Case Report

# Primary testicular leiomyosarcoma

Mohd Shafi Moona, Dzumbaeva Fatima, Abduldaev Turezbek
Department of Medical Oncology, Kyrgyz State Medical Academy, Bishkek, Kyrgyzstan.

## Abstract

Primary leiomyosarcoma of the testis is a rare condition with only 13 cases reported. We present a case of primary intra-testicular leiomyosarcoma in a 45 year old patient who presented with painless testicular enlargement. Ultrasound revealed a large heterogeneous right testicular solid mass with moderate hydrocele and huge retroperitoneal mass. Serum alpha-fetoprotein (AFP) and beta-human chorionic gonadotrophin (β-HCG) were normal. A right radical orchidectomy with histopathology confirmed primary leiomyosarcoma of testis. A leiomyosarcoma should be one of the differential diagnosis of a sero-negative testicular mass.

**Keywords:** Testis, Intratesticular tumour, Leiomyosarcoma.

## Introduction

Leiomyosarcoma is a soft tissue tumour (STS) arising from smooth muscle cells of mesenchymal origin. Leiomyosarcoma of testis is known to occur following radiotherapy, with use of anabolic corticosteroids and in association with testicular germ cell tumours. However, occurrence of testicular leiomyosarcoma without these predisposition (primary intratesticular leiomyosarcoma) is extremely rare. Although paratesticular leiomyosarcoma are reasonably common, primary testicular leiomyosarcoma is extremely rare. Only thirteen cases have been reported in literature, with metastatic disease only in one case.[1-13]

We report a case of testicular leiomyosarcoma with metastasis to para-aortic lymph nodes.

## Case Report

A 45 year old male was admitted with a right scrotal mass of 5 months duration. He had no significant past medical history. On physical examination there was a 4x8 cm right scrotal mass. The X-ray chest was normal. CT scan revealed huge retroperitoneal lymphadenopathy. Tumour markers including Lactate dehydrogenase (LDH), alpha-fetoprotein (AFP) and beta-human chorionic gonadotrophin (β-HCG) were within normal ranges. An ultrasonography showed a solid testicular mass with hypoechoic and hyperechoic components. Ultrasound abdomen revealed huge retroperitoneal lymphadenopathy. The patient underwent a right radical orchidectomy. The size was 3.0X8.0x7.8 cm. The pathologic examination was high grade leiomyosarcoma, with frequent mitoses. Immunostaining was done which was positive for smooth muscle actin and desmin.

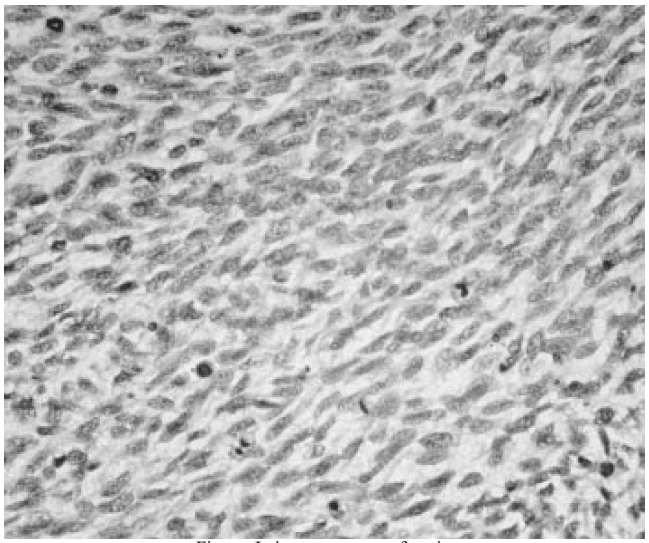

Figure: Leiomyosarcoma of testis.

**Table: Summary of the 13 reported cases.**

| Case No. | Authors | Age | Side | Clinical stage | Treatment | Follow-up (months) | Outcome | Risk factors | Levels of tumour markers |
|---|---|---|---|---|---|---|---|---|---|
| 1 | Yachida | 55 | R | I | Orchidectom-y | 24 | survived | - | normal |
| 2 | Pellice | 37 | L | I | Orchidectom-y | 24 | survived | - | normal |
| 3 | Washec-ka | 47 | R | I | Orchidectom-y | 49 | survived | - | normal |
| 4 | Washec-ka | 40 | R | I | Orchidectomy | 42 | survived | - | normal |
| 5 | Froehner | 32 | R | I | Orchidectomy+RPLND | 79 | survived | anabolic steroid | unknown |
| 6 | Rui Pedro | 19 | L | 1 | Orchidectomy+Adj Chemo+RT | 16 | Survived (metastasis to Lt kidney & ileum) +salvage chemo. | _ | normal |
| 7 | Hachi | 70 | L | I | Orchidectomy | 14 | death (lung metastases) | - | normal |
| 8 | Ali | 65 | R | I | Orchidectomy | 12 | survived | chronic inflammat-ion | normal |
| 9 | Takiza-wa | 76 | L | I | Orchidectomy | 12 | survived | - | normal |
| 10 | Canales | 30 | R | I | Orchidectomy | 6 | survived | radiation | unknown |
| 11 | M.Satta-ry | 27 | L | 1 | Orchidectomy | 30 | Survived | _ | normal |
| 12 | Shunsu-ke case | 73 | L | III | Orchidectomy+Chemo (CYVADIC) | 9 | survived | - | slightly elevated |
| 13 | Labanaris | | | 1 | Orchidectomy | | | | |
| | Current case | 45 | R | 11 | Orchidectomy+ Chemo | | Survived (para Aortic L.N mets) | _ | normal |

R = right; L = left; RPLND = retroperitoneal lymph node dissection.

Metastasis was present in the para-aortic lymph nodes.

## Discussion

Leiomyosarcoma are malignant soft-tissue tumours, arising from the undifferentiated smooth muscle cells of the mesenchymal origin. They may arise anywhere in the body from tissues containing smooth muscles. Leiomyosarcoma of the scrotum have been classified into the paratesticular and the intratesticular, the latter being an uncommon neoplasm. Intratesticular leiomyosarcoma is believed to arise from the smooth muscle elements of the testicular parenchyma such as the blood vessels or the contractile cells of the seminiferous tubules. Leiomyosarcoma is a rare tumour and only 13 cases have been reported (Table).[1-12] Only one case was reported in an infant.[13] The age range was between 19 to 70 years. Six patients had a right-sided tumour while six patients had left-sided tumour. Twelve cases had stage I tumour. Among these thirteen cases only one case had stage III Leiomyosarcoma testis with subcutaneous metastatic nodules of 1 to 4 cm on chest and abdomen.[11]

In all reported cases of intratesticular Leiomyosarcoma a radical orchidectomy was performed.[1-12] Eleven patients who had stage I disease did not receive any adjuvant treatment. One patient received adjuvant chemotherapy (gemcitabine plus docetaxel) and radiotherapy to prevent distant and local metastasis after orchidectomy as the histopathology was high grade intratesticular leiomyosarcoma stage I.[9] After 16 months of follow up CT scan was done which revealed a bulky mass in the retroperitoneum. This mass, kidney and the segment of ileum were removed en bloc. Microscopy confirmed distant recurrence (with invasion of left kidney, ileum, peri-intestinal fat and serosa). Patient received salvage chemotherapy.[9] One patient had clinical stage III disease and received additional CYVADIC (cyclophosphamide, vincristine, adriamycin, decarbazine) chemotherapy.[11]

Our case had a similar presentation. The final diagnosis was primary leiomyosarcoma of the right testes with metastasis to the para-aortic lymph nodes. All tumour markers were within normal range. Treatment was right-orchidectomy followed by chemotherapy.

The aetiology of testicular Leiomyosarcoma is unknown. High doses of anabolic steroids and chronic inflammation are reported to be risk factors for intra-testicular leiomyosarcoma.[1,2] One patient developed intratesticular leiomyosarcoma after receiving testicular radiation for the treatment of leukaemia.[8] Patients with intratesticular leiomyosarcoma are usually older than 40 years. The majority of these patients present with painless testicular enlargement and inguinoscrotal discomfort. These tumours might spread via three routes: local invasion, lymphatic dissemination and haematogenous metastasis.[3]

## Conclusion

Testicular leiomyosarcoma is a rare tumour. Based on a review of literature, the treatment for an intratesticular leiomyosarcoma is a radical orchidectomy and surveillance followed by radiological examination in cases at stage I. There are no available data regarding the management of stage II or Stage III disease. These tumours are rare and radical orchidectomy is the treatment of choice, however standard therapy is difficult to recommend.

## References


1. Froehner M, Fischer R, Leike S, Hakenberg OW, Noack B, Wirth MP. Intratesticular leiomyosarcoma in a young man after high dose of doping Oral-Turinabol: a case report. Cancer 1999; 86: 1571-5.
2. Ali Y, Kehinde EO, Makar R, Al-Awadi KA, Anim JT. Leiomyosarcoma complicating chronic inflammation of the testis. Med Princ Pract 2000; 11: 157-60.
3. Takizawa A, Miura T, Fujinami K, Kawakami S, Osada Y, Kameda Y. Primary testicular leiomyosarcoma. Int J Urol 2005; 12: 596-8.
4. Washecka RM, Mariani AJ, Zuna RE, Honda SA, Chang CD. Primary intratesticular sarcoma. immunohistochemical, ultrastructural and DNA flow cytometric study of two cases with a review of the literature. Cancer 1996; 77: 1524-8.
5. Pellice C, Sabate M, Combalia A, Ribas E, Cosme M. [Leiomyosarcoma of the testis]. J Urol (Paris) 1994; 100: 46-8.
6. Yachia D, Auslaender L. Primary leiomyosarcoma of the testis. J Urol 1989; 141: 955-6.
7. Hachi H, Bougtab A, Amhajji R, Otmany F, al Bouzidi A, Laalou L, et al. A case report of testicular leiomyosarcoma. Med Trop (Mars) 2002; 62: 531-3.
8. Canales BK, Lukasewycz SJ, Manivel JC, Pryor JL. Postradiotherapy intratesticular leiomyosarcoma. Urology 2005; 66: 657.
9. Borges RP, Vila F, Cavades V, Queros J, Marcelo F, Ercinas A. Primary testicular leiomyosarcoma, a case report. Acta Urologica 2007; 4: 45-7.
10. Sattary M, Hazraty B, Saraii MB. Primary pure testicular low-grade leiomyosarcoma. IJMS 2003; 28: 48-50.
11. Yoshimine S, Kono H, Nakagawa K, Kikuchi E, Miyajima A, Kameyama K, et al. Primary intratesticular leiomyosarcoma. Can Urol Assoc J 2009; 3: E74-6.
12. Labanaris AP, Zugor V, Smiszek R, Nutzel R, Kuhn R. Primary leiomyosarcoma of the testis. A case report. Anticancer Res 2010; 30: 1725-6.
13. Wakhlu A, Chaudhary A. Massive leiomyosarcoma of the testis in an infant. J Pediatr Surg 2004; 39: e16-7.