Undifferentiated Pleomorphic Sarcoma with Focal Myogenic Differentiation. A Rare Case of Testicular Sarcoma

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Abstract

Undifferentiated pleomorphic sarcoma (UPS) of the testis is an extremely rare entity worldwide. To the best of our literature, this is the first case from Oman. Here in, We, present a case of UPS in a 68-year-elderly male who presented with a left testicular solid mass and underwent left radical orchiectomy was performed. The Histopathology shows undifferentiated pleomorphic sarcoma with focal myogenic differentiation. The patient followed up after 6 months and one year, there was no recurrence of diseases. Surgical management with radical orchiectomy is the preferred treatment, but the role of postoperative radiotherapy and chemotherapy remains unclear.

Keywords: Testis, Undifferentiated, Pleomorphic, Sarcoma, Case report

Introduction

Undifferentiated polymorphic sarcoma (UPS), which is known as malignant fibrous histiocytoma (MFH), is one of the most common soft tissue sarcomas of deep tissues in old age,[1] and MFH is a subtype of rare malignant soft tissue sarcoma of Testis, especially in mid and late age. (MFH) was first reported by O'Brien and Stout in 1964.[2] Authors1 understand that the MFH previously developed as pleomorphic variants of liposarcoma, fibrosarcoma, and rhabdomyosarcoma as previous examples of MFH. The extremities are the most affected sites with undifferentiated high-grade pleomorphic sarcomas, even more than the trunk or retroperitoneum; however, incidents in the gastrointestinal tract are sporadic.[1,3-5] UPS resists a worse prognosis than other types of MFH.[6] Myxofibrosarcoma (MFS), histologically similar to UPS, was separated from UPS and reclassified as a single entity in 2002 due to its clinical pathology.[7] However, comprehensive and integrated genomics shows that UPS and MFS are largely indistinguishable in a cross-platform molecular context.[8] UPS/MFS, characterized by great genetic complexity, has always been a headache in clinical practice. Thorough knowledge of the pathogenesis of UPS/MFS is lacking and precise diagnostic features and treatment strategies are required. To better understand UPS/MFS, advances are needed in the identification of abnormal signals associated with sarcoma origin, advances in conventional management, and the development of targeted therapy and therapy. Immunotherapy, as well as new therapies with promising future management, and the development of targeted therapy and therapy. Immunotherapy, as well as new therapies with a promising future. We report a case of a rare entity of testicular undifferentiated pleomorphic sarcoma and discuss its management.

Case Report

A 68-year-old male, married, non-smoker, with a past surgical history of coronary artery bypass grafting 10 years back and left hydrocelectomy in another hospital one year ago. He presented to our urology outpatient department in March 2023, with presenting complaints of left testicular swelling and mild pain for five months. According to the patient, this swelling was smaller initially and has progressively increased in size. On examination left testis was palpable with a craggy feeling of large epidydimal swelling, no local sign of inflammation, and no hernia. A contrast-enhanced computed tomography (CT) of the abdomen and pelvis was done outside and reported as a heterogeneous left para-testicular mass with no inguinal or para-aortic lymphadenopathy (Fig 1). Magnetic resonance imaging (MRI) and showed a heterogenous 5.1 x 5.2 cm, left testicular mass (Fig 1). The mass appears heterogeneous with foci of high T1 signal and shows high T2 signal and foci of diffusion restriction. The mass is likely arising from the epididymis. Significantly increased vascularity within the spermatic cord extending from the inguinal region down to the mass and epididymis. There is no obvious invasion of the scrotal wall. The right testicle appears normal. There are no significant inguinal lymph nodes. No significant pelvic or retroperitoneal lymphadenopathy and no lung pathology were identified. All tumor markers (Alpha-fetoprotein, Beta HCG, and LDH were within normal ranges. So, based on clinical and radiological findings, he underwent a left radical inguinal Orchiectomy. Histopathology reported as; The whole specimen is inked blue. On slicing, the testis is markedly atrophic and measures 3.5x2.2x3 cm. Epididymis is replaced by a well-circumscribed mass that has a necrotic yellow and hemorrhagic surface. The mass measures 5.5x 5.4. 4 cm. A gross pathological examination of the orchiectomy revealed a 5.5 cm × 5.4 cm × 3 cm well-delineated tumor with a tan solid surface arising from the epididymis (Figure 2). Microscopically epididymal mass shows a malignant spindle cell neoplasm exhibiting fascicles of high-grade tumor cells with extensive tumor necrosis. The tumor cells have enlarged hyperchromatic nuclei with prominent nucleoli and spindly cytoplasm. The mitotic rate is variable but measures up to 25 mitotic figures per 10 high-power fields. Marked cytological atypia are present, with tumor-type giant cells. The French Federation of Cancer Centers Sarcoma Group grading system score was 8 out of 8 (Sum of 3 for degree of differentiation, 2 for necrosis, and 3 for No lymphovascular space invasion). Figure 3 Hematoxylin and Eosin stain at x10 magnification showing highly pleomorphic tumor cells arranged in to storiform pattern. The immunohistochemistry stains (Figure 4) show, focal positivity for desmin, Caldesmon, and smooth muscle actin and are negative. So, in conclusion, the report mentioned that the morphology and immunophenotype findings are in keeping with undifferentiated pleomorphic sarcoma with focal myogenic differentiation., Grade (FNCLCC): 3. Pathological stage (TNM 8th edition): pT2, grade 3 (according to National Federation of French Cancer Centers and National Cancer Institute system). The patient followed up after six months and one year and was free from any recurrence or metastasis of the disease.

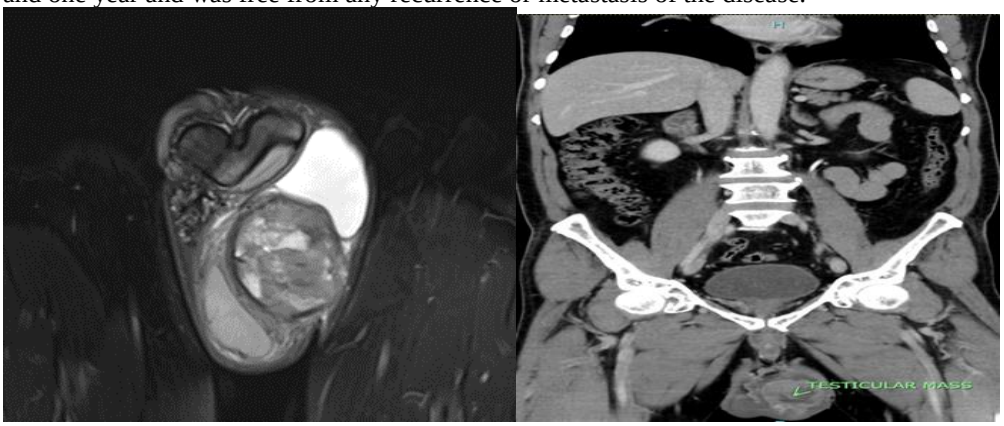

Figure 1: MRI & CT SCAN showing left testicular solid heterogenous mass.

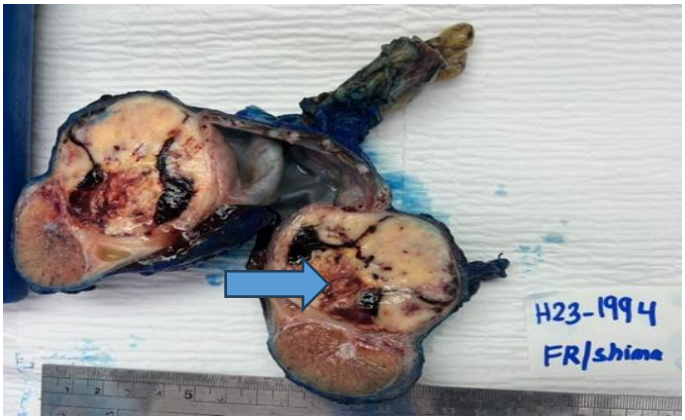

Figure 2: A gross pathological examination of the orchiectomy revealed a 5.5 cm × 5.4 cm × 3 cm well- delineated tumor with a tan solid surface arising from the epididymis

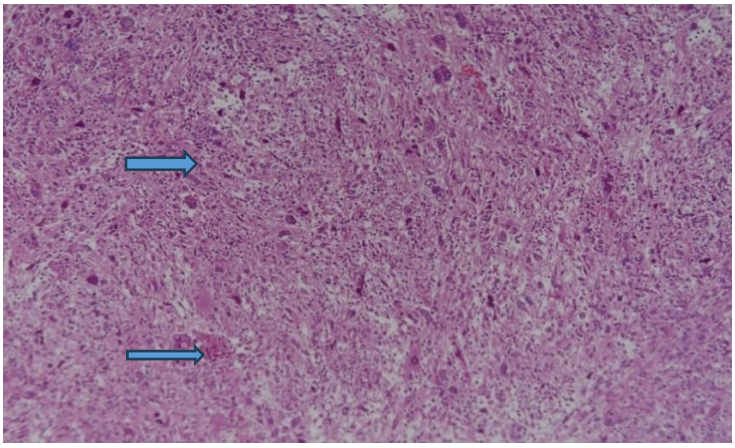

Figure 3: Hematoxylin and Eosin stain at x10 magnification showing highly pleomorphic tumor cells arranged in to storiform pattern.

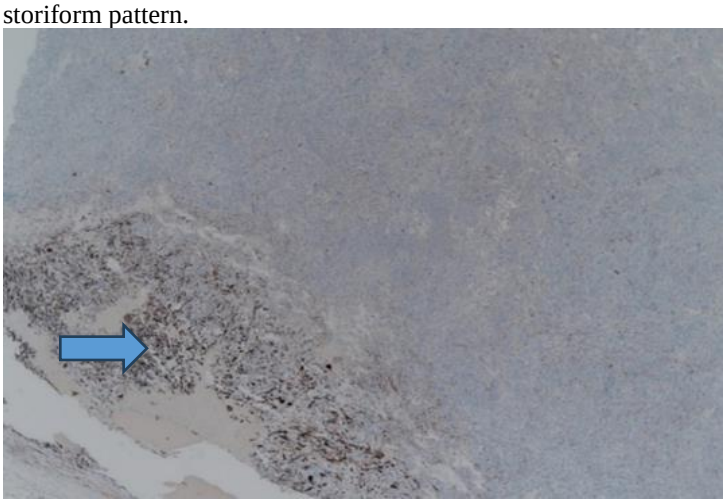

Figure 4: Tumor is focally positive for Desmin immunohistochemistry stain.

Discussion

Testicular tumors, which make up 1% to 2% of testicular tumors, are a relatively uncommon type of cancer. Rare diseases include testicular MFH. To the best of our knowledge, four cases of testicular MFH were documented in the literature; one case was in English and the other two were in China, and the patients' ages ranged from 56 to 78.[5] Literature shows a case of an MFH in which the patient presented with a painless mass, more over another study of a patient with soft tissue sarcoma complained of mild pain at the site of tumor.[9] However, our patient, the case presented with left testicular swelling and mild pain for the last 5 months. Studies have reported that the age between 14 to 78 years at the initial presentation with giant malignant fibrous histiocytoma of the testis, in our patient was 68 years old.[5,6,10-13] Diagnosis of undifferentiated pleomorphic sarcoma (UPS) is challenging and can be recognized on CT scans and MRI. The final diagnosis is usually a diagnosis of exclusion confirmed by histological study and post-operative examination.[6,9] For our patient, a contrast-enhanced computed tomography (CT) of the abdomen and pelvis was done outside and reported as a heterogeneous left para-testicular mass with no inguinal or para-aortic lymphadenopathy (Fig 1). Magnetic resonance imaging (MRI) and showed a heterogenous 5.1 x 5.2 cm, left testicular mass (Fig 1). The mass appears heterogeneous with foci of high T1 signal and shows high T2 signal and foci of diffusion restriction. No metastasis in the lungs was identified. The treatment of sarcoma is surgical resection of the mass with or without radiotherapy.[6,9] As in our case, the treatment was left radical inguinal Orchiectomy without radiotherapy as the resection margin was negative. Literature has revealed, the same microscopic examination findings of pleomorphic cells with multi-nucleated giant cells, as in our case.[6,9,14] The immunohistochemical staining, is important to review for smooth muscle actin, desmin, and Caldesmon, in our patient these immunohistochemical findings were positive.[6,9] Moreover, our case does not mention the Ki-67, while other cases revealed a high expression of Ki-67.[5,8,13] Authors13 have reported that radiotherapy in addition to surgical treatment, reduces the chances of local recurrence, and Nascimento AF et al, have observed the positive response of 55% and 66% with the use of adjuvant chemotherapy.[15] There are certain reasons associated with metastasis and survival, and these factors are tumor depth, size, grade, necrosis, and local recurrence. For example, length and intensity look like covariables due to the fact big tumors tend to be deep-seated tumors. In the multivariate analysis by Engellau et al.,[16] necrosis and local recurrence were significant predictors of metastasis within the first 2 years of diagnosis and throughout a longitudinal follow-up period, while tumor intensity and recurrence locally, have been widespread predictors past 2 years.

Conclusion

In conclusion, Undifferentiated polymorphic sarcoma (UPS) is classified as a rare testicular tumor and the first line of management is radical orchiectomy, followed by adjuvant chemotherapy is advisable to improve survival, if indicated.

Disclosure

The authors have not received any funding for this case. We, could not find any conflict of interest for these case series research, authorship, or publication of this article.

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