DOI: 10.7860/JCDR/2014/8147.4045

Case Report

Pathology Section

Paratesticular Liposarcoma-Masquerading as a Testicular Tumour

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ABSTRACT

Paratesticular liposarcomas are rare tumours which account for 12% of all liposarcomas. Probably there are about 186 cases which have been reported till date. They must be differentiated from tumours of testicular origin which have extension to the spermatic cord. We are reporting a case of a 50-year-old male who had presented with a painless swelling in the right hemiscrotum, which was of 20 years' duration. Inititally, a clinical diagnosis of testicular tumour was made; however, CT of the scrotum revealed paratesticular tumour? liposarcoma and testis being normal and displaced postero-inferiorly. Metastatic work-up, which included CT of the abdomen and pelvis, thorax and whole body scan, did not reveal any distant metastasis. Patient underwent high orchidectomy, hemiscrotectomy. Histopathological studies confirmed the diagnosis of well-differentiated liposarcoma (atypical lipomatous tumour of sclerosing type).

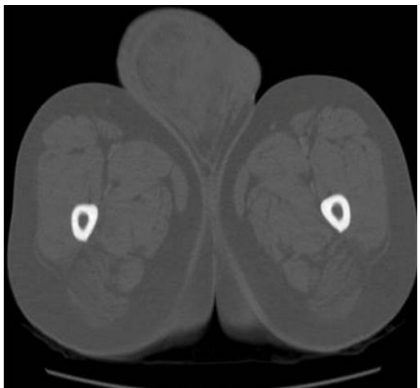
Keywords: Paratesticular liposarcoma, Atypical lipomatous tumour, High orchidectomy

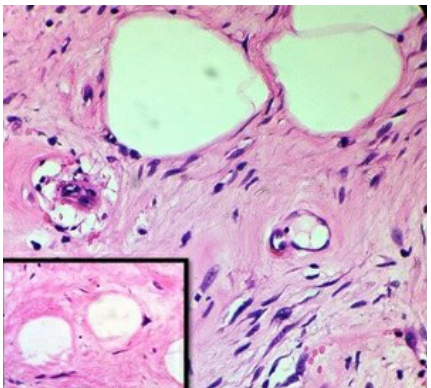
CASE REPORT

A 50-year-old male presented with a swelling in the right hemiscrotum, which had duration of 20 years, which had gradually increased in size and had developed dull aching pain since 4 months. On examination, a hard, non-tender mass was felt in right hemiscrotum, which measured 20×15 cm. Clinically, testis was not felt separately. With a clinical diagnosis of testicular tumour, patient was subjected to investigations. Ultrasonography indicated a right testicular tumour. CT of abdomen, pelvis and scrotum [Table/Fig-1] was advised, which reported it as right extratesticular mass-probably liposarcoma, the right testis being normal and displaced postero-inferiorly, with no retroperitoneal extension or pelvic lymph node enlargement. With a clinical suspicion of paratesticular tumour, patient was subjected to metastatic work-up, including whole body bone scan and CT of thorax. These investigations did not reveal any distant metastasis. Other blood investigations were within normal limits. High orchidectomy, i.e. ligating and dividing the spermatic cord at the deep inguinal ring, was undertaken, along with removal of the entire testis. Operative findings were a large, hard tumour which arose from the paratesticular region, right testis and spermatic cord being normal. He also had right direct inguinal hernia. He underwent high orchidectomy, hemiscrotectomy and right hernioplasty. The patient's post-operative course was uneventful and he was discharged on the 5th post-operative day, though mild infection of the scrotum which was present, was treated with antibiotics on outpatient basis. The tumour, on gross examination, was 20×14.5×9.5 cm in size. The testis was identified separately. Cut surface was solid, yellowish white, along with interspersed fatty areas and myxoid change [Table/Fig-2]. Microscopy revealed an infiltrative tumour amidst dense fibrocollagenous tissue, which had a fibrillary appearance. Scattered amongst them were mature adipocytes and bizarre hyperchromatic stromal cells. Scanty atypical lipoblasts with hyperchromatic, irregular nuclei, with indented, scalloped nuclear membranes were noted. Interspersed areas showed dense sclerotic tissue with mixed inflammatory infiltrate [Table/Fig-3]. Margins were free from tumour. Lymphovascular invasion was absent. The right testis and spermatic cord were normal. A diagnosis of atypical lipomatous tumour, sclerosing subtype (well differentiated liposarcoma) was reported. Since there was no metastasis and as it was a low grade tumour, patient was not advised further adjuvant radiotherapy or chemotherapy. He is on regular follow-up since 2 months after surgery.

DISCUSSION

Paratesticular liposarcomas are extremely rare malignant tumours which account for approximately 3-7% of all paratesticular sarcomas. They are clinically indistinguishable from testicular tumours, which thus result in difficulty in diagnosis and management. There are 186 similar cases which have been reported till date [1]. Lesauvage reported the first case of sarcoma of spermatic cord [2]. The paratesticular region includes contents of the spermatic





[Table/Fig-1]: CT pelvis shows a large heterogenous extra-testicular mass lesion in right scrotal region with areas of fat density
[Table/Fig-2]: Gross photograph showing right paratesticular mass with solid, yellow-white, with fatty and myxoid areas. Also seen are testis and scrotal skin
[Table/Fig-3]: Microphotograph shows mature adipocytes and bizarre hyperchromatic stromal cells amidst dense fibrocollagenous tissue. Inset- shows atypical lipoblasts with scalloped nucleus. (H & E, X 400)

cord, testicular tunics, epididymis and vestigeal remnants. The site of origin of paratesticular tumours may be difficult to determine , the spermatic cord being the most common one, accounting for 90% of cases [3]. These tumours may arise de novo from the adipose tissue around the spermatic cord or by malignant transformation of a pre-existing lipoma [3]. Liposarcomas are the most common soft tissue sarcomas. They are located in the extremities and retroperitoneum in about 70% of cases. About 12% of the liposarcomas are paratesticular in location [4]. Almost 70% of paratesticular tumours are benign, and 30% are malignant. Most of the paratesticular liposarcomas are well-differentiated and they present in the fifth and sixth decades of life in the form of slow-growing painless inguinal or inguinoscrotal masses .They can be mistaken for inguinal hernia, lipoma, hydrocoele, spermatocoele, haematocoele, epididymo-orchitis or testicular tumour [1,3,4]. Three distinct types of liposarcomas have been described, well differentiated (WDLPS) or atypical lipomatous tumour, myxoid/ round cell type and pleomorphic type. Well-differentiated liposarcomas grow slowly , but they tend to recur if they are incompletely excised. The histologic subtypes of WDLPS are, adipocytic (lipoma-like), sclerosing, inflammatory and spindle cell types [5]. The sclerosing type is more commonly seen in retroperitoneum and spermatic cord. Microscopy of sclerosing type of WDLPS reveals fibrocollagenous tissue with a fibrillary appearance. Scattered amongst it are mature adipocytes and bizarre hyperchromatic stromal cells, with few lipoblasts [5]. Special stains for lipids play no essential role, except that occasionally, S100 protein staining highlights lipoblasts. Fluorescence in situ hybridization (FISH) for MDM2 amplification is a sensitive and specific tool which can be used for distinguishing difficult and doubtful cases of well differentiated liposarcoma/atypical lipomatous tumours from benign lipomatous neoplasms [6]. P16 an immunohistochemical marker, is sensitive for and is positive in atypical lipomatous tumours [7]. IHC analysis of liposarcomas shows S100 positivity, while CD 34, actin, keratin, desmin all show negativity [8]. In our case, the histopathological features were diagnostic and in favour of WDLPS (atypical lipomatous tumour)- sclerosing subtype. The differential diagnosis of low grade WDLPS includes spindle cell lipoma, pleomorphic lipoma, neurofibroma, dermatofibrosarcoma protruberans. Spindle cell lipomas are commonly seen in neck and upper back. Spindle cells are bland, with ropy collagen and they have atypical cells. Immunohistochemistry (IHC) reveals CD34 positivity, whereas MDM2(0-12% +) CDK4(0-6%+) and S100 show negativity [5]. Pleomorphic lipomas are seen in posterior neck and shoulder. Circumscription with encapsulation is seen, unlike in liposarcoma. Floret cells are present. IHC reveals CD34 positivity [8]. Neurofibroma, a nerve sheath tumour, is seen in skin and subcutaneous tissues. Microscopically, it is a hypocellular lesion, fat is not an integral part of it, no pleomorphism is noted. On IHC, S100 protein is found to be positive. Dermatofibrosarcoma protruberans is a cellular tumour with storiform pattern without pleomorphism, with entrapped fat in periphery only. No lipoblasts are seen. IHC reveals CD 34 positivity [8].

Pathologic features that increase risk of recurrence include large tumour size, inguinal location, degree of nuclear differentiation, and depth of invasion. Positive surgical margin is a risk factor for early recurrence and distant metastasis [9]. The optimum local and systemic treatment of paratesticular sarcomas in adults includes complete resection, including high ligation of the spermatic cord. The reported local recurrence rate in the scrotum and groin after orchidectomy is 25%-37%. Retroperitoneal lymph node dissection should be limited to patients with only radiologically suspicious lymph nodes [3]. Some authors recommend adjuvant radiotherapy only for high grade tumours or for patients who have a high risk of local recurrence [9]. Currently, use of polychemotherapy (Vincristine, Cyclophosphamide and Doxorubicin) is advocated in high grade tumour subtypes or metastatic disease [9]. Our case highlights the rarity of paratesticular liposarcomas and the diagnostic difficulties which the surgeons encounter, as they mimic testicular tumours.

CONCLUSION

In conclusion, paratesticular liposarcomas represent a rare type of tumours, which are often misdiagnosed pre-operatively, as they mimic testicular tumours. We have reported a rare subtype of paratesticular liposarcoma which occurred at a rare site. Well differentiated liposarcoma must be considered in the differential diagnosis of paratesticular tumours. Our case highlights the importance of an integrated approach, which includes clinical findings, imaging studies and histopathological examination and which includes immunohistochemistry in the definitive diagnosis of paratesticular tumours.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Nov 24, 2013**
Date of Peer Review: **Dec 20, 2013**
Date of Acceptance: **Jan 11, 2014**
Date of Publishing: **Feb 03, 2014**