Case Report

Paratesticular Liposarcoma in an Adolescent Male: Case Report of a Rare Tumor

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Abstract

Paratesticular liposarcomas (PLs), first reported in 1952, are rare tumors that comprise approximately 3%–7% of all paratesticular sarcomas. PL of unknown etiology typically affects adults aged 50–60 years and rarely occurs in the young. Here, we report one case of an 18-year-old male patient presenting with a mobile, firm, painless, and nontender testicular mass measuring about 7 cm in greatest dimension. Testicular tumor markers were negative. On cytological (fine-needle aspiration cytology) examination, features were suggestive of a malignant mesenchymal tumor. Subsequently, he underwent radical orchiectomy along with high ligation of the spermatic cord. A wide local scrotal excision was also done, which was followed by histopathological examination. A diagnosis of PL was made which was confirmed by immunohistochemical examination with vimentin positive and negative for desmin and myogenin. Since there is no definite consensus of opinion as regards to the role of adjuvant radiotherapy and chemotherapy, the patient received none. He remained recurrence free after an 18-month regular follow-up.

Keywords: *Desmin, paratesticular liposarcoma, vimentin*

Introduction

Most masses encountered in the scrotal sac are within the testis and are neoplastic. However, a subset of these tumors is extratesticular and mostly arises from paratesticular tissue. The paratesticular region is a complex anatomical area which includes the contents of the spermatic cord, testicular tunics, epididymis, and vestigial remnants. Primary paratesticular tumors are rare, accounting for only 7%–10% of all intrascrotal tumors. More than 75% of these lesions arise from the spermatic cord.[1-3] The most frequently reported benign scrotal tumors are hemangioma, lymphangioma, leiomyoma, and lipoma.[4] Among the malignant tumors, the most common histological type is liposarcoma (46.4%), followed by leiomyosarcoma (LMS) (20%), malignant fibrous histiocytomas (13%), and embryonal rhabdomyosarcoma (9%).[5] Paratesticular liposarcomas (PLs) are rare tumors that comprise approximately 3–7% of all paratesticular sarcomas[6,7] and typically affect adults aged 50–60 years.[8]

Case Report

An 18 years old male presented in the surgery outpatient department with a left-sided painless scrotal swelling that has gradually increased in size for the past 6 months. On examination, a firm swelling was felt adjacent to left testes. The testicular sensation was preserved and there was no significant palpable inguinal lymphadenopathy. On ultrasonography, left testis was normal in size and shape, but a solid space occupying heterogeneous and hypoechoic lesion of 7.2 cm × 4.5 cm was noted in the epididymis [Figure 1a]. Right testis and adnexa were normal. The mass involved the body and tail of epididymis. Moderate amount of left-sided hydrocele was noted. Preoperative blood parameters were within normal range (serum beta-human chorionic gonadotropin <1.2 mIU/ml, alpha-fetoprotein - 1.04 ng/ml [normal range - 0.89–8.79 ng/ml], and lactate dehydrogenase - 257 U/l [normal range - 248 U/l]). Fine-needle aspiration cytology revealed cellular cytosmears showing mostly dispersed population of spindled to ovoid cells with hyperchromatic nuclei [Figure 2a]. Many large cells with multivacuolated cytoplasm and scalloped nuclei were noted in small clusters as well as dispersed singly. Fine anastomosing capillary vessels and myxoid matrix were appreciated. A provisional diagnosis of pleomorphic spindle cell sarcoma was made.

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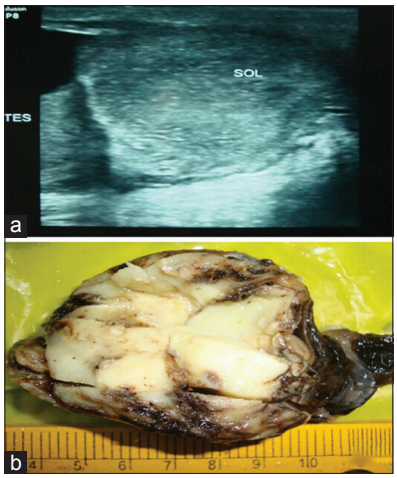

Figure 1: (a) Ultrasonography image showing a heterogeneous and hypoechoic lesion separate from the testis. (b) Gross image showing a lobulated solid white mass almost obliterating the normal testicular tissue

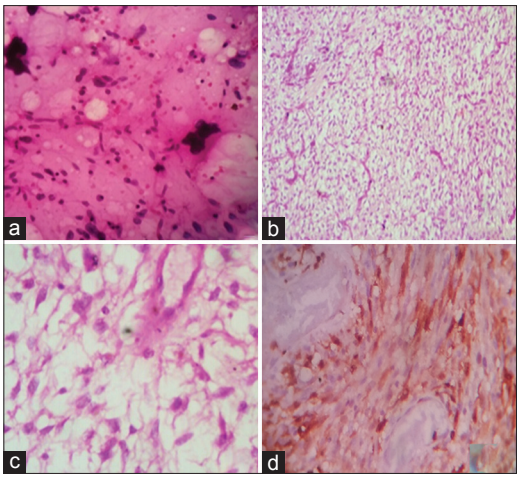

Figure 2: (a) Cytology image (×400) showing dispersed pleomorphic cells. (b) H and E image (×100) showing spindle-shaped cells and fine arborizing vasculature. (c) H and E image (×400) showing pleomorphic spindle-shaped cells with hyperchromatic nuclei and cytoplasmic vacuolation along with myxoid degeneration of the stroma. (d) Immunohistochemistry showing S-100 positivity in tumor cells

High inguinal orchiectomy was done subsequently and the specimen was sent for histopathological examination to the pathology department. Grossly, the specimen measured 8 cm × 4 cm × 3 cm and attached spermatic cord was 2 cm in length. On slicing, a lobulated solid white mass noted [Figure 1b]. In naked eye examination, normal testicular tissue could not be separated from the mass. Representative sections from the tumor mass were taken.

On histopathological examination, sections from spermatic cord, tunica, and testis showed the presence of a tumor tissue composed of pleomorphic spindle-shaped cells with hyperchromatic nuclei and cytoplasmic vacuolation. Multivacuolated cells with indented hyperchromatic [Figure 2c] nucleus (resembling lipoblast) were also noted. Fine arborizing vasculature and myxoid degeneration of the stroma were identified [Figure 2b]. Mitotic figures including atypical forms were also present. Normal testicular tissue was focally identified. The above histological features were that of a malignant mesenchymal tumor and suggestive of well-differentiated liposarcoma. Immunohistochemical staining was done keeping in mind the differential diagnosis of spindle cell variant of embryonal rhabdomyosarcoma. Immunohistochemistry panel of markers such as myogenin and desmin were done which came out as negative. S100 was positive focally [Figure 2d]. Postoperative ultrasonography (USG) and computed tomography scan reports were unremarkable.

The patient was placed in a multidisciplinary tumor board, and no adjuvant therapy was offered as tumor was of low grade and the margins were negative. The patient was observed, and till 18 months postoperative, there was no recurrence on clinical examination and routine USG.

Discussion

Primary tumors of the paratesticular region are rare, with paratesticular sarcomas constituting a major proportion of these tumors, particularly in the elderly. Of these, liposarcoma and LMSs are most common.

PL is a rare entity making it difficult to have a universal consensus on the natural history and management even in large institutions. It typically affects adults with most common age of presentation being 50–60 years, as per available literature.[8] However, in the present case, the patient is an adolescent male, aged 18 years. Liposarcoma is classified into four subtypes – well-differentiated, myxoid/round cell, pleomorphic, and dedifferentiated.[9] The prognosis of PLs depends on the histological cell type. The well-differentiated and myxoid/round cell types have a better prognosis, but they tend to have a high incidence of local recurrence.[10] The reported recurrence rate ranges between 46% and 57%.[11]

These tumors generally present as painless scrotal swelling easily mistaken for inguinal hernia or hydrocele. An USG of the scrotum generally reveals heterogeneous scrotal mass with varying echogenicity separate from normal appearing testes. They commonly involve the spermatic cord or testicular tunics but can also rarely involve the epididymis as was the present case. The diagnosis is mainly clinicoradiological that is confirmed postoperatively by histopathology. Radical orchiectomy remains the mainstay of initial treatment for PLs. Since they have high tendency (60%) for local recurrence after inadequate resection, complete resection, including high ligation of the spermatic cord, is indicated.[12] An intralesional biopsy or surgery should be avoided to prevent spillage of malignant cells. Some authors have even discussed the possibility of the dedifferentiation.[13] PL shows local invasion of the crus of penis, in some cases. Visceral metastasis and bone metastasis may occur in rare instances.

The differential diagnosis from lipoma, spindle cell lipoma, and dedifferentiated liposarcoma is based on light microscopic examination because immunohistochemistry is of little value in distinguishing these neoplasms.[14] Rhabdomyosarcoma is the most common malignant tumor (17%) and may arise from the distal spermatic cord and appear as a scrotal mass or hydrocele in prepubertal age group.[15] Embryonal rhabdomyosarcoma comes close as a diagnostic possibility. Such tumors are composed of primitive mesenchymal cells in various stages of myogenesis. Sheets of moderate to poorly diffrerentiated round to spindloid cells having scant cytopplasm and eccentric oval nucei are found, along with occasional rhabdomyoblasts. Highly cellular areas around blood vessels alternate with paucicellular mucoid regions (resembles normal embryogenesis). Cross-striations are seen frequently. Rarely anaplastic features, clear cell change, rhabdoid features, and neuronal, melanocytic, or schwannian differentiation (ectomesenchymoma) may be present.

There is no established role of adjuvant therapy in PL as there is a scarcity of evidence suggesting the role of adjuvant radiotherapy or chemotherapy in decreasing recurrence. The role of chemotherapy remains limited in cases of metastatic tumors. Radiation therapy may be employed as an adjunct to surgical resection in cases following incomplete resection, in an attempt to avoid local recurrence.[16] Doxorubicin-based chemotherapy with contradicting results has also been reported.[17] Since being a rare disease and having long course before recurrence, surgery with adequate margin followed by observation is the most rational approach, which was adopted. However, in the present case, since it has occurred in a young patient, long-term surveillance is mandated.

Conclusion

We report a case of liposarcoma arising from epididymis, proved by histopathology and immunohistochemistry in an 18-year-old male for its rarity and uniqueness. Furthermore, PL should feature in the differential diagnosis of all paratesticular masses, even in younger age group, as it has a propensity for late recurrences and metastases after surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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