, stage, histologic type, and lymph node involvement were independently predictive of prognosis [23].

IV. Conclusion

Leiomyosarcoma should be considered as a differential diagnosis in any elderly male presenting with an intrascrotal mass. The diagnosis and treatment of paratesticular leiomyosarcoma remain challenging with a lack of information due to the rarity of this disease. Careful follow-up of patients is needed to prevent recurrence and distant metastases. Further research to develop evidence is required to determine the optimal management for this disease.

Acknowledgment

We thank our oncologists colleagues at national institute of oncology of Rabat who provided care and support for the patient.

REFERENCES

[1] Moloney J, Drumm J, Fanning DM. A rare case of paratesticular leiomyosarcoma. Clin Pract 2012;2:e29.
[2] Pérez Juan, Licham Milagros, Godoy Mariela, et al. Sarcomas paratesticulares en el paciente adulto. Manejo y evolución de la enfermedad. Actas urológicas españolas. 2009;33(6):639e645.
[3] Dangle P, Basavaraj DR, Bhattarai S, Paul AB, Biyani CS et al. Leiomyosarcoma of the spermatic cord: case report and literature review. Can Urol Assoc J 2007; 1: 55–8.
[4] J. Moloney, J. Drumm, and D. M. Fanning, "A rare case of paratesticular leiomyosarcoma," Clinics and Practice, vol. 2, p. e29, 2012.
[5] Folpe AL, Weiss SW. Paratesticular soft tissue neoplasms. Semin Diagn Pathol 2000;17:307-18.
[6] P. Mohammadi Torbati and H. Zham, "Epithelioid type of paratesticular leiomyosarcoma: a case report and literature review," Journal of Urology, vol. 1, no. 3, pp. 215–217, 2004.
[7] P. Dangle, D. R. Basavaraj, S. Bhattarai, A. B. Paul, and C. S. Biyani, "Leiomyosarcoma of the spermatic cord: case report and literature review," Canadian Urological Association Journal, vol. 1, no. 1, pp. 55–58, 2007.
[8] V. T. Yuen, S. D. Kirby, and Y. C.Woo, "Leiomyosarcoma of the epididymis: 2 cases and review of the literature," Journal of the Canadian Urological Association, vol. 5, no. 6, pp. E121–E124, 2011.
[9] Rodriguez D, Olumi AF. Management of spermatic cord tumors: a rare urologic malignancy. Ther Adv Urol 2012;4: 325–34.
[10] Joana Alfarelos , Gustavo Gomes et al. Paratesticular Leiomyosarcoma: A Case Report and Review of the Literature. Urology Case Reports 11 (2017) 30e32.
[11] H. Hashimoto, Y. Daimaru, M. Tsuneyoshi, and M. Enjoji,"Leiomyosarcoma of the external soft tissues. A clinicopathologic, immunohistochemical, and electron microscopic study," Cancer, vol. 57, no. 10, pp. 2077–2088, 1986.
[12] Fisher C, Goldblum JR, Epstein JI, Montgomery E. Leiomyosarcoma of the paratesticular region: a clinicopathologic study. Am J Surg Pathol 2001; 25: 1143–9.
[13] Mohamad Moussaa, Mohamed Abou Chakra. Leiomyosarcoma of the spermatic cord: A case reportand literature review. International Journal of Surgery Case Reports 57 (2019) 175–178.
[14] J. Coleman, M. F. Brennan, K. Alektiar, and P. Russo, "Adult spermatic cord sarcomas: management and results," Annals of Surgical Oncology, vol. 10, no. 6, pp. 669–675, 2003.
[15] L. B. Weinshank, D. Kellner, and I. Nash, "Paratesticular leiomyosarcoma: case report and review," ConnecticutMedicine, vol. 75, no. 7, pp. 399–403, 2011.
[16] N.H. Kolev, V.R. Dunev et al. Paratesticular leiomyosarcoma: A clinical case report. Urology Case Reports 27 (2019) 100913
[17] Kucuk U, Bayol U, Pala EE, Akman O, Divrik RT. Paratesticular sarcomas: our case series. Turk J Urol 2013;39: 84–9.
[18] M. A. Fagundes, A. L. Zietman, A. F. Althausen, J. J. Coen, and W. U. Shipley, "The management of spermatic cord sarcoma," Cancer, vol. 77, no. 9, pp. 1873–1876, 1996.
[19] H. C. Bauer, T. A. Alveg˚ard, ¨ O. Berlin et al., "The Scandinavian Sarcoma Group Register 1986-2001," Acta Orthopaedica Scandinavica, Supplement, vol. 75, supplement 311, pp. 8–10, 2004.
[20] C. Catton, B. J. Cummings, V. Fornasier, B. O'Sullivan, I. Quirt, and D. Warr, "Adult paratesticular sarcomas: a review of 21 cases," Journal of Urology, vol. 146, no. 2, pp. 342–345, 1991.
[21] Adjuvant chemotherapy for localized resectable soft-tissue sarcoma of adults: meta-analysis of individual data. Sarcoma Meta-analysis Collaboration. Lancet 1997; 350:1647-54.
[22] Woll PJ, Van Glabbeke M, Hohenberger P, et al. Adjuvant chemotherapy (CT) with doxorubicin and ifosfamide in resected soft tissue sarcoma (STS): interim analysis of a randomised phase III trial. J Clin Oncol 2007; 25 Suppl 18:10008.
[23] Rodríguez D, Barrisford GW, Sanchez A, Preston MA, Kreydin EI, Olumi AF. Primary spermatic cord tumors: disease characteristics, prognostic factors, and treatment outcomes. Urol Oncol. 2014 Jan;32(1):52.e19–52.e25