Case Report

Abstract Page

Title of the article: Pleomorphic sarcoma in paratesticular region

[Abstract]:Primary tumors of the paratesticular region are rare, with paratesticular sarcomas constituting a major proportion of these tumors, particularly in the elderly. The paratesticular region consists of mesothelial, various epithelial and mesenchymal cells and may therefore give rise to a number of tumors with various behaviors.[1]We present a case of a 63-year-old male with a paratesticular mass clinically diagnosed as Epididymal cyst. Histopathological examination finally led to the diagnosis of Undifferentiated pleomorphic sarcoma. We report this case due to its rarity and due to the limited nature of data and definitional variations.

[Key-words]:Pleomorphic sarcoma, sarcoma, paratestis.

[Key Message]: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.

Defining the association between the paratesticular mass and the testicle, and differentiation between benign and malignant masses using radiology is challenging, therefore the mass is usually considered to be malignant and radical orchiectomy with high ligation is performed.

[Introduction]

As the paratesticular region contains various structures, including the epididymis, spermatic cord, tunica vaginalis and strong fat-ligament-muscle supporting tissues, it may give rise to a number of tumor types with various behaviors [1]. Tumor variability may also be due to the Wolffian duct origin of the testis appendages, including the spermatic cord. The most significant feature of the paratesticular region is that it is the origin of a small number of tumors with rich diversity.

The majority of the masses within the scrotum in adults are of testicular origin. Paratesticular masses account for 2-3% and sarcomas account for ~30% of all scrotal masses . The most common type of sarcoma is liposarcoma, followed by leiomyosarcoma (LMS), rhabdomyosarcoma (RMS), undifferentiated pleomorphic sarcoma and fibrosarcoma. To the best of our knowledge, only one instance of low-grade fibromyxoid sarcoma in a paratesticular location has been previously reported. 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 63-year-old man.The prognosis is often poor, as recurrence and metastasis are common, and the mechanism and outcome of regional lymph node resection, radiotherapy and chemotherapy is unclear.We report this case due to its rarity and due to the limited nature of data and definitional variations.

[Case History]

A 63 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, mass was found in the scrotum with normal appearing scrotal skin. Inguinal lymph nodes were not palpable. With a clinical diagnosis of Epididymalcyst,scrotal exploration was done andbiopsy specimen sent for histopathological examination. On gross examination solid grey white, soft tissue were found. Histopathological examination revealed a tumor arranged in sheets. Tumor cells showed marked nuclear pleomorphism with a round to oval to spindled nuclei, with moderate amount of eosinophilic cytoplasm,pleomorphic nuclei and vesicular chromatin. Brisk mitotic activity (with more than 10 mitoses/10 hpfs) was seen . A diagnosis of Pleomorphic sarcoma (FNCLCC grading system) was thus made and immunohistochemistry was performed. Immunohistochemical examination showed diffuse strongpositivity for vimentin in tumor cells. Tumor cells were negative for smooth muscle actin, desmin, cytokeratin, CD 34,and S100. 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.

Lab findings:

Lab findings showed a haemoglobin of 12.1gm/dl , white blood cell count of 9,300/mm^3 with predominantly neutrophils. ESR was 45 mm/hr, Platelet count 2.06 lakhs. All other investigations were within normal limits.

USG scrotum-

-Thickened right spermatic cord with three hypoechoic well encapsulated collections with low level internal echoes within.

-Bilateral Grade II varicocele.

-Right inguinal hernia.

. We received high inguinal orchidectomyspecimen showing an encapsulated mass in the paratesticular region.Mass measures 8x5.5x4 cm and cut surface is solid, grey white, fleshy with yellowish areas and tiny cystic spaces.Adjacent testis showed normal morphology.

Histologically, section showed a neoplasm composed of cells arranged haphazardously.Cells showed marked pleomorphism with high N/C ratio, pleomporphic vesicular nucleus with clumped chromatin.Few spindly and stellate cells seen. Many binucleate and multinucleate cells with numerous mitotic figures (0-2/ HPF) noted.Tumour cells showed diffuse and strong positivity for Vimentin and were negative for smooth muscle actin (SMA), S100, Desmin, CD34.

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. In paratesticular region, Liposarcoma and LMS are the most common sarcomas[5]. RMS is more frequent in younger patients[6].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.

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 adenomatoidtumors 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 otherwisespecified(13%).[7],[8],[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 tumornecrosis[2]. 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

All paratesticular sarcomas in adults are managed with complete resection, including high ligation of the spermatic cord.[1] 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.[4]

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.[5] Prognosis of these sarcomas depends on tumor size, grade, stage, histologic type, and lymph node involvement.

In conclusion, the paratesticular region consists of complex structures that can develop various neoplastic formations and patterns. Sarcomas comprise a significant part of paratesticular masses and may exhibit an aggressive clinical course. In older patients, paratesticular sarcomas must be considered for the differential diagnosis of scrotal masses, which do not exhibit a clear association with the testes.Furthermore, clinicians and patients must be informed about the high probability of local recurrence and distant metastasis in paratesticular sarcomas.

[References]

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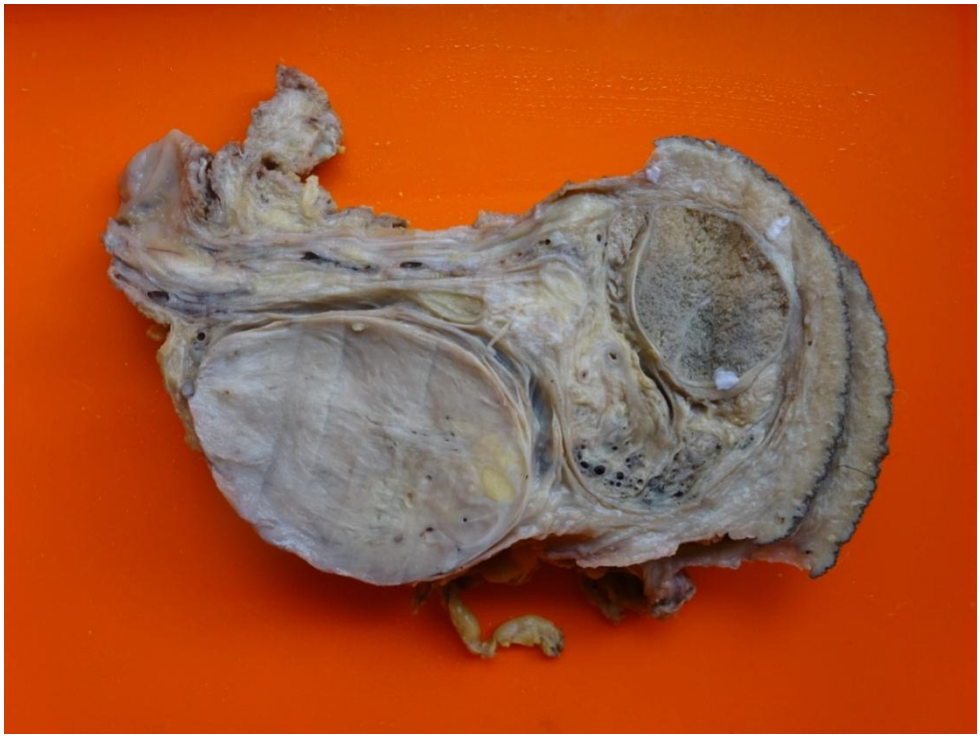
Figures



. **Figure 1**.Gross photograph showing an encapsulaed mass in the paratesticular region, which is greywhite , firm with yellowish areas.(original image)

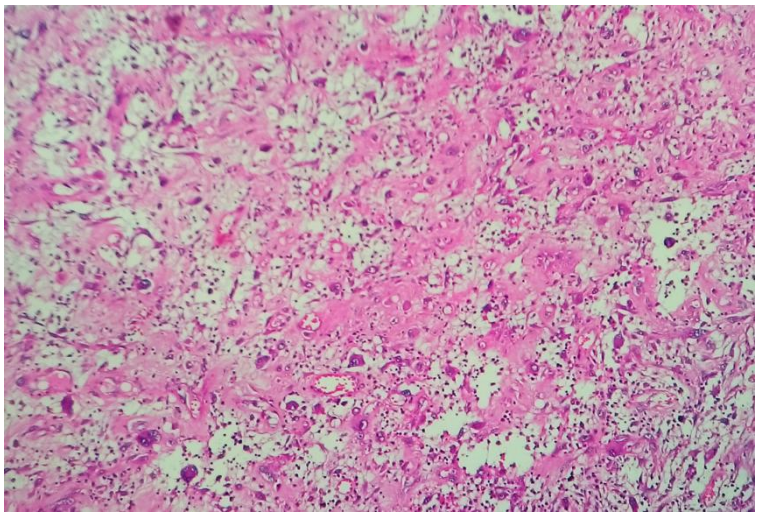


Figure 2: shows cells arranged haphazardously with pleomorphic vesicular nuclei with clumped chromatin

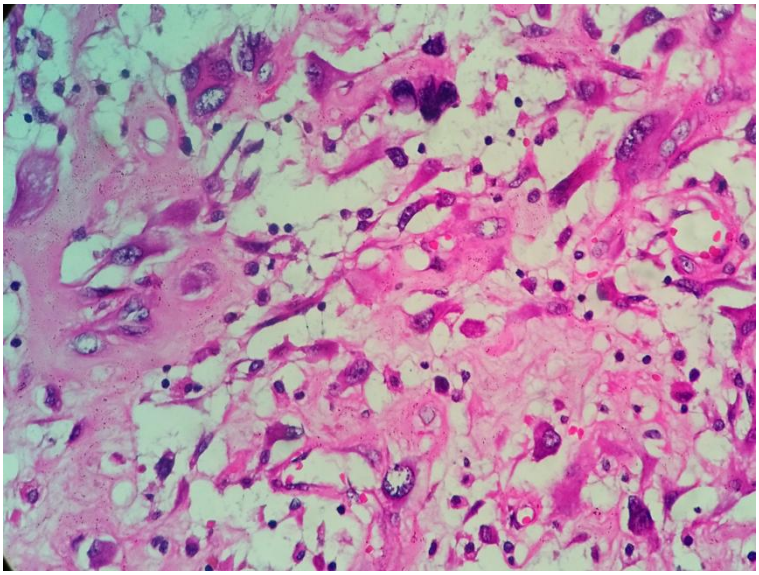


Figure 3: High power view showing pleomorphic cells and spindle cells

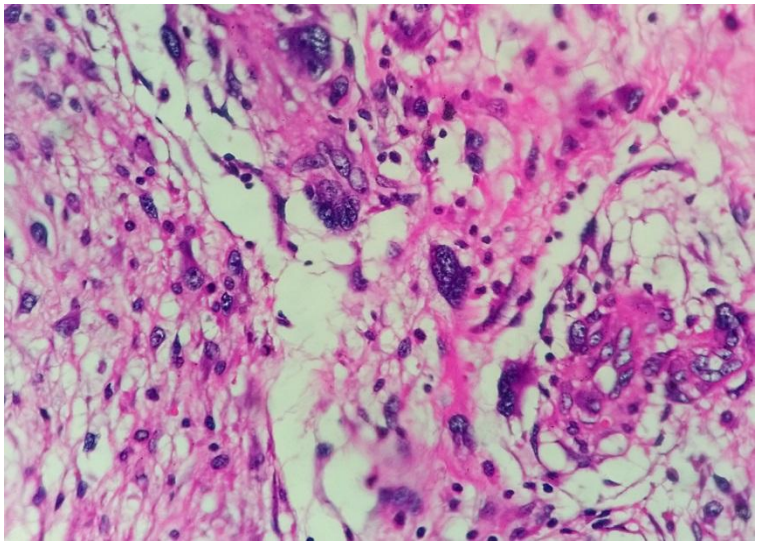


Figure 4: High power view showing pleomorphic cells

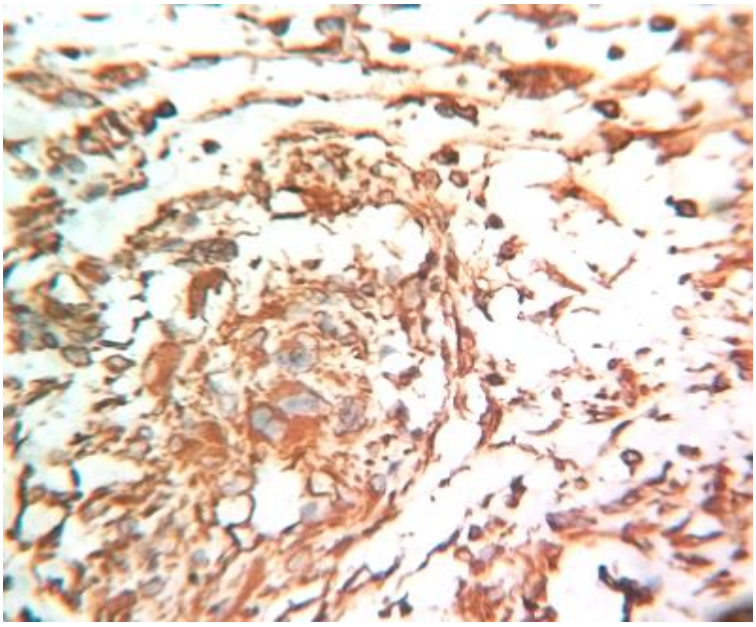


Figure 5 : Tumor cells showed diffuse and strong positivity for Vimentin

Discussion