Case Report

Pleomorphic sarcoma, not otherwise specified, of scrotum mimicking Fournier's gangrene

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ABSTRACT

Pleomorphic sarcoma, not otherwise specified/undifferentiated pleomorphic sarcoma (UPS) is one of the most common sarcomas occurring between the ages of 50 and 70 years. We present a case of a 50-year-old male with a paratesticular mass clinically diagnosed as Fournier's gangrene due to ulceration and necrosis of the overlying scrotal skin. Histopathological examination finally led to the diagnosis of UPS.

Key words: Pleomorphic sarcoma, sarcoma, scrotal sarcoma

INTRODUCTION

Pleomorphic sarcoma, not otherwise specified/ undifferentiated pleomorphic sarcoma (UPS), earlier known as malignant fibrous histiocytoma (MFH), is a common soft-tissue tumor in adults. The most frequent primary sites include the lower (49%) and upper (19%) extremities, and the retroperitoneum and abdomen (16%). However, paratesticular/scrotal UPS is a rare entity.[1,2] It constitutes approximately 13% of all malignant tumors in the paratesticular region. More than 75% of the tumors in this region arise from the spermatic cord and are mostly benign.[3] We report a case of UPS of the scrotum in a 50-year-old man.

CASE REPORT

A 50-year-old male presented with a gradually increasing left scrotal mass since 10 months. The swelling was associated with pain. The patient did not give any history of loss of weight or appetite. On examination, a firm, nodular, ulcerative mass was found in the scrotum with sloughing, ulceration, and necrosis of the scrotal skin. Inguinal lymph nodes were not palpable. With a clinical diagnosis of Fournier's gangrene, debridement of the necrotic tissue was done and sent for histopathological examination. On gross examination grey, white, and necrotic soft -tissue pieces were found. Histopathological examination revealed a tumor arranged as fascicles, storiform pattern, and in sheets. Tumor cells showed marked nuclear pleomorphism with a round to oval to spindled nuclei, vesicular nuclear chromatin, and a moderate amount of eosinophilic cytoplasm. Brisk mitotic activity (with more than 10 mitoses/10 hpfs) was seen. Areas of necrosis were prominent [Figures 1 and 2]. A diagnosis of grade 3 sarcoma (FNCLCC grading system) was thus made and immunohistochemistry was performed. Immunohistochemical examination showed diffuse positivity for vimentin and CD68 in tumor cells [Figures 3 and 4]. Tumor cells were negative for smooth muscle actin, desmin, cytokeratin, leukocyte common antigen, S100, and HMB 45. Thus, a final diagnosis of undifferentiated high-grade pleomorphic sarcoma was rendered. The patient then underwent a radical orchiectomy and had been referred for a postoperative chemoradiation.

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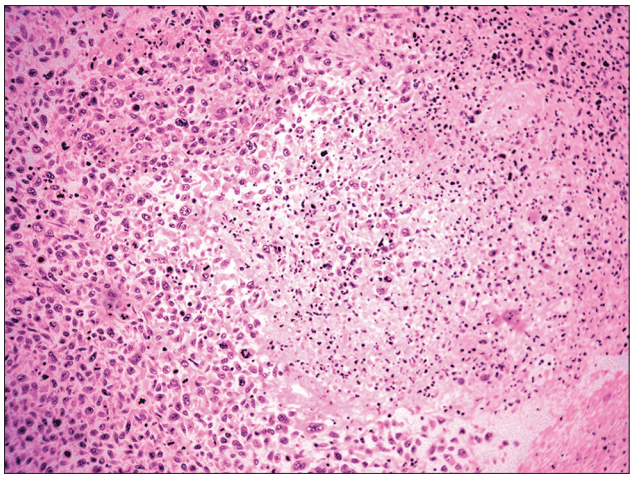

Figure 1: Tumor cells showing marked nuclear pleomorphism, hyperchromatic nuclei and areas of necrosis. (H and E, ×100)

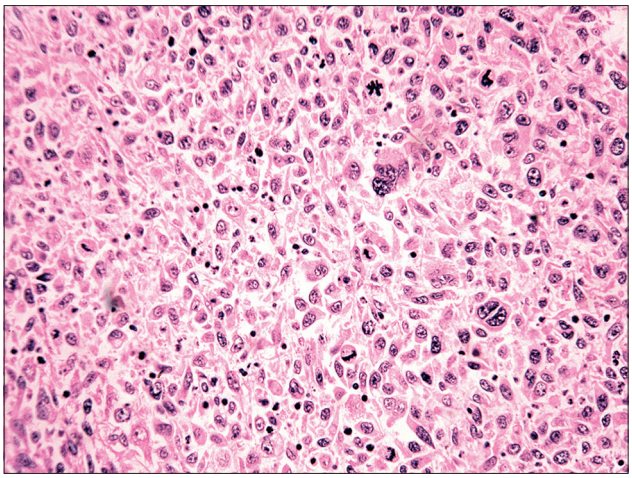

Figure 2: Photomicrograph showing tumor giant cells and atypical mitotic figures. (H and E, ×400)

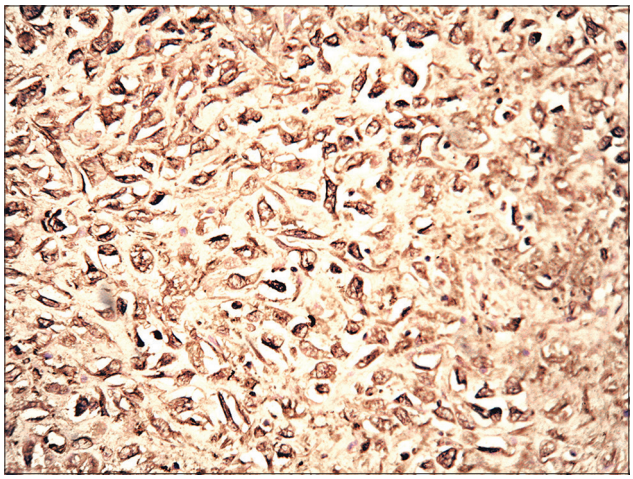

Figure 3: Tumor cells showing diffuse cytoplasmic positivity for vimentin. (×400)

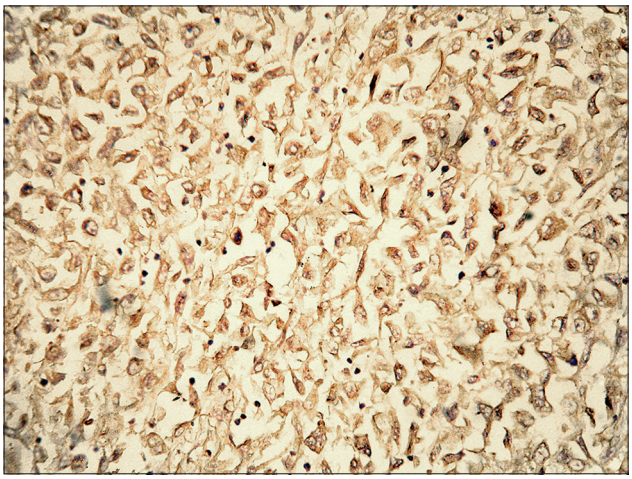

Figure 4: Tumor cells showing positivity for CD68. (×400)

DISCUSSION

Pleomorphic sarcoma, not otherwise specified/UPS is one of the most common sarcomas occurring between the ages of 50 and 70 years. The sites of predilection include the lower extremities and retroperitoneum. UPS of the spermatic cord and the paratesticular area is rare.[1,2] Preoperative diagnosis of spermatic cord MFH/UPS is challenging, as the origin and nature of an inguinal mass cannot be precisely determined by clinical presentation and physical examination. Most tumors occur as a painless, slowly growing, localized solitary mass, although some cases may present as multiple satellite tumor nodules, with the involvement of scrotal skin in rare cases.[1,4] The tumor is often misdiagnosed as a benign condition such as a hernia, spermatocele or hydrocele. Our patient, however, presented with a paratesticular mass with ulceration and necrosis of overlying scrotal skin leading to a diagnosis of Fournier's gangrene clinically. An extensive search of the literature in PubMed database revealed only 22 cases of pleomorphic sarcoma of the scrotum and only one case with the clinical presentation as Fournier's gangrene.[5]

Fournier's gangrene is a rare, life-threatening necrotizing fasciitis that usually involves the perineal or genital areas and may extend up to the abdominal wall. The organisms usually implicated are hemolytic streptococci, which cause fulminating inflammation of the subcutaneous tissues resulting in obliterative arteritis of the arterioles to the scrotal skin. Although many malignancies including squamous cell carcinoma, colorectal carcinoma, lymphoma, and multiple myeloma have been known to be associated with Fournier's gangrene, its occurrence in a case of scrotal sarcoma has rarely been mentioned in the literature.[6]

Important differential diagnoses need to be considered at this site before reaching a diagnosis of UPS. Unlike testicular masses, paratesticular masses are usually benign. They may be either solid or cystic. Cystic masses include hydrocoele, epididymal cyst, and varicocoele. The most common

cause of abscess in the scrotal region is epididymitis or epididymo-orchitis. Most of the solid masses are also benign, with the prevalence of malignancy being only 3%.[7] The most common paratesticular neoplasms are spermatic cord lipomas and adenomatoid tumors of the epididymis. Malignant tumors of the paratesticular region can have mesenchymal, epithelial, germ cell, mesothelial, and lymphoid origin.[1] Spermatic cord is the most common site of genitourinary sarcomas, which include leiomyosarcoma (32%), rhabdomyosarcoma (24%), liposarcoma (20%), pleomorphic sarcoma, not otherwise specified (13%).[7-9]

Ultrasonography is the primary modality for imaging scrotal lesions. It provides excellent spatial resolution and has been shown to be nearly 100% sensitive in distinguishing testicular and paratesticular pathologies. Magnetic resonance imaging further helps in the establishment of a more specific diagnosis.[1,9]

Morphological variants of spermatic cord MFH/UPS include the pleomorphic (83%), giant-cell (9%) and inflammatory (6%) and, very rarely, myxoid subtypes.[1] The French Federation of Cancer Centers Sarcoma Group grading system is used for the grading of sarcomas. In this system, the grade is obtained by scoring the degree of tumor differentiation, mitotic count, and the amount of tumor necrosis.[2] In our case, the score of differentiation was three, the score of necrosis one, and the score of mitotic activity three, giving a total score of seven. Differentiation of UPS from other malignant tumors that exhibit a comparable degree of cellular pleomorphism is of particular importance. The differential diagnosis thus includes anaplastic carcinoma, malignant melanoma, and pleomorphic forms of liposarcoma, leiomyosarcoma, and rhabdomyosarcoma. Careful examination of the histopathological features and immunohistochemical profile helps in reaching a definitive diagnosis.

A diagnosis of liposarcoma can be made by the identification of typical multiloculated lipoblasts and areas of lipomatous differentiation. Dedifferentiated liposarcomas can have areas showing high-grade sarcoma similar or identical to MFH. In difficult cases, in which lipoblasts are very rare, the cells can be identified by positive immunostaining for S100 or leptin.[2,9] In rhabdomyosarcoma, pleomorphic cells with round to elongated nuclei, and abundant eosinophilic cytoplasm are seen. Areas of necrosis are common. Immunohistochemistry is necessary to demonstrate cytoplasmic positivity of desmin and nuclear positivity for myogenin.[10] The epithelial and biphasic types of mesothelioma can be excluded by the lack of epithelial cell features and the absence of keratin immunostaining. Sarcomatous mesotheliomas can have an immunophenotype similar to MFH and may be a diagnostic challenge. In difficult cases, ultrastructural studies and additional immunostains may be helpful.[2]

Being a rare entity, the management of UPS of the scrotum has not been well-defined in the literature. All paratesticular sarcomas in adults are managed with complete resection, including high ligation of the spermatic cord.[11] Radical inguinal orchiectomy, along with a wide *en bloc* resection of adjacent soft-tissue, has been used for spermatic MFH/UPS. Various studies have underscored the importance of adjuvant radiation therapy to prevent recurrence of intrascrotal sarcoma.[12]

Tumor size over 5 cm has been found to be an important prognostic factor.[13,14] In a review of 22 cases of locally recurrent MFH/UPS, Froehner *et al*. found the prognosis for these patients to be extremely poor. Only two of 22 patients survived past 3.5 years of follow-up.[15] Prognosis of these sarcomas depends on tumor size, grade, stage, histologic type, and lymph node involvement.

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Conflicts of interest
There are no conflicts of interest.

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