



Paratesticular Liposarcoma: A Diagnostic Challenge

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Abstract

Liposarcoma of the spermatic cord is rare. It usually presents as a painless slowly-growing scrotal mass of consistency like lipoma. Inguinal orchiectomy is an adequate surgical approach. Retroperitoneal lymphadenectomy is not indicated owing to the low malignancy potential of the sarcomas. Value of adjuvant radiotherapy/chemotherapy remains uncertain. Recurrences are frequent, owing to incomplete surgical removal of the tumor. We report on a 66 year old male who presented with one year history of a soft painless mass in the left scrotum. Radical orchiectomy was performed. Histological examination revealed a well-differentiated liposarcoma of sclerosing subtype. No evidence of recurrence or metastases has been noted during the 6-month and one year follow-up without any postoperative adjuvant therapy.

Keywords: liposarcoma, scrotum.

Introduction

Liposarcoma of the spermatic cord is a rare malignant neoplasm that accounts for 3% to 7% of all sarcomas. Although 20% of liposarcomas originate in the retroperitoneum, only 0.1% manifest as incidental scrotal swelling. The majority of spermatic cord sarcomas begins their development just below the external inguinal ring and therefore grow as a scrotal mass, rather than as an inguinal mass. Radiation, immunodeficiency, some of the drugs, some hereditary indexes and some viruses might have roles in their pathogenesis^[1]. Sarcoma in the paratesticular region would be extremely rare^[2,3] and its treatment could not be clear^[4]. Paratesticular sarcomas have happened rarely. Due to the infrequency of this malignant disease and its diverse histopathologic subtypes, no standard

treatment would be available. Multiple treatments have reported in literature with different results.

Case history

We report a case of a 66-year-old male patient with left hemi -scrotal swelling mimicking as a testicular swelling that turned out to be a paratesticular liposarcoma. Our patient presented with complaints of a progressively increasing swelling in his left scrotal region for one year that had grown rapidly during the last three months. Physical examination revealed a mass in the left scrotal region approximately size of a walnut which was irregular, firm, non-tender, irreducible, and painless. Routine laboratory results and tumour markers for testicular malignancy were within normal limits. Initially, a clinical diagnosis of testicular tumour was made; however, CT of

the scrotum revealed bulky spermatic cord with paratesticular tumour and testis being normal and displaced poster-inferiorly. The mass was surgically removed, along with the left testis and spermatic cord (figure 1). Histopathological studies confirmed the diagnosis of well-differentiated liposarcoma (atypical lipomatous tumour of sclerosing type) (figure 2).



Fig 01

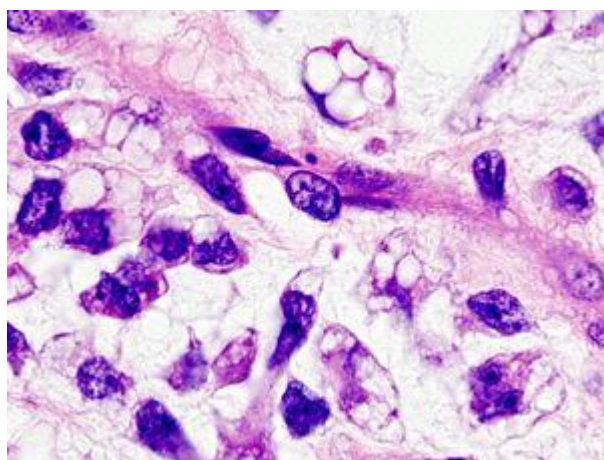


Fig 02

Discussion

Patients with paratesticular liposarcoma usually present in their fifties or sixties, with an age range of 16 to 87 years. A palpable scrotal or inguinal mass is the most common finding; the differential diagnosis includes inguinal hernia, hydrocele, and chronic epididymitis. Liposarcomas are more often located in the lower extremities (41%), the retroperitoneum (19%), and the inguinal region (12%). They are clinically indistinguishable from

testicular tumours, which thus result in difficulty in diagnosis and management. The typical clinical characteristics of a well-differentiated liposarcoma are a painless, slow-growing (months to years) soft-tissue mass. However, pain and tenderness have been reported in 10-15% of the cases. Rapid growth, large size, and symptomatic presentation are features suggestive of malignancy. A well-differentiated liposarcoma has no metastatic potential unless dedifferentiation occurs, but lesions may recur locally. The prognosis and treatment of well-differentiated liposarcomas are therefore closely related to the anatomical locations of the lesions. With adequate initial resection, local recurrence of subcutaneous lesions is rare. The rate of local recurrence is 70% for groin lesions and 91% for retroperitoneal lesions. In these cases, radiation therapy may be employed as an adjunct to surgical resection in an attempt to avoid local recurrence.

Soft tissue sarcomas have not been common among the humans and have constituted only 1% of adult malignancies. This disease has affected the males more than the females. Most of sarcomas have not associated with risk factors, but some environmental and genetic predispositions have proposed in a minority of patients^[1]

Sarcoma in tunica albuginea and vaginalis, spermatic cord and epididymis (collectively, paratesticular region), scrotal skin and testicles have been very rare^[2,3]. Patients usually have not diagnosed as sarcoma preoperatively, and have often misdiagnosed as benign lesions^[2,5,6]. Novosel has believed that these malignant lesions have usually found during hernia operations^[8]. Soler, in a review of the literature, has believed that most paratesticular liposarcomas have presented as mass^[5]. In most reviewed reports, there were long histories of scrotal mass or treatments as benign lesion. Paratesticular sarcomas might also have other presentations. Guo et al. has reported a 63 year old man who has presented with Fournier gangrene. After 10 months and multiple operations, pathologic evaluation has shown malignancy. The patient had

malignant fibrous histiocytoma of scrotum with lung metastasis and only 1 month survival ^[9]. In some reports, paratesticular sarcomas have presented as primary testicular tumors ^[10]

After suspicion of sarcoma, the next step was surgical tumor removal. Extent of surgery for paratesticular sarcoma has not been clear. In most reports, radical orchiectomy has been carried out. Kochman has reported local recurrence 3 months after local excision alone for well-differentiated liposarcoma of scrotal wall. After funiculoorchiectomy, the patient was disease free for 2 years ^[11]. But Crespo Atín has suggested that only mass excision was enough ^[12]. Lymph node dissection has not usually been necessary ^[6]. Sarcomas have been a diverse group of diseases with different behaviors. Catton et al has reviewed 21 cases during 1958 to 1987 with paratesticular sarcoma and has suggested retroperitoneal lymph node dissection for those patients with rhabdomyosarcoma, intermediate or high grade malignant fibrous histiocytoma, or fibrosarcoma ^[13]

The definite patterns of spread of paratesticular sarcomas are not well defined. Most sites for recurrence are local ^[4,5,7]. Some authors propose hematogenic routes as important for spread in paratesticular sarcomas ^[2,7]. Yol et al. reported a 63 years old man with a 2 year scrotal mass and no treatment. Their patient developed abdominal mass; biopsy and the subsequent operation showed a 42 kg retroperitoneal myxoid liposarcoma ^[14].

Paratesticular liposarcomas have usually well differentiated with good prognosis ^[15]. Schwartz has reported 6 cases with spermatic cord liposarcoma, one of whom was disease free for 23 years ^[16]. Liposarcoma was a radiosensitive tumor but its role in paratesticular liposarcoma was not clear. No adjuvant treatment has been considered standard for paratesticular sarcomas ^[4,6,7]. Some believe radiotherapy would be beneficial in recurrent or high grade liposarcoma ^[5,15]. Some authors have reported paratesticular sarcomas with a relatively prolonged disease free survival without adjuvant therapy. Hagiwara has reported a

78 years old man with spermatic cord liposarcoma. The patient has not received adjuvant treatment, and was disease free for 6 years before developing local recurrence ^[16].

The role of chemotherapy in paratesticular sarcoma has not well studied. Fujita has reported a 50-year-old man who had malignant mesenchymoma (consisting of osteosarcoma, leiomyosarcoma, and liposarcoma) of spermatic cord. Their patient has presented with painless swelling in the inguinal area. After orchiectomy, the patient has received 2 courses of the CYVADIC regimen. No recurrence has observed after 12 months when they have reported this patient again ^[17].

Conclusion

The rarity of a paratesticular liposarcoma makes it very difficult to differentiate it from other causes of scrotal swelling. Paratesticular liposarcoma should be considered as a possibility during the differential diagnosis of scrotal mass. Paratesticular liposarcoma would be so rare to have a well-studied clinical presentation, clinical course, treatment and survival. It has seemed presenting good prognosis, but further studies are needed to draw a conclusion.

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