Paratesticular Dedifferentiated Liposarcoma with Rhabdomyoblastic Differentiation: A Case Report and Review of the Literature

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Abstract: Liposarcomas of the paratesticular tissue is a rare pathological entity. The symptoms are similar to inguinal hernias or hydroceles. We present the case of an 84-year-old man with a rare paratesticular liposarcoma that manifested as painless right hemiscrotal swelling. Testicular tumour markers were negative. Imaging revealed a heterogeneous mass with a fat component. He underwent a radical orchiectomy on the left side to remove the associated mass. This revealed dedifferentiated liposarcoma (DDLS) with rhabdomyoblastic differentiation and MDM2 amplification. The surgical margins were negative, and the patient had a metastatic workup that included magnetic resonance imaging (MRI) of the abdomen and pelvis. Because of the disease's rarity, there is no clear agreement on radiotherapy and chemotherapy roles.

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Introduction

Most paratesticular solid lesions in adults are benign, although malignant tumours can be seen in 3% of cases (Rafailidis et al., 2021). The most common types of malign paratesticular tumours are rhabdomyosarcoma and liposarcoma (Cardenosa et al., 1990). A more common form of paratesticular liposarcoma is well-differentiated and dedifferentiated types (Li et al., 2022). Here, we present the clinical history and management of a case with primary paratesticular dedifferentiated liposarcoma in light of the literature.

Case report

An 84-year-old man was referred to our urology outpatient clinic in May 2022 after complaining of swelling in his left scrotum for two weeks. A painless, slow-growing, fixed mass in the left scrotum was not accompanied by any obvious promoting or alleviating factors. There are no additional symptoms or signs. According to the patient, there were no discernible personal conditions related to the current clinical manifestation. There was also no evidence of family history. The only positive finding on physical examination was a rigid mass in the left scrotum, about 5 cm in diameter (Figure 1).

The laboratory examinations, including testicular tumour markers, reveal no specific abnormalities (hemogram, urinalysis, ESR – erythrocyte sedimentation rate, brucella agglutination test, liver, and kidney function tests, and chest X-ray).

Scrotal ultrasound revealed a large left scrotal heterogeneous mass ultrasounds-predominantly hyperechoic in echotexture; however, these findings were not specific. However, contrast-enhanced magnetic resonance imaging (MRI) revealed

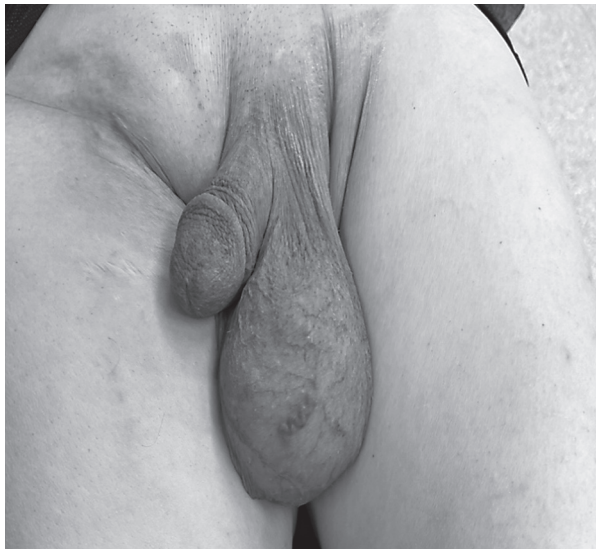

Figure 1 – Physical examination. A left scrotal mass measuring 6 cm was noted.

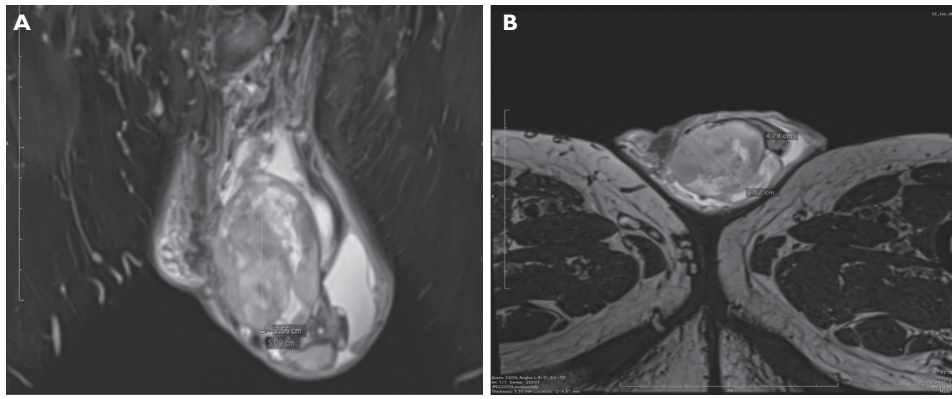

Figure 2 – Sagittal and transverse magnetic resonance images.

a 48×38×50 mm nonhomogeneous space-occupying lesion of the left testis. There was no lymphadenopathy or distant metastasis found (Figure 2).

Following the examinations, the patient underwent a radical left orchidectomy with wide local excision that included the paratesticular mass as well as the left testicle and all left inguinal canal contents up to the deep inguinal ring while sparing the left ilioinguinal nerve. A 5×4×6 cm enlarged left testicle was removed from the scrotum (Figure 3). There is no visible inflammatory adhesion to the organs.

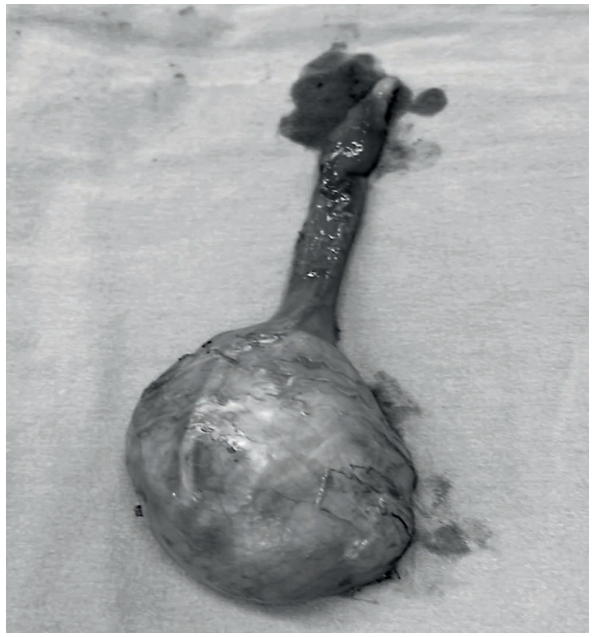

Figure 3 – Testis and cord after orchidectomy.

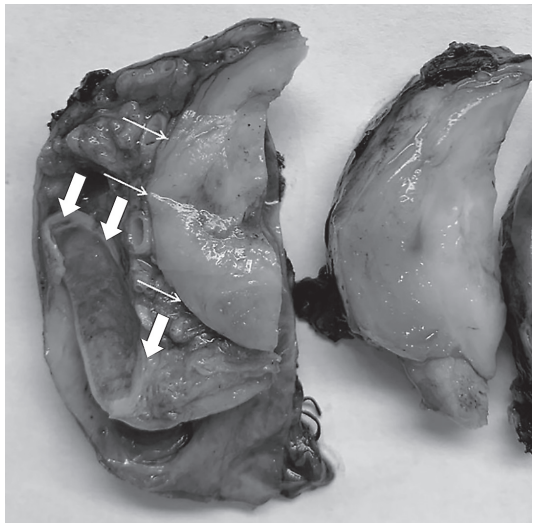

Figure 4 – Gross appearance of dedifferentiated liposarcoma (thin arrows) and testis (thick arrows).

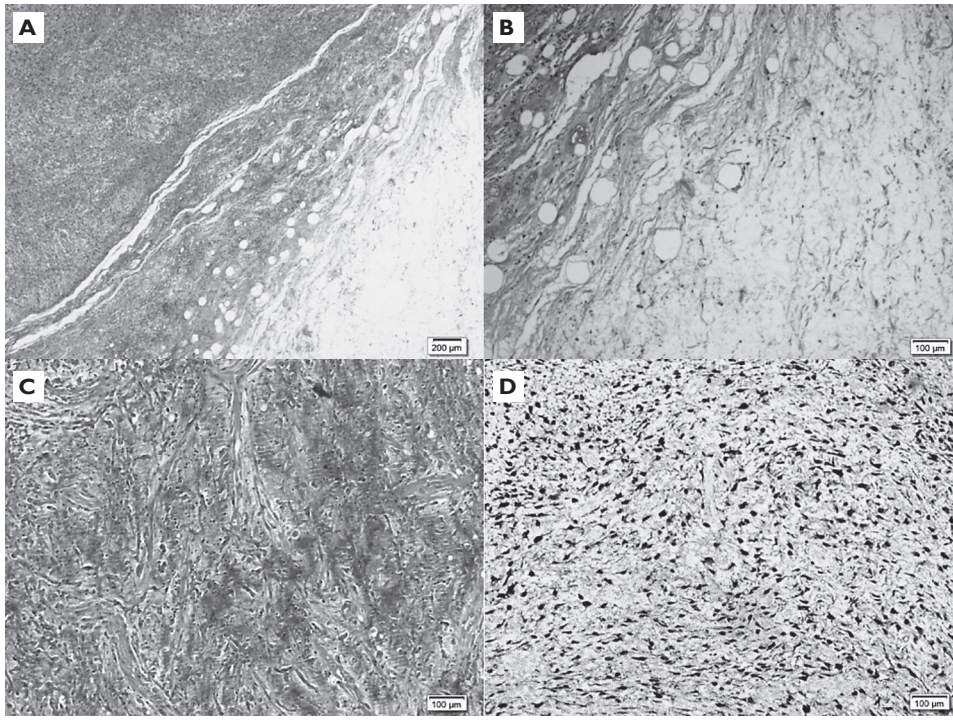

Figure 5 – Histologic and immunohistochemistry analysis of dedifferentiated liposarcoma specimens. (A) The transition zone of well-differentiated to the dedifferentiated tumour (hematoxylin-eosin stain). (B) Atypical lipomatous tumour area (hematoxylin-eosin stain). (C) Areas of liposarcoma on the myxoid floor with more spindle appearance (hematoxylin-eosin stain; 10×). (D) CDK-4 diffuse positive staining supports the diagnosis of dedifferentiated liposarcoma (immunohistochemistry).

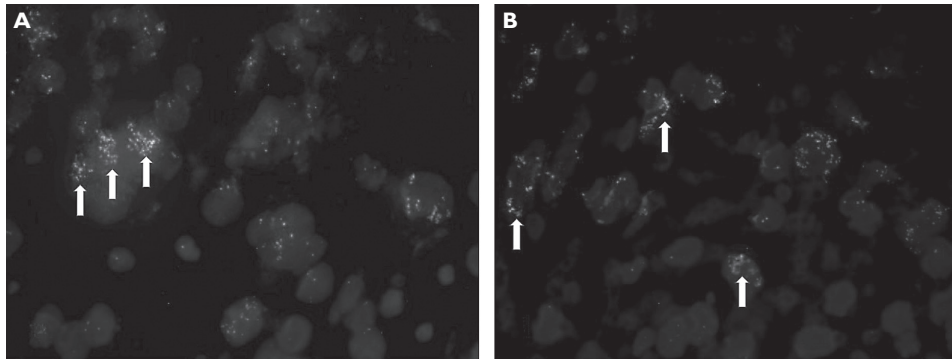

Figure 6 – Fluorescence in situ hybridization (FISH) of the CDK-4 (A) and MDM-2 (B) genes. FISH analysis confirmed CDK4 and MDM2 gene amplification in the nuclei of atypical cells (clustering of green signals indicated by arrows).

Histopathology revealed that the tumour was a DDLS (dedifferentiated liposarcoma) with rhabdomyoblastic differentiation, measuring 5×5×6 cm. CDK4(+); MDM2(+); ALK(–) immunohistochemical analysis supported this diagnosis (Figures 4 and 5). Fluorescence in situ hybridization (FISH) analysis showed the presence of CDK4 and MDM2 gene amplification supported the diagnosis of DDLS (Figure 6).

Discussion

Liposarcomas are classified into four types based on their histological appearance: myxoid (the most common; 40%), round cell, well-differentiated (subdivided into lipoma-like, sclerosing, inflammatory, and dedifferentiated), and pleomorphic (Logan et al., 2010). DDLS accounts for 18% of liposarcomas and was first described by Evans in 1979 as a well-differentiated liposarcoma adjacent to a cellular nonlipogenic sarcoma. Dedifferentiated, round cell and pleomorphic liposarcomas are high-grade, aggressive tumours with metastatic potential, whereas well-differentiated and myxoid liposarcomas are low-grade tumours with a more indolent clinical course (Dalal et al., 2006). DDLS can be identified on imaging as a heterogeneous non-lipogenic mass within an area of abnormal-appearing fat.

The diagnosis of DDLS is dependent on histology and immunohistochemistry. CDK4 and MDM2 were significant markers for diagnosing well-differentiated liposarcoma in one investigation (Pănus et al., 2015). FISH is especially beneficial when the presence of a well-differentiated liposarcoma component is unknown (Nishio et al., 2021). MDM2 and CDK4 amplification is helpful for differential diagnosis (Nishio et al., 2015).

Localized paratesticular liposarcoma can be treated with radical orchidectomy with a negative surgical margin. After orchidectomy, nearly one-third of the patients had local persistent lesions (Chiodini et al., 2015). Although some researchers

advocate adjuvant radiation therapy because of increased local control, its routine usage remains controversial (Li et al., 2018). Anthracycline-based therapy is the conventional first-line treatment for advanced DDLS. Trabectedin and eribulin are the two second-line therapeutic alternatives (Nishio et al., 2021). Several other medications were tested for advanced illnesses and with promising results. The prognosis and survival rate vary. Khandekar et al. (2013) reported a recurrence-free survival rate of 76% at 3 years and 67% at 5 years. Another study showed a 5-year survival rate of 75% and a recurrence rate of 50–70%. Large tumour size (>5 cm), pathologic degree of nuclear differentiation, and depth of invasion are the risk factors for recurrence (Schoonjans et al., 2016).

Conclusion

Paratesticular DDLS is a rare tumour with a painless scrotal mass. Radical orchidectomy with wide excision and high ligation is the standard treatment for localized disease. Although the prognosis is favourable, long-term follow-up is needed because the probability of recurrence is high. We must continue to examine the molecular mechanisms underlying liposarcomagenesis and work toward the development of novel therapeutic strategies for liposarcoma patients.

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