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Case Report

A rare case of para-testicular dedifferentiated liposarcoma: a diagnostic dilemma

Biren P. Modi, Himanshu U. Patel*

Department of Pathology, Nirali Cancer Hospital, Navsari, Gujarat, India

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***Correspondence:**
Dr. Himanshu U. Patel,
E-mail: himanshupatel831@gmail.com

ABSTRACT

Paratesticular dedifferentiated liposarcoma (DD-LPS) are rare neoplasms. It has been estimated that 12% of DD-LPS occur in the paratesticular region, while the retroperitoneum is the most common site for this tumor. Herein, we presented a case of a 66-year-old male with a history of pedal edema and right scrotal swelling, CT scan shows a solid mass arising from the right side of the scrotum. Following this, a radical orchiectomy of the right testis with mass was performed, and after thorough microscopic and immunohistochemistry positive for MDM2 and CDK4, the diagnosis of low-grade dedifferentiated liposarcoma was given. This case report expands the morphologic spectrum of dedifferentiated liposarcoma and emphasizes the need to consider these tumours in the differential diagnosis of other neoplasms.

Keywords: Paratesticular, Liposarcoma, Spermatic cord, De-differentiated

INTRODUCTION

The most frequent mesenchymal tumour that occurs in the paratesticular region is liposarcoma (40%) followed by leiomyosarcoma, histiocytoma, and rhabdomyosarcoma. Paratesticular liposarcoma (PLS) is an unusual form of liposarcoma that consists of about 12% of all cases. It typically develops in the spermatic cord and then expands to the testicular tunics and epididymis.[1] These tumours are typically misinterpreted as a lipoma, hydrocele, cyst, hernia, or hematocele.[2] Approximately 18% of liposarcomas are dedifferentiated. This tumour mostly occurs in the retroperitoneum; the lower extremities or paratesticular regions are less commonly involved.[2] The head, neck, trunk, and spermatic cord collectively account for less than 20% of DLs, while the subcutis is only seldom affected. Dedifferentiated liposarcoma (DD-LPS) begins to manifest around the same time as atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDL) and peaks in the early seventh decade. Both men and women are equally affected.[3] DD-LPS is an unusual tumour that can emerge in this area, and because of its rarity and morphologic diversity, it can be difficult for surgical pathologists to diagnose.[4] Here we present a case of dedifferentiated liposarcoma of the paratesticular region.

CASE REPORT

A 66-year-old, male having bilateral pedal oedema and right scrotal swelling for 1 month presented to us. On clinical examination, a right-side hard mass was felt in the spermatic cord region. Ultrasonography and CT scan showed approx. 82×65×49 mm^3 sized irregular, heterogeneously enhancing soft tissue lesion the in the right para-testicular region. The lesion shows multiple coarse calcifications, and no evidence of any macroscopic fat was seen within the lesion. The lesion is abutting the right testis. However, the testis was separately visualized from the lesion. Multiple irregulars and nodular enhancing areas and calcifications were seen in the epididymis, separate from the bulk of the primary lesion. A bilateral hydrocele was noted. Routine laboratory examinations

were within normal limits. His vital signs were noted to be stable and consistent intra-operatively and postoperatively. The resected specimen was received in a histopathology laboratory for a frozen section and routine histopathological examination. On gross examination, the Whole specimen measuring 15.0×8.0×7.0 cm^3 in size comprising paratesticular mass, spermatic cord, and testis. A paratesticular mass measures 9.5×7.0×5.5 cm^3 in size, on serial cutting, the cut surface was glistening, solid, bony hard, and greyish to slightly yellowish in colour (Figure 1). Multiple sections were routinely processed and stained with hematoxylin and eosin (H and E). Microscopic examination revealed a diffusely proliferative tumor composed of spindle cells predominantly arranged in sweeping and acute angle cutting fascicles with areas of hyalinization and collagenization (Figure 2). Cells show mild hyperchromasia and mild pleomorphism (Figure 3). Mitosis was inconspicuous and no evidence of lipoblast, necrosis, or haemorrhage. Adjacent para-testicular tissue shows foci of metaplastic bone formation, changes of congestion, fibrosis, oedema, myxoid degeneration, mild inflammation and inspissated urate crystals. Tumour was limited by capsule and testis; vas deference and epididymis were free from the tumor. Immunohistochemistry was performed, tumour cells were immunoreactive for MDM2 (focally), CDK4, p16, SMA and immunonegative for Desmin, CD34, S-100, SOX10, beta-catenin, C-kit, DOG-1, STAT-6 and H-caldesmon. After a thorough histopathological and immunohistochemistry examination, the DD-LPS was given.

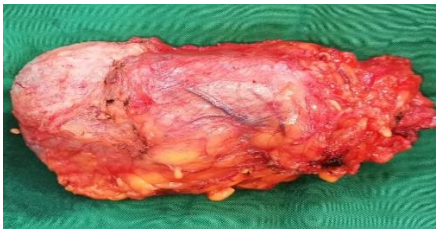

Figure 1: Grossly, paratesticular mass was glistening, solid, bony hard and greyish to slightly yellowish in colour.

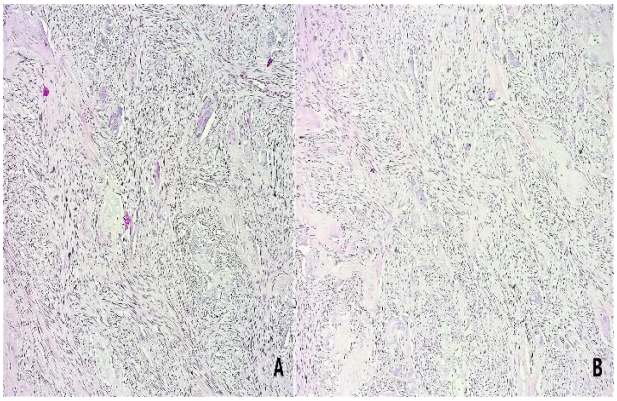

Figure 2: Spindle cells in sweeping and acute angle cutting fascicles with areas of hyalinization and collagenization (H and E, 10X).

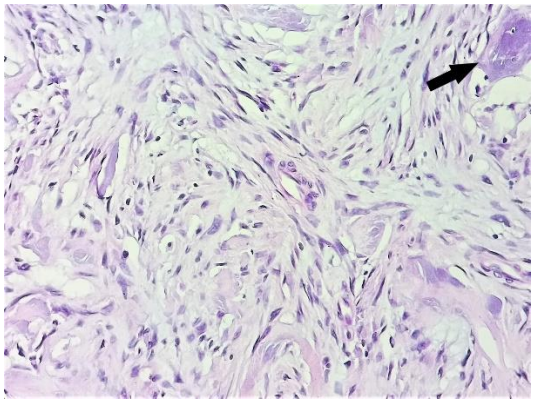

Figure 3: Cells displaying mild hyperchromasia and mild pleomorphism with foci of ossification (black arrow) (H and E, 40X).

DISCUSSION

The spermatic cord accounts for 75% of the origin of paratesticular neoplasms. The vas deferens, smooth muscle, nerves, fibrous tissue, and fat make up the spermatic cord. This anatomic structure seldom develops tumours. They can have a wide range of benign etiologies, however, they very rarely can develop malignant manifestations.

Lipoma and well-differentiated liposarcoma are the most frequent neoplasms that develop from this structure.[4] WHO divides liposarcomas into four subtypes-ALT/WDL, myxoid/round cell, dedifferentiated and pleomorphic liposarcoma.[5] But, it is useful to view liposarcomas into the following three large groups conceptually: (a) atypical lipomatous tumor, or well-differentiated liposarcoma, with or without dedifferentiation (DD-LPS); (b) myxoid and round cell and/or cellular myxoid spectrum; and (c) pleomorphic liposarcoma.[6] While the majority of DD-LPS exhibit high-grade dedifferentiation, a small number of cases have either contain only low-grade areas or both low- and high-grade areas.[3]

Microscopically, low-grade DD-LPS has a less variable morphology and is typically made up of an even proliferation of atypical spindle cells arranged in fascicles having mild to moderate pleomorphism with cellularity intermediate between that of WD-sclerosing liposarcoma and that of high-grade LPS. DD-LPS has minimal to no mitotic activity and an abundance of stromal collagen.[4,7] The most common morphologies of the dedifferentiated element in DD-LPS are fibroblastic and pleomorphic patterns. Leiomyosarcoma, osteosarcoma, and rhabdomyosarcoma are the three most commonly seen divergent differences.[8]

Myogenic, osteo/chondrosarcomatous, or angiosarcomatous elements, as well as meningothelial-like whorling and metaplastic bone formation, have all been reported as additional components of heterologous

differentiation which may occur in around 5-10% of cases.[9-11] Paratesticular liposarcomas are shown as heterogeneous, solid, and hypoechoic lesions on ultrasonography. Any painless echogenic tumour with diverse ultrasonic architecture and only moderate vascularity should raise liposarcoma concerns. The more precise CT and MRI scans can differentiate between areas having fatty and soft tissue components.[2]

MDM2 and CDK4 immunohistochemistry is very accurate and useful in the diagnosis of DD-LPS. In cases when a WDL component is absent, fluorescence in situ hybridization for amplification of MDM2 could be utilized to further validate the diagnosis. The cytogenetic study suggests that MDM2, TP53, CDK4, and chromosomal translocations (12q14-15) were equally positive for WDL. It is important to consider that the dedifferentiated component may also express other markers, especially those associated with myogenic differentiation.[1,2]

The objective of surgical intervention is complete excision with negative margins, and radiation and chemotherapy should be used when required. Only a small number of patients have systemic disease, and local recurrence is common.[4] Compared to well-differentiated liposarcomas (WDLPS), DD-LPS have a higher propensity to metastasis to distant sites, a higher rate of local recurrence, and a higher risk of death.[2]

CONCLUSION

DD-LPS are a relatively uncommon type of tumour. The diagnosis can be difficult, however a thorough histological analysis and immunohistochemistry for MDM2 and CDK4 aid in providing a reliable conclusion. Radial orchiectomy, with wide local excision and negative margins for overall survival, is the main surgical procedure. Due to the high chance of local recurrence and distant metastases, multimodality therapy and long-term follow-up are advised. This particular case emphasises the widespread distribution of these tumours and distinguishes them from other types.

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