A Rare Presentation of Liposarcoma of Spermatic cord: A Case report

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Paratesticular tumors represent 7-10% of intra scrotal masses. Sarcomas account for 90% of malignant lesions of the spermatic cord; of these approximately 3-7% are liposarcomas. They are most commonly found in the extremities, in the retroperitoneum and less often in the head and neck area. This report describes a patient with liposarcoma of the spermatic cord. The clinical presentation, pathogenesis, and prognosis of this unusual sarcoma is reviewed. The low-grade malignant potential and irregular growth characteristics of liposarcoma render preoperative diagnosis difficult. The role of radical orchiectomy, retroperitoneal lymph node dissection, radiation therapy and chemotherapy in the treatment of spermatic cord liposarcomas is discussed.

Keywords: Liposarcoma, Radical orchiectomy, Radiation therapy, Chemotherapy.
INTRODUCTION

Liposarcoma of the spermatic cord is a rare condition, representing approximately 7% of paratesticular sarcomas. Around 181 cases are reported in the world literature among which half of the cases have been reported from Japan. There have been occasions when an effort had to be made to differentiate them from inguinal hernias, funicular cysts, lipomas of the cord, etc. Like in this case, preoperative diagnosis is not common and usually they present as operative or histological surprises. The recommended treatment is surgery in the form of wide local excision which in most cases turns out to be a radical high orchiectomy. There is no indication for routine lymph node dissections as the loco-regional lymph nodes are rarely involved. The outcome is fairly good in most cases if the resection is R0. Adjuvant radiotherapy is usually not required except in cases with positive margins or local recurrence and poor prognostic factors. There is no definite role of chemotherapy and most of the recommendations are based on cases with recurrences. These cancers are, however, known for local recurrences and long-term follow-up of up to 10 years is mandatory; as even recurrences after 20 years have been reported.

CASE REPORT

A 62 year old presented with the complaint of an insidiously progressing right inguinal swelling of 6 months’ duration. To begin with, the swelling was painless but developed pain in the few days prior to presentation. Cough aggravated the pain. He was hypertensive and diabetic and was on treatment for the same. He was also on treatment for tricuspid valve dysfunction post percutaneous transluminal coronary angioplasty. Clinical examination revealed a right inguinoscrotal swelling which measured around 8x4cm. It was firm to hard in consistency. No cough impulse or fluctuation could be elicited. The swelling was irreducible and the trans-illumination test was negative. Right testis was distinctly felt separate from the swelling. There was no associated lymphadenopathy. The left inguinoscrotal region was normal. Abdominal examination did not reveal any other palpable lump. Systemic examination did not reveal any other abnormality. Routine investigations including chest x-ray were normal. Echocardiogram showed post angioplasty status. Ultrasound of the region revealed large right inguinoscrotal swelling incarcerated inguinal hernia or lipoma of the cord. Patient was planned for surgery wide local excision/ or right radical orchiectomy for the remote possibility of sarcoma under general anesthesia and appropriate consent was obtained. Through an inguinal approach, the spermatic cord was dissected and delivered out. It appeared as a firm to hard lipomatous mass (11x6cm) involving the entire circumference and was inseparable from the cord. The testis was also delivered into the wound and was found to be normal. An occlusion clamp was applied to the spermatic cord close to the deep ring and in view of the high suspicion of malignancy; a right radical high orchiectomy was done. The histopathological examination revealed dedifferentiated liposarcoma of the cord with clear margin of resection. The patient was not given any adjuvant treatment and he is on regular follow up from last 3 months without loco regional recurrence or progression of tumor.

DISCUSSION

The first case of a spermatic cord sarcoma was reported in 1845 by Lesauvage and subsequently liposarcoma was first described in 1952. It is an uncommon neoplasm that accounts for 3-7% of paratesticular sarcomas. It grows slowly, and may reach 30 cm or more in size. 80% of spermatic cord tumors are benign and usually originate from lipomatous tissue. This tumor is difficult to diagnose preoperatively and is often mistaken
for incarcerated hernia, lipoma or hydrocele or so in our case also. The most common age of presentation is between 50-60 years (range 16-82 years). Regarding the origin of this cancer there are various theories, but most arise de novo. Simple lipomas that constitute nearly 80% of all neoplasm almost never transform into sarcomas. Various authors have hypothesized that a mesenchymal origin rather than malignant transformation of lipomatous cells leads to liposarcomas. Despite being a rare kind of tumor, liposarcomas of the spermatic cord presumably have good prognosis and a high 5-year survival rate. Due to the rarity of this condition, it is difficult to accumulate sufficient cases to learn its natural history and reach conclusions regarding treatment. For example, a 1981 series studied sarcomas of the spermatic cord at the Massachusetts Hospital. Only ten cases were collected between 1940 and 1977, and of these, two were liposarcomas. In a publication of Johns Hopkins Hospital, from 1980 to 2000 only three cases were identified.

USG provides little information on paratesticular sarcomas, as some are visualized as homogenous and isoechochogenic, others as inhomogeneous and echo-density being quite variable. The use of CT scans is not widely reported, but seems to be promising, as liposarcomas are of low density and can be well demarcated, but no pathognomonic features for the differentiation of benign versus malignant masses are defined. Use of MRI provides good information on the local extent, but an exact evaluation of any such mass again cannot be ascertained. FDG–PET scan may be of use in recurrent cases but their routine use is not indicated.

The tumor is often classified histologically into 4 subtypes: Well-differentiated, Myxoid and round cells, Pleomorphic, Dedifferentiated. Most paratesticular liposarcomas are well-differentiated (40-45%), sclerosing and inflammatory according to current criteria. This has a good correlation with the prognosis of these tumors, notwithstanding their large size according to Montgomery. For liposarcoma the most specific immunohistochemical marker is the S100 protein which is positive in up to 90% of cases. High-grade tumors are often positive for desmin.

Liposarcomas are locally aggressive tumors and recurrence is quite common after incomplete excision. The inguinal radical orchiectomy with wide resection margin is the standard approach for sarcomas of the spermatic cord. Some authors favor a hemiscrotectomy in addition to the inguinal orchiectomy. Sometimes a second resection is advised if the margins are positive. Local radical excision alone seems to be insufficient for liposarcomas since local recurrence is a major problem occurring in up to 50% of the patients. Due to the rarity of this condition, the role of radiotherapy and chemotherapy has not yet been well established. A review by Stephen in 1995 reports the results of treatment in six cases of liposarcoma of the spermatic cord; the study shows that there is no response to radiotherapy and chemotherapy (doxorubicin). The article also mentions that retroperitoneal lymphadenectomy does not offer any therapeutic benefits. This is explained by the fact that the
main pattern of dissemination of sarcomas of the spermatic cord is by contiguity through the inguinal canal into the abdominal cavity; the blood and lymph routes are less common\textsuperscript{30}. In a study of all histological types of sarcomas of the spermatic cord, the 5-year survival rate was 75\%, 50\% of patients had recurrence and of patients with liposarcoma, only 4\% had metastases at the time of diagnosis\textsuperscript{7}. A long periodic follow-up is recommended because the local control of this disease may be compromised by very late recurrences. These check-ups should include a chest x-ray and a bone scan (if there are suspicious bone symptoms) because although the incidence of metastatic dissemination via blood is low, it may still occur\textsuperscript{16,23,24}.

CONCLUSION

Sarcomas of the spermatic cord are rare neoplasms that should be considered in the differential diagnosis of scrotal masses. Imaging techniques such as scrotal ultrasound, axial tomography and magnetic resonance should be used to assess these masses. Initial management is surgical. We recommend even a trans-operative assessment of the surgical edges in order to extend the margins of the excision if needed in order to try to reduce the chance of recurrence. Complementary treatment for recurrences is controversial as on date and must be assessed on an individual basis for each patient: for this reason we also recommend that these patients be followed-up closely for a longer period of time.

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