CASE REPORT

Liposarcoma of the Spermatic Cord Presenting as an Indirect Inguinal Hernia

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Liposarcoma of the spermatic cord is a rare condition with a limited number of published reports in literature. These tumors are mostly diagnostic dilemmas where they are mistaken for other inguino-scrotal masses preoperatively and are only accurately diagnosed after surgery. This case report presents a liposarcoma initially mimicking an indirect inguinal hernia on presentation and a definitive diagnosis was confirmed via histopathologic examination. A literature review of other cases that have been previously reported is also presented.

Key words: liposarcoma, spermatic cord, inguinal hernia

Introduction

Liposarcoma is the most common soft tissue malignancy, accounting for 20% of all mesenchymal tumors.[1] However, this tumor appearing in the spermatic cord is a rare occurrence. As of 2016, liposarcoma of the spermatic cord (LSC) has only been published by 185 times all over the world, mostly as case reports.[2]

Limitations in research are also partly due to the fact that this disease is a diagnostic dilemma. In most case reports, liposarcoma of the spermatic cord presents as an inguino-scrotal mass, initially mistaken to be inguinal hernias, hydroceles, chronic epididymitis or lipomas of the cord. Ultrasound is able to confirm the consistency of the mass, but has no pathognomonic features to differentiate benign from malignant masses. CT scan and MRI studies have been found to be useful but a definitive diagnosis cannot be done with these studies alone and they are therefore not widely used.[3]

With the limited literature on LSCs, this case report is presented to provide further information on this malignancy.

The Case

This is a case of a 77 year old male, diagnosed case of right inguinal hernia who underwent right-sided hernioplasty last 2013, who came in due to a right scrotal mass. History of present illness started about two months prior to admission when the patient noted a fist-sized slow-growing, non-tender, non-reducible mass in the right scrotum which was more apparent during coughing. He also complained of moderate lower urinary tract symptoms with frequency, nocturia, intermittency and incomplete bladder emptying. Patient denied any history of fever, vomiting, abdominal pain,

trauma or infection. Physical examination showed a large, non-tender, mobile, right inguinoscrotal mass ~10 cm that could be delineated from the right testis. He had no palpable inguinal lymph nodes. Digital rectal examination showed a tight sphincter tone with an enlarged prostate gland (20-30g, non-nodular, non-tender).

Prior to admission, an ultrasound was done and it showed echogenic structures exhibiting some gliding movement of Valsalva maneuver through an inguinal defect. It also showed multiple hyperechoic tubular structures noted within the right scrotal sac displacing the testis superomedially, which was thought to represent bowel loop segments. Interestingly, color Doppler study showed no definite vascularity in the mass (Figure 1). The radiologist's sign out was an inguinoscrotal hernia on the right. The ultrasound of the kidneys, urinary bladder and prostate showed enlarged prostate of 28 grams.

With these findings, patient was admitted to undergo hernioplasty on the right and cystoscopy and transurethral resection of the prostate gland. After an unremarkable cystoscopy, transurethral resection of the prostate gland, an inguinal exploration on the right was done. Intraoperatively, a firm, round mass, well-delineated from the testis was noted. Patient was also found to have an indirect inguinal hernia on the right with a note of a soft, fatty tissue on the spermatic cord. Patient underwent radical orchiectomy, right with excision of the extratesticular mass and the fatty mass on the spermatic cord (Figure 2). Tissue repair of the indirect inguinal hernia was also done. Specimen was sent for biopsy. The patient's post-operative hospital stay was unremarkable and he was discharged with no pain, fever or hematoma on the 4th post-operative day and the histopathology report was provided on follow-up.

The gross appearance of the extratesticular mass adjacent to the the right testis consists of a tan, brown, irregular, doughy tissue, measuring 11.0cm x 8.5cm x 7.7cm, weighing 230 g. Cut sections showed a yellowish brown, homogenous surface (Figure 3). The gross appearance of the mass on the spermatic cord consisted of a yellowish brown, irregularly ovoid, doughy tissue measuring 9.0cm x 8.5cm x 6.5cm.

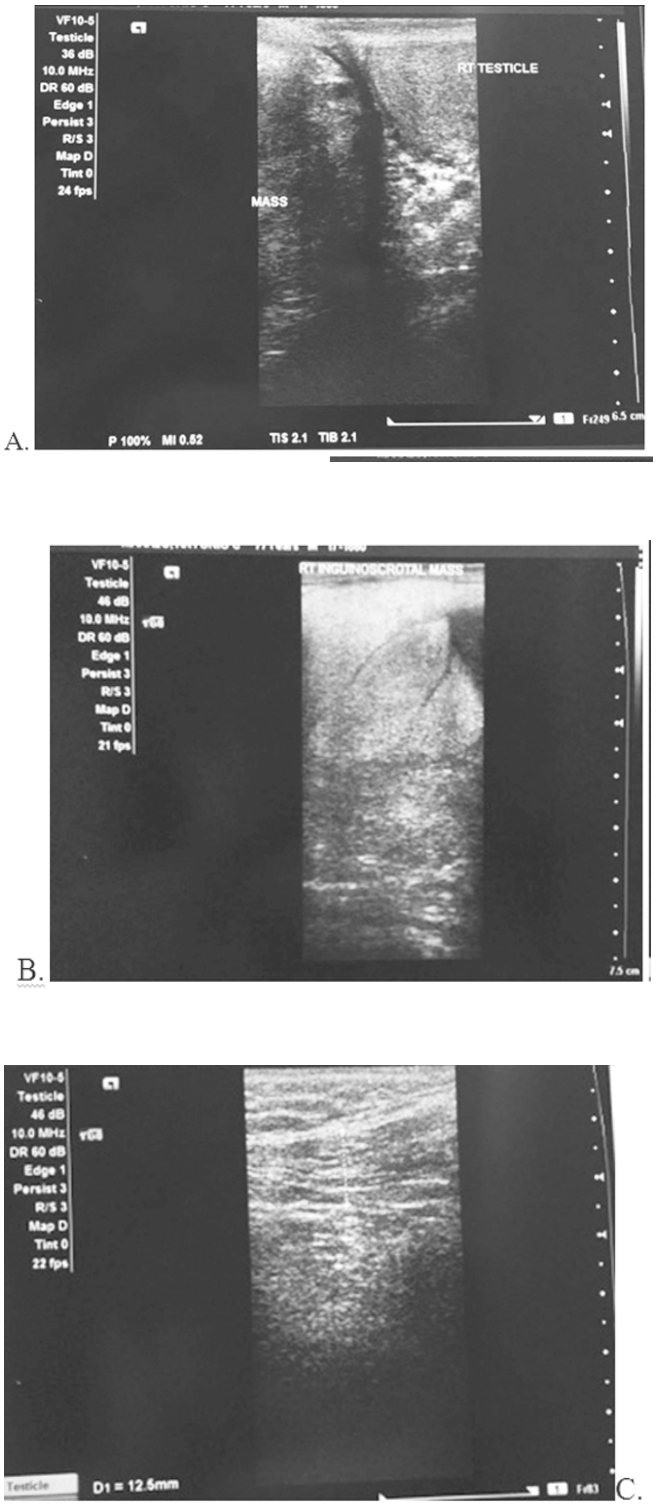

Figure 1. Pre-operative ultrasound findings of the inguinoscrotal mass.
The inguinoscrotal mass was well-delineated from the testis, displacing it superomedially (A). It was composed of echogenic structures exhibiting some gliding movement of Valsalva maneuver (B) through an inguinal defect (C).

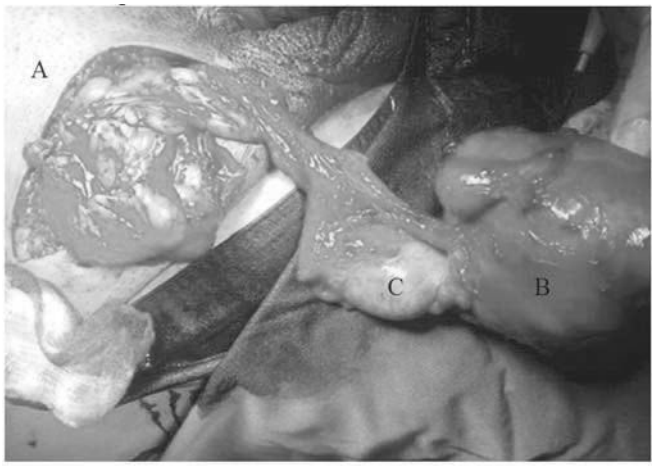

Figure 2. Intraoperative findings.
The mass on the spermatic cord consisted of a yellowish brown, irregularly ovoid, doughy tissue measuring 9.0cm x 8.5cm x 6.5cm (A). The gross appearance of the extratesticular mass from the right testis consists of a tan, brown, irregular, doughy tissue, measuring 11.0cm x 8.5cm x 7.7cm (B), well-delineated from the grossly normal looking right testis (C).

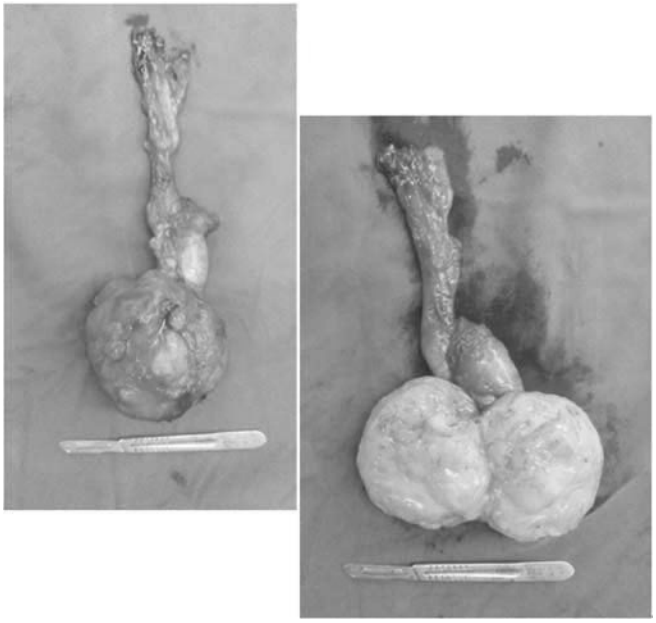

Figure 3. Gross and cut specimen of the extratesticular mass.
Cut sections of the extratesticular mass showed a yellowish brown, homogenous surface.

Histologic examination showed similar features for both the extratesticular mass and the spermatic cord mass. It showed differentiated adipose tissue with some variation in adipocyte size and scattered cells with enlarged hyperchromatic nuclei. There were also irregular fibrous septa that traverse the fatty tissue. This was signed out as a well-differentiated liposarcoma of the cord (Figure 4). Surgical margins were also noted to be negative for tumor involvement (Figure 5). Histopathology of the testis showed testicular atrophy (Figure 6). Prostate tissue resected turned out to be benign prostatic hyperplasia on histopathology (Figure 7).

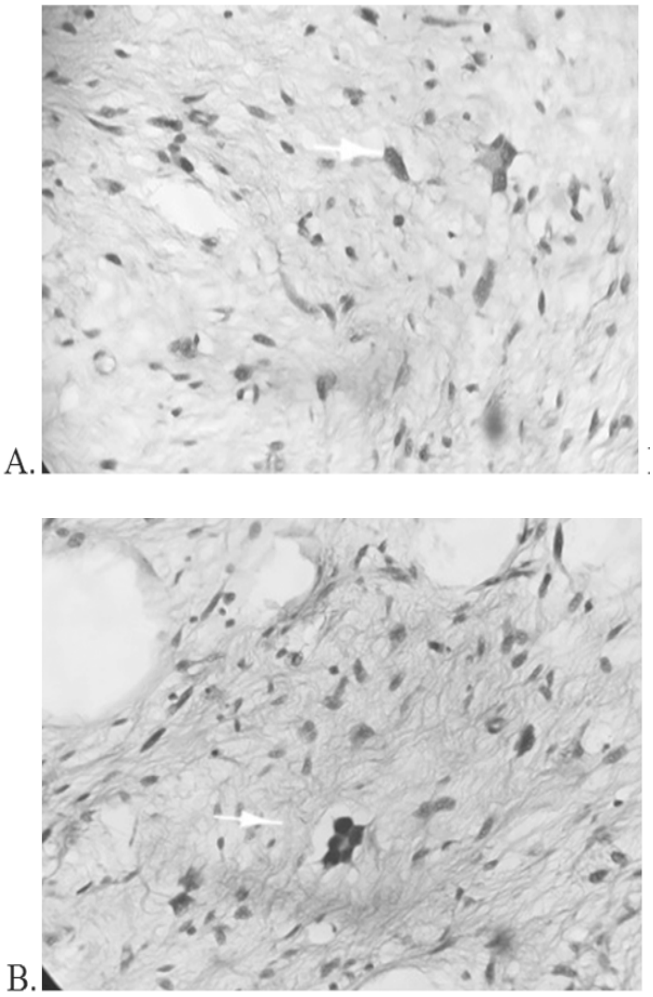

Figure 4. Histopathology of the extratesticular mass and spermatic cord mass.
Histologic examination showed similar features for both the extratesticular mass (A) and the spermatic cord mass (B). It showed differentiated adipose tissue with some variation in adipocyte size and scattered cells with enlarged hyperchromatic nuclei. The specimen had sparse lipoblasts (arrow). There were also irregular fibrous septa that traverse the fatty tissue. They were both signed out as well-differentiated liposarcoma.

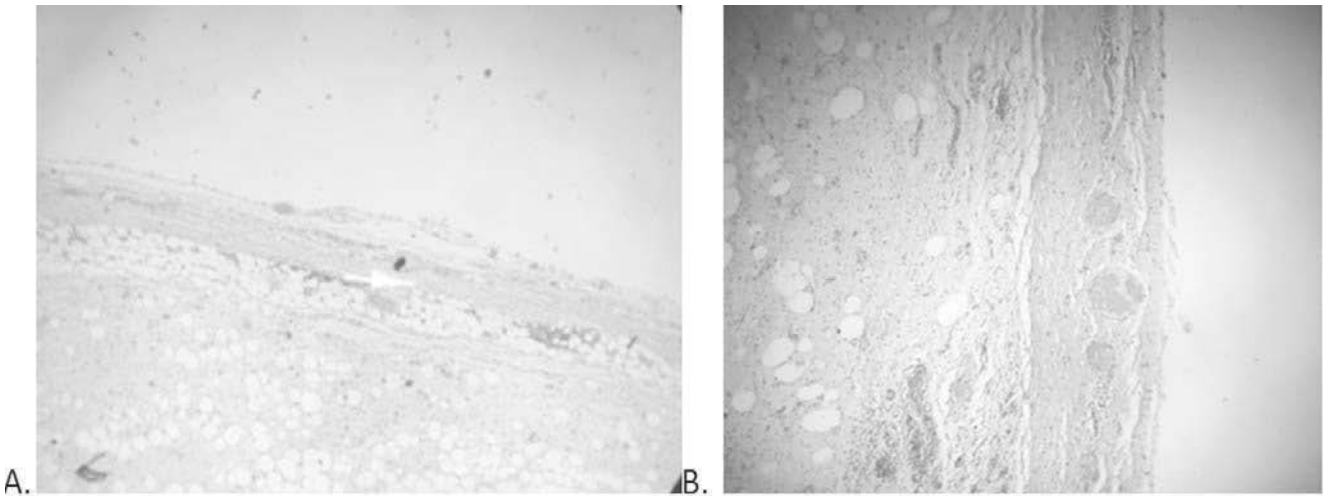

Figure 5. Histopathology of the surgical margins. Surgical margins were negative for tumor involvment.

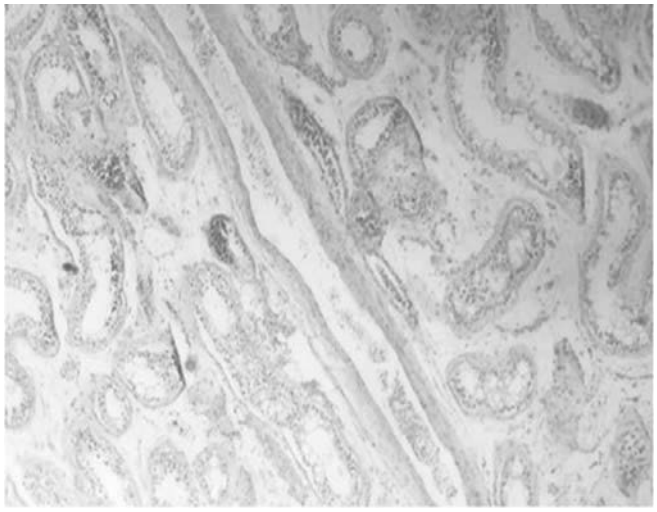

Figure 6. Histopathology of the testis.
Sections from the testis showed small tubules, thick basement membranes and interstitial fibrosis. No tumor involvement was seen. This was signed out as testicular atrophy.

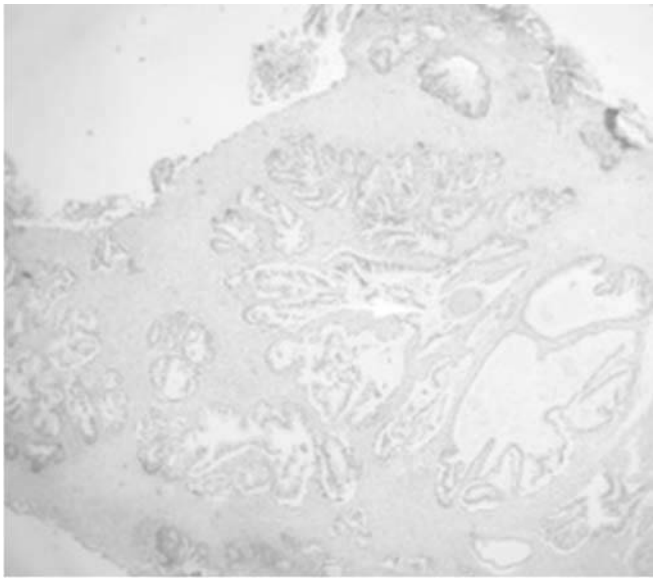

Figure 7. Histopathology of the prostate chips.
Sections of the prostate chips revealed lush and hyperplastic prostatic acini, with complex papilliform structures. This is lined by tall columnar epithelium and basal cells. The glands are aggregated in incomplete nodules by transversing hyperplastic fibromuscular stroma. There are focal inflammatory mononuclear infiltrates with no crowding of glands, no stratification of lining and no disruption of basal cells. No mitosis or necrosis seen. There was no evidence of malignancy in the sections evaluated. This was signed out as benign prostatic hyperplasia.

With the biopsy results, patient was referred to oncology service for further co-management. On metastatic work-up, a chest and upper abdomen CT scan with IV contrast was done which revealed scattered ill-defined faintly hypodense nodules on both lobes of the liver which showed arterial enhancement, the largest of which measured 2.9cm x 4.4cm x 1.9cm at hepatic segment 7, which was signed out as metastatic liver disease. No metastasis was seen on the lungs, no enlarged lymph nodes were noted and no retroperitoneal masses were seen. Patient was started on chemotherapy, given his first cycle of doxorubicin. Two months after the surgery,

patient had no evidence of local recurrence of the mass.

Discussion

Liposarcomas of the spermatic cord are rare. While liposarcomas are the most common soft tissue tumors, sarcomas of the genitourinary tract only represent 2% of soft tissue sarcomas. In the spermatic cord, liposarcomas account for only 3% of malignant lesions appearing in this area.[4]

In a review of 38 cases previously done by Fubiao et al, this tumor occurred more frequently in adults with a mean age of presentation at 61 years old. While the population of the study involved men from ages 24 to 79 years old, majority of the cases (57.9%) were of 60 years and above.[5]

Reports published in different countries noted a high incidence in Japanese men.[5] A liposarcoma of the epididymis in a Filipino man has been reported in one publication from India.[6] In the Philippines, however, no case report on a liposarcoma of the spermatic cord has been published yet.

The common presentation of LSCs is a painless scrotal swelling that is slowly increasing in size throughout a period of months or years.[7]

Imaging studies provide limited diagnostic value for liposarcomas of the spermatic cord. The most common and most convenient imaging modality that can be done is an ultrasound. As mentioned, sonography can provide information on the consistency of the mass and the status of the testes and the cord.[3] For extratesticular masses, it may be used to distinguish the mass from the testis with a high sensitivity (95-100%). Unlike the findings in the patient, liposarcomas are usually seen as hypervascular solid heterogenous lesions on color Doppler study. Still, there are no clear diagnostic criteria for the diagnosis of liposarcoma via ultrasound because of the operator-dependent variable findings.[2] In this case, for example, ultrasound was unable to accurately diagnose the tumor. Computed tomography scanning is not widely used but has been found to be useful to work-up possible liposarcomas. In reports, they appear as heterogeneously low intensity enhancing masses separate from the testis. Again, no pathognomonic features are reliable to differentiate benign from malignant masses.[3,6] Magnetic resonance imaging, which is ideal for soft tissue tumors, may also aid in the characterizing the anatomical extent of the tumor and delineating extratesticular from testicular lesion.[6] PET scanning using FDG has also been reported to have use in evaluation of recurrent cases. However, it has not been established in routine clinical practice. In general, liposarcomas of the spermatic cord do not show specific radiographic patterns therefore a definitive diagnosis should only be made post-opeartively once with histopathology results.[8]

While there is no grading or staging system established specifically for liposarcomas of the spermatic cord, the Federation Nationale des Centres ed Lutte Contre le Cancer (FNLCLCC) grading system for liposarcomas in general is being used in its place as it was found to have to better correlation with overall and metastasis-free survival.[7] This system assigns three grades to the tumor: well-differentiated, de-differentiated and high grade (pleomorphic) liposarcoma. Grade 1 or well-differentiated LSCs histologically consist of mature fat cells mixed with atypical hyperchromatic cells with lipoblasts. This was the histopathologic finding seen in this case's patient. Grade 2 or dedifferentiated LSCs histologically show a progression from a well-differentiated sarcoma with less production of mature cells but more hyperchromatic spindle cells. These already start to have a metastatic potential. In grade 3 or pleomorphic adenomas, there is notable necrosis which correlates with poorer prognosis.[7]

Immunohistochemical stains have also been used to confirm the diagnosis of liposarcoma. In general, the S100 protein is the most specific marker for liposarcomas (90% specificity). For well-differentiated LSCs, MDM2 and CDK4 have been used. High grade liposarcomas on the other hand are often positive for desmin.[4]

Due to the rarity of the tumor, there is still no consensus regarding the management of LCs. Instead, surgeons follow the general recommendations in the surgery of sarcomas. That is, there should be a complete excision of

the tumor with negative microscopic margins. In liposarcomas of the spermatic cord, the main primary surgical procedure is complete excision of the tumor with radical inguinal orchiectomy and high ligation of the cord.[8] Simple excision was found to be inadequate where there was a 27% residual disease in those who underwent repeat wide excision.[9] Local recurrence rates were relatively high after resection alone with 30% recurring at 10 years and 42% recurring at 15 years. A much extensive surgical management such as hemiscrotectomy has been advised in cases where there is clear involvement of the scrotum in histopathology.[8] Currently, there is also a consensus against prophylactic retroperitoneal lymphadenectomy. Aside from the additional morbidity that is brought about by the procedure, there is still no sufficient evidence that prophylactic retroperitoneal lymph node dissection improves prognosis.[8] This may be due to the low incidence of lymph node involvement (5.5%) compared to local recurrence (19.4%) and hematogenous spread (11.1%).[10]

Risk of recurrence is increased in patients with a large tumor size, inguinal location, narrow or positive margins, surgery or manipulation of the tumor, degree of nuclear differentiation and the depth of invasion.[8] In tumors with these pathologic features, the role of adjuvant chemotherapy and radiotherapy has also been investigated but results and recommendations remain controversial.

High rates of local recurrence after surgery have been reported for spermatic cord sarcomas. Recent studies have shown improvement in patients who underwent radiotherapy after surgery (combined modality treatment), with distant metastasis-free rates of 63-85% and overall survival rates of 52-85%.[10] Adjuvant radiotherapy is currently recommended only for high-grade tumors or for patients who are believed to be at high risk of local recurrence. However, there is no uniformity of criteria to indicate when adjuvant therapy is necessary.[11]

There are even more limited studies supporting adjuvant chemotheraphy for patients with liposarcoma of the spermatic cord. The use of chemotherapy regimens have been attempted in treatment of high-grade subtypes or metastatic disease, based mainly on treatment of liposarcomas in general. In a systemic meta-analysis of RCTs of adjuvant chemotherapy for localized resectable soft tissue sarcomas, it was shown that improvement in adjuvant chemotherapy with doxorubicin with ifosfamide reduced the local recurrence rate by 5%. Doxorubicin alone also showed reduction in the recurrence rate but the difference was not statistically significant. Similarly, doxorubicin alone did not significantly improved survival but doxorubicin with ifosfamide showed a reduction of death rate by 11%.[12] However, while improvement could be seen in liposarcomas in general, spermatic cord tumors have been known to be relatively resistant against chemotherapy. It is therefore currently advised for high-grade histologic subtypes and metastatic disease and it is not recommended to be given routinely.[8]

Due to the high risk of recurrence, a close follow-up is advised. Imaging is advised at 3, 6, 12 and 24 months.[13] Some authors suggest close monitoring for a minimum of 36 months with imaging such as chest x-rays, CT scans or bone scans if symptoms arise.

Liposarcoma of the spermatic cord is a rare entity with limited studies, making it difficult for urologists to come up with a consensus regarding diagnosis and treatment. However, with the growing literature on the disease, guidelines for management are continually being developed.

References

1. Karadayi K, Yildiz C, Karakus S, Kurt A, Bozhurt B, Soylu S, Cicekli A, Egilmez R and Cetin A. Well-differentiated abdominal liposarcoma: experience of a tertiary center. World J Surg Oncol 2015; 13: 166 doi: 10.1186/s12957-015-0580-z

2. Draeger DL, Erbersdobler A, Protzel C and Hakenberg O. Malignant spermatic cord tumor as a rare scrotal lesion: case report and literature. J Genital System Disorders 2016; http://dx.doi.org/10.4172/2325-9728.S2-004

3. Chintamani MT, Khandelwal R, Jain S, Narayan N, Kumar Y and Saxena S. Liposarcoma of the spermatic cord a diagnostic dilemma. J Royal Soc Med 2010; 1(6):49. doi: 10.1258/shorts.2010.010058

4. Panus A, Mesina C, Plesea I and Dragoescu P. Paratesticular liposarcoma of the spermatic cord: a case report and review of literature. Romanian J Morphol Embryol 2015; 56(3):1153-1157. Available from: http://www.rjme.ro/RJME/resources/files/56031511531157.pdf

5. Li F, Tian R, Yin C, Dai X, Wang H, Xu N and Guo K. Liposarcoma of the spermatic cord mimiching a left inguinal hernia: a case report and literature review. World J Surg Oncol 2013; 11:18. doi: 10.1186/1477-7819-11-18

6. Ramanathan S, Raghu V, Kumar D and Sempiege VR. Sclerosing liposarcoma of epididymis: Role of chemical shift magnetic resonance imaging. Indian J Radiol Imag 2016; 26:356-9. Available from: http://www.ijri.org/text.asp?2016/26/3/356/190425

7. Logan J, Williams M, Shaves M and McCammon K. Liposarcoma of the spermatic cord: a case review and review of management. (2010) UroToday Int J 1944-5784.2010.08.08. DOI: 10.3834/uij.1944-5784.2010.08.08f2b

8. Rodriguez D and Olumi A. Management of spermatic cord tumors: a rare urologic malignancy. Ther Adv Urol 2012; 4 (6): 325-34. doi: 10.1177/1756287212447839

9. Catton C, Jewett M, O'Sullivan B and Kandel R. Paratesticular sarcoma: failure patterns after definitive local therapy. J Urol 1999; 16(16): 1844-7. doi: http://dx.doi.org/10.1016/S0022-5347(05)68823-4

10. Ballo MT, Zagars GK, Pisters PW, Feig BW, Patel SR, von Eschenbach AC. Spermatic cord sarcoma: outcome, patterns of failure and management. J Urol 2001; 166: 1306-31. https://doi.org/10.1016/S0022-5347(05)65758-8

11. Coleman J, Brennan MF, Alektiar K, Russo P. Adult spermatic cord sarcomas: management and results. Ann Surg Oncol 2003; 10: 669-75. https://doi.org/10.1245/ASO.2003.11.014

12. Pervaiz N, Colterjohn N, Farrokhyar F, Tozer R, Figueredo A, Ghert M. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft tissue sarcomas. Cancer. 2008; 113(3): 573-81. doi: 10.1002/cncr.23592.

13. Chalouhy C, Ruck J, Moukarzel M, Jourdi R, Dagher N, Philosophe B. Current management of liposarcoma of the spermatic cord: a case report and review of literature. Molec Clin Oncol 2007; 6(3):438-40. doi: 10.3892/mco.2017.1157