Paratesticular liposarcoma of the spermatic cord: A rare case

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ABSTRACT

Spermatic cord liposarcomas (LPS) are rare representing 7% of paratesticular sarcomas. The most present as slow growing inguinal or scrotal masses that need to be differentiated from inguinal hernias, testicular tumors, hydrocele, and benign lipomas. A 57-year-old male presented with right scrotal swelling since 1 year that grew rapidly in last 4 months. On examination, a firm, nontender mass was felt in the right scrotum measuring 12 cm × 8 cm. A clinical diagnosis of right testicular tumor was made. Ultrasound scrotum—large, heteroechoic mass separate from right testis. Magnetic resonance imaging scrotum well-defined, homogenously enhancing extra testicular mass with fat component? Paratesticular lipoma/liposarcoma. Right radical orchiectomy, high ligation of spermatic cord, and excision of mass en bloc were performed. Gross findings: Multiple, globular, gray yellow, firm masses seen in the paratesticular area and spermatic cord, the largest measuring 11 cm × 7 cm × 5 cm. Testis, epididymis-normal. Cut surface—solid, yellowish white with myxoid areas. Microscopy-mature adipocytes of varying sizes separated by dense fibrillary fibrocollagenous stroma with pleomorphic, bizarre stromal cells, and multinucleate giant cells. Interspersed areas showed extensive mast cell infiltration and myxoid change. Diagnosis, well-differentiated liposarcoma/atypical lipomatous tumor sclerosing type of paratesticular area (spermatic cord). Paratesticular LPS of spermatic cord are rare tumors often misdiagnosed clinically resulting in incomplete excision and high recurrences. This report highlights the need of considering liposarcoma as a differential diagnosis in inguinocrotal masses and stresses the role of histopathological examination in making a definitive diagnosis, which cannot be established clinically/radiologically.

KEY WORDS: Aggressive, paratesticular liposarcoma, recurrence, spermatic cord

INTRODUCTION

Liposarcomas (LPS) are malignant mesenchymal neoplasms of adipocytic origin [1]. Well-differentiated LPS are common in deep soft tissues of limbs, retroperitoneum, paratesticular area, and mediastinum [1].

Paratesticular LPS of the spermatic cord are rare tumor, accounting for about 7% of all paratesticular sarcomas [2]. Until date, about 200 cases have been reported in literature, most being reported as isolated cases; hence, the natural history and biological behavior of this rare tumor are less understood [3,4]. Most patients usually present with a slowly growing painless inguinocrotal mass mimicking hernias or testicular tumors clinically [3].

Herein, we report a rare case of paratesticular well-differentiated LPS of spermatic cord sclerosing type, which was clinically diagnosed as a testicular tumor.

CASE REPORT

A 57-year-old male presented with a history of swelling in the right scrotum for 1 year duration that had started growing rapidly in last 4 months and had reached the size of 5 cm × 5 cm. On examination, a firm, nontender, smooth surfaced mass were felt in the right scrotal sac measuring 12 cm × 8 cm. Transillumination test was negative. There was no associated inguinal lymphadenopathy. Left scrotum and testis were normal. A clinical diagnosis of the right testicular tumor was made. Ultrasound scrotum revealed a large heteroechoic mass lesion measuring 11 cm × 6 cm seen separate from the right testis. Magnetic resonance imaging (MRI) of the scrotum revealed a multiple, well-defined, homogenously enhancing extra testicular mass with fat component? Paratesticular lipoma/liposarcoma with testis being normal and pushed to the left [Figure 1]. A right-sided radical inguinal orchiectomy with high ligation of spermatic cord and excision of the mass en bloc was performed and sent for histopathological examination. GROSS—multiple, globular, gray yellow to gray white, firm, rubbery masses of varying sizes were seen in the spermatic cord and paratesticular area with the largest measuring 11 cm × 7 cm × 5 cm [Figure 2]. The testis and epididymis were free of the tumor. Cut surface was solid, yellowish-white to gray white with myxoid areas. Microscopy showed a tumor composed of mature adipocytes of varying sizes separated by dense fibrillary fibrocollagenous stroma...
Furthermore, noted were pleomorphic, bizarre stromal cells which showed nucleomegaly, irregular nuclear membranes, and smudgy chromatin with multinucleated giant cells [Figure 4]. Interspersed areas showed dense sclerosis, extensive mast cell infiltration, and myxoid areas. Margins were free from tumor. A diagnosis of well-differentiated liposarcoma/atypical lipomatous tumor–sclerosing type of the paratesticular area spermatic cord was made. Since the tumor was of low grade with negative margins with no evidence of metastasis, no adjuvant radiotherapy was advised. The patient was discharged on the 7th post-operative day with an uneventful post-operative course. The patient is on regular follow-up since 6 months.

DISCUSSION

Atypical lipomatous tumor/well-differentiated LPS are an intermediate (locally aggressive) mesenchymal neoplasm of adipocytic origin [1]. They most frequently occur in the deep soft tissue of the limb followed by retroperitoneum, paratesticular area, and mediastinum [1]. On microscopy, they are of 5 types–well-differentiated LPS, dedifferentiated LPS, myxoid LPS, pleomorphic LPS, and mixed type LPS [1].

Paratesticular LPS of spermatic cord are rare tumors, accounting for 3-7% of malignant paratesticular tumors. The first case was reported by Lesauvage in 1845 [5]. Paratesticular region includes spermatic cord, testicular tunics, epididymis, and vestigial remnants. About 90% of all paratesticular LPS arise from the spermatic cord. Most cases occur in the age group of 41-87 years [6].

Clinically, most cases present with a slow growing, a painless inguinoscrotal mass which is clinically indistinguishable from testicular tumors, inguinal hernia, lipomas, hydrocele, spermatocele, and hematocoele [7,8]. No specific diagnostic procedure can establish the diagnosis. Ultrasonography provides a little information on paratesticular LPS, as some are homogenous and isoechogetic, whereas others are inhomogenous and have variable echodensity.
On CT, LPS are of low density and well-demarcated, and there are no pathognomonic features differentiating benign from malignant masses. MRI provides good information on the local situation, but an exact evaluation of any masses again cannot be obtained [9].

Morphologically, well-differentiated LPS can be subdivided into 4 main subtypes—adipocytic (lipoma-like), sclerosing, inflammatory, and spindle cell types [1]. Sclerosing LPS consist largely of collagenous fibrous tissue that tends to have a somewhat delicate, almost fibrillary appearance. Scattered within this fibrous tissue are mature adipocytes and bizarre, hyperchromatic stromal cells which are often multinucleated; in many cases, lipoblasts are hard to find [10]. Paratesticular LPS usually have a combination of lipoma-like and sclerosing patterns. Multivacuolated lipoblasts may be present but are not required for diagnosis [11].

Immunohistochemistry plays a very minute role in the differential diagnosis of LPS; adipocytes usually exhibit S-100 protein immunoreactivity that may be helpful in highlighting the presence of lipoblasts and are negative for CD34, actin, keratin, and desmin [1,12].

Fluorescence in situ hybridization and comparative genomic hybridization show MDM2 gene amplification which is a sensitive and specific tool for diagnosis of LPS [13].

The well-differentiated and myxoid LPS have favorable prognosis, whereas dedifferentiated, pleomorphic, and mixed LPS are associated with multiple recurrence and metastasis [14].

The microscopic differential diagnosis is considered as spindle cell/pleomorphic lipoma, lipoma with degenerative/atrophic changes, sclerosing inflammatory lesion, fibromatosis, neurofibroma, and dermatofibrosarcoma protuberans. Spindle cell lipoma is common in the neck and upper back, microscopically composed of bland spindle cells and thick rope collagen. They are CD34 positive, rarely positive for S-100. Pleomorphic lipoma is common in the neck and upper back with floret-like cells and is CD34 positive [1]. Lipoma with degenerative/atrophic changes and sclerosing inflammatory lesion are differentiated from LPS by the absence of adipocytic nuclear atypia and bizarre hyperchromatic stromal cells [10]. Fibromatosis are more common in the abdomen, shoulder, and chest wall, and are more cellular with uniform spindle cells without atypia; they are CD34 negative but positive for muscle specific actin and smooth muscle actin [1]. Neurofibroma, a nerve sheath tumor is hypocellular without any nuclear pleomorphism/mitoses. S-100 is positive [12] DFSP is highly cellular with storiform patterns and entrapped fat at the periphery; they lack lipoblasts [12].

The most important prognostic factor for well-differentiated LPS is the anatomic location. Paratesticular LPS tend to recur repeatedly, especially after incomplete excision with positive margins [1]. The reported recurrence is in 50% cases, making it mandatory to follow-up these cases for a very long time period [14,15].

The treatment of choice for paratesticular LPS of the spermatic cord is radical orchidectomy with high ligation of the spermatic cord and en bloc removal of the tumor [16]. The benefit of adjuvant radiotherapy for margin-positive cases is not well-established and controversial.

The present case highlights the rarity of paratesticular LPS of spermatic cord which are often misdiagnosed clinically as testicular tumors. We have also discussed the microscopic differential diagnosis, which have to be considered, especially in the absence of lipoblasts.

CONCLUSION

Paratesticular LPS of the spermatic cord are rare types of tumors which are highly aggressive with local recurrences when excised incompletely. Clinically, they are often mistaken for testicular tumors; hence, LPS’s should always be considered in the differential diagnosis of inguinoscrotal masses. Microscopically, a sclerosing variant of LPS is common in the paratesticular area, and it is difficult to find lipoblasts in this variant and lipoblasts are not always mandatory for the diagnosis of LPS in this location.

We have reported a rare case of paratesticular LPS of spermatic cord and have highlighted the role of histopathological examination in the diagnosis of this tumor, as most often, it cannot be diagnosed clinically and radiologically.

REFERENCES

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