

Case Report

Recurrent Giant Paratesticular Myxomatous Neoplasm

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الورم المخاطي العملاق الجار خصوي المتكرر

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الملخص العربي:

ان المنطقة الجار خصوية من المناطق المعقدة تشريحيا وخلويا لذا نجد ان الاورام الناشئة منها قد تكون حميدة او عدوانية للانسجة المجاورة او حتى خبيثة . هنا نسجل نص سريري مفصل ومتابعة علاجية لحالة نادرة لورم مخاطي عملاق جار خصوي مع مناقشة التشريح المرضي النسيجي المثير للجدل . لقد قدمنا حالة ورم جار خصوي مخاطي مع الوصف للتاريخ السريرية المرضي ، والنتائج على ضوء الفحص المجهرى والمناعية، والمرضية المحتملة. على الرغم من ان الورم الجار خصوي المخاطي آفة نادرة، الا انه ينبغي النظر في ذلك في التشخيص التفريقي للأورام في هذه المنطقة .

ABSTRACT

The paratesticular region is complex in both anatomical and histogenetically. eoplasms arising from this region therefore might be benign, locally aggressive or even malignant. We present a 20 year old male case of giant paratesticular myxomatous tumor and describing clinical history, findings on light microscopy and immunohistochemistry, and possible pathogenesis and the controversial histopathological features of a rare giant paratesticular myxomatous neoplasm. Although primary paratesticular myxoma is a rare lesion, it should be considered in the differential diagnosis of intrascrotal mesenchymal tumors. The treatment of choice is radical inguinal orchidectomy and wide tumor excision with safe margins. Adjuvant therapy probably not necessary but long follow up is essential.

Keywords: Myxoid neoplasm, Myxomatous neoplasm, Myxolipoma, Spindle cell lipoma, Myxofibrosarcoma, Aggresive angiomyxoma, paratesticular tumor.

INTRODUCTION

The paratesticular region is complex anatomical area which includes the contents of the spermatic cord, testicular tunics, epididymis and vestigial remnants^{1,2}. Histogenetically this area is composed of a variety of epithelial, mesothelial and mesenchymal elements. Neoplasms arising from this region therefore form a heterogeneous group of tumors with different behavioral patterns³. It may harbor rare varieties ranging from pure benign to locally aggressive lesions. Although malignant tumors are rare, their involvement in the differential diagnosis renders them clinically important⁴. Paratesticular myxomatous tumors are rare tumors that may arise in this area, it occurs in middle aged men⁵. These tumors are prone to local recurrence following inadequate extirpation, even with adjuvant radiotherapy⁶. We report a rare entity of giant paratesticular myxomatous neoplasm which created some histopathological controversies. Initially mislabeled as an aggressive angiomyxoma. Then the possibility of myxofibrosarcoma was raised. Lastly, two soft tissue experts supported the view of angiomyofibroblastoma-like tumor, with final impression of myxomatous neoplasm with features of myxolipoma and spindle cell lipoma. The clinical scenario, work up, management, follow up and the differences in histopathological features are presented in this report.

CASE

A 20 year old male patient presented to us complaining of gradual and significant enlargement of his left scrotum associated with dragging dull ache pain. One and half year prior to presentation, he had undergone repeated partial excision of a left scrotal mass in a different hospital where he first presented as a case of hydrocele or/and hernia. According to the histopathological report from that hospital, diagnosis was "an aggressive angiomyxoma". On physical examination he had a huge left inguino-scrotal soft mass measuring around 20 x 17 x 5 cm., not reducible or compressible. No lymph nodes were felt in inguinal or adjacent regions. MRI and CT scan (Fig. 1) showed no deeper lesions. The patient underwent left radical orchidectomy and wide inguinoscrotal mass excision (Fig. 2) with safe margins. Grossly, on slicing, the tumor contained mucoid and gelatinous material. Microscopically, general sections showed a hypocellularity with focal increase of cellularity and cytological atypia. The tumor appeared to be quite infiltrative of the soft tissue. The tumor cells showed mild pleomorphism although focally. Occasional large tumor cells were seen with irregular or multilobulated, hyperchromatic nuclei. Mitoses were not seen. (Fig. 3) The tumor cells were positive for vimentin, CD34 and for Bcl2. They were negative for SMA, MSA, desmin and for S100 protein. An impression of myxofibrosarcoma was raised as a possibility. The slides were reviewed at by two other international soft tissue experts who gave the impression of myxomatous neoplasm with features of myxolipoma / spindle cell lipoma. A possibility of angiomyofibroblastoma-like tumor was also considered. Due to the histopathological report of multiple projections and the adhesions from previous surgery, the patient was referred for radiation therapy to prevent possible recurrence. The patient refused this plan. Three years later, on follow up, no local recurrence was seen. This would favor the myxomatous neoplasm.

DISCUSSION

Benign and malignant tumors of the paratesticular tissues present an interesting spectrum of diagnostic entities often encountered in routine surgical pathology practice. Despite of the infrequent, it has been estimated that 70% of paratesticular tumours are benign and 30% are malignant. Although it is often difficult to determine with certainty the exact site of origin of paratesticular tumors, it is thought that the spermatic cord is the most common, accounting for 75- 90% of which 75-80% are primarily comprised of lipomas, with nearly all malignant counterparts being sarcomas^{2,7}. There has been substantial research in the paratesticular soft tissue pathology with increasingly support by cytogenetics and molecular genetics, making it more logical, reproducible and substantially less confusing than it has ever been. As a consequence, the major beneficiaries are patients. Without question there will be continuing advances and conceptual shifts, as in any other area of pathology, and any future WHO classification will undoubtedly bring further improvement^{8,9,10,11}.

In our case, at one stage, aggressive angiomyxoma was thought to be the diagnosis. This tumor is a rare but well described neoplasm. It was recognized as a separate histopathological entity in 1983. This is a distinctive tumour with a characteristic clinical course and specific gross and microscopic features. It has been reported in men, but mostly reported in women. In men it occurs over a wide variety of sites around the genital tract such as perineum, perianal region, scrotum, spermatic cord, inguinal region, and pelvic soft tissues. The final diagnosis is usually provided by the pathologist. Aggressive angiomyxoma occurs in ages ranging from 15 to 63 years. It is characteristically a slow growing, focally infiltrative tumour, lacking a capsule. Histologically, the tumour shows vascular channels of varying sizes distributed haphazardly in the myxoid stroma. Nuclear atypia and mitosis are not present. Although occasionally well circumscribed, they are usually not encapsulated and microscopically infiltrative. It is a locally aggressive but non-metastasising neoplasm. After excision, a significant rate of local recurrence of 20% is reported¹¹.

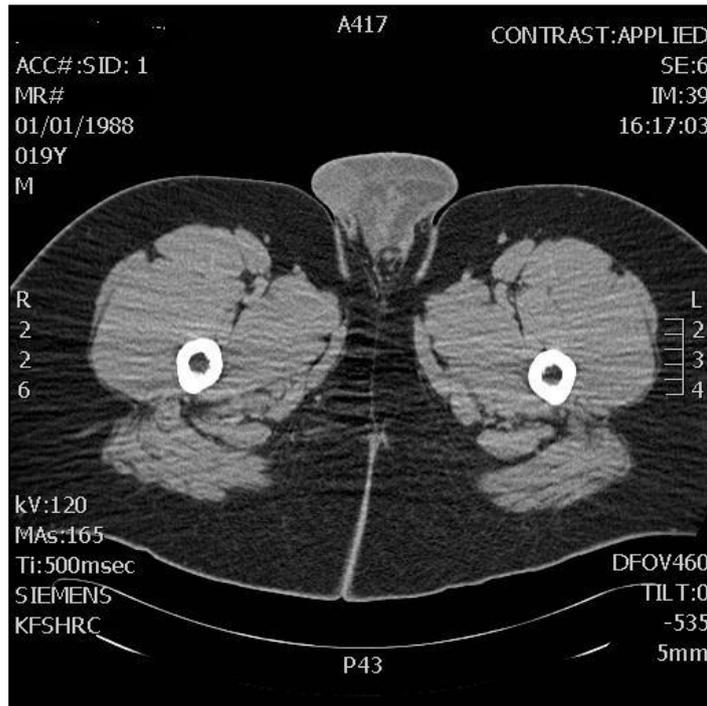
On the other hand, myxofibrosarcoma is a myxoid subcategory of malignant fibrous histiocytoma, which is, in general, the most common soft tissue sarcoma of late adulthood (50-70 yrs) and second most common sarcoma of the retroperitoneum. Mostly a tumor of soft tissues, with 20% found in the abdomen and retroperitoneum. The Myxoid subtype represents 15% of this whole group. Composed of loose myxoid stroma which constitutes 50% of the tumor mass, interspersed with areas of higher cellularity, abundant plexiform vascular network, scattered giant cells. Metastases present at diagnosis in 25%. Five year survival is more than 50%.¹² Angiomyofibroblastoma, shows a predilection for the vulvovaginal region during the reproductive years but may also occur in the inguino-scrotal region in men and is distinguished from aggressive angiomyxoma. Angiomyofibroblastoma is a recently described benign tumor in the scrotum and inguinal region of men. Microscopically it has high cellularity and vascularity. Unless there is a sarcomatous transformation, they behave benignly and are adequately treated by local excision^{2,8,12}.

Myxolipoma is a very old term reported since 1857 in many different parts of the body but rarely reported in the scrotum¹³. Spindle cell lipoma is characteristically located at the upper trunk or rarely at the lower limbs. It shows microscopally a mixture of mature lipocytes and uniform, primitive, bland, S-100 negative spindle cells in a mucinous and fibrous background with frequent mast cells. It can be distinguished from myxoid liposarcoma by the thick collagen bundles, the absence of lipoblasts and a plexiform vascular pattern. No matter what are the underlining histological subtypes, the clinical presentation is mostly a mass or

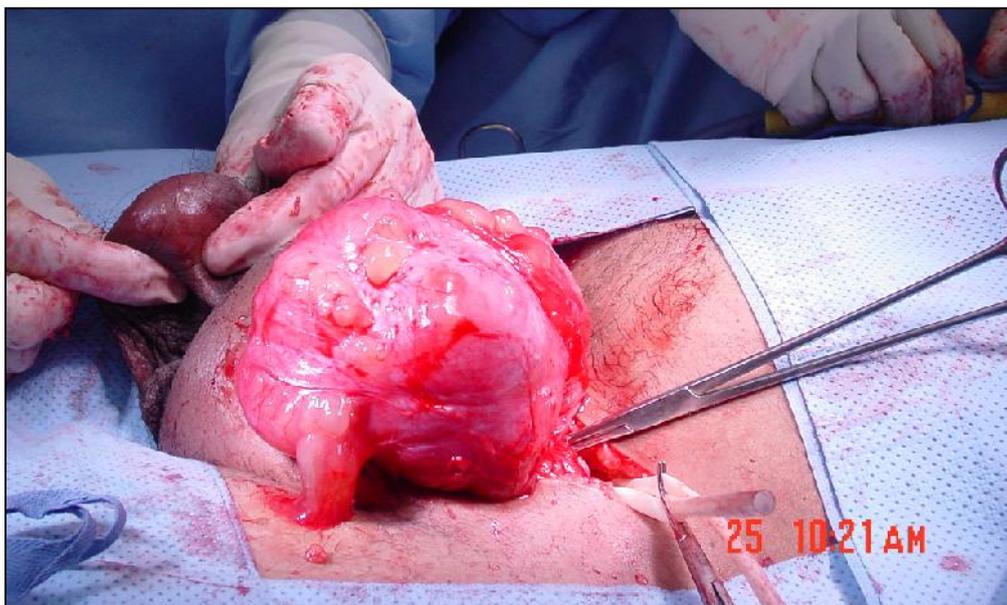
swelling, which might be painless or painful and is occasionally accompanied by a hydrocele. These findings are by no means specific to a tumor type and it cannot distinguish a benign from a malignant tumor. High-resolution ultrasonography of the scrotum provides evidence of the location, extent, and relationship of intrascrotal masses with the testis, epididymis, or spermatic cord; the echogenicity and the internal echo patterns obtained allow cystic, fluid-filled lesions to be distinguished from solid ones. However, the technique cannot be used to discriminate between benign and malignant lesions. Unlike sonography, CT Scan can furnish densitometric evidence of tumor texture. MRI may locate the tumor better and define its relationship to various paratesticular structures in greater detail, which is not always possible with ultrasonography¹⁴. Careful histological assessment with the knowledge of the various pathological subtypes is crucial to formulating adequate treatment and predicting clinical outcome. All benign tumors of the paratesticular region are amenable to adequate surgical resection, but a close follow-up for some of these tumors has been recommended, because of occasional recurrences and rare malignant transformation, or even misdiagnosis for a rare unusual locally aggressive pathology.

CONCLUSION

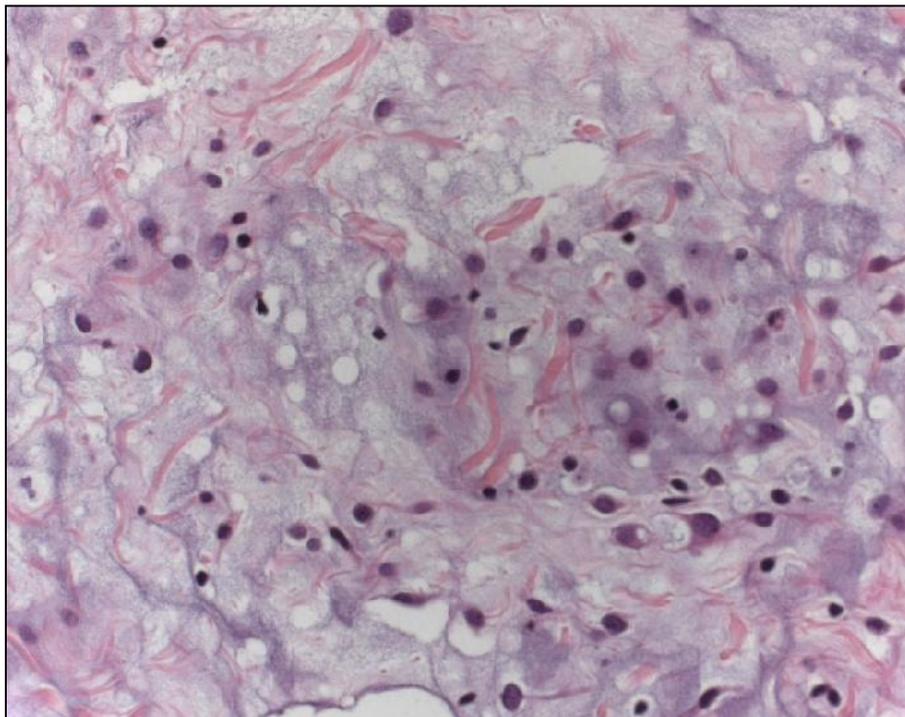
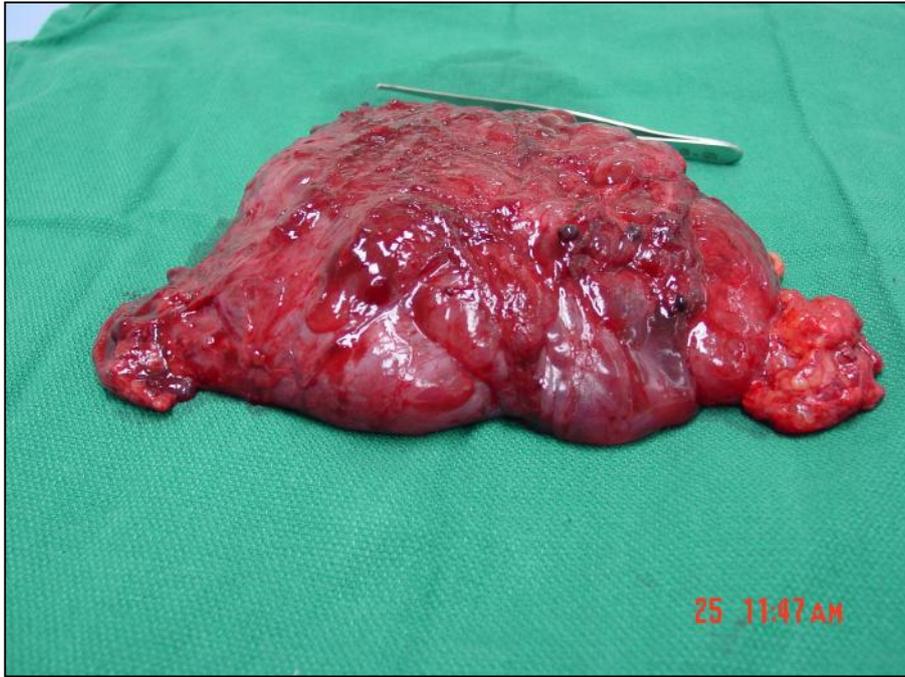
Myxomatous neoplasm of the spermatic cord is a very rare tumor that can present during adulthood as a painless scrotal or inguinal mass that can be misdiagnosed as inguinal hernia, hydrocele or spermatocele. The new case that we present is an example of how similar tumor can be misleading to both surgeon as well as to general pathologist. Imaging can be useful, although definite diagnosis cannot be made without histological examination. Yet Careful histological assessment with the knowledge of the various pathological subtypes is crucial to formulating adequate treatment and predicting clinical outcome. The treatment of choice is radical inguinal orchidectomy and wide tumor excision with safe margins. However, conservative limited tumor excision has the risk of recurrence. Surgery seems also enough for local recurrences. Adjuvant therapy is probably not necessary since paratesticular myxomatous neoplasm have very good prognoses if completely excised and life-long follow up is accessible. The present case, which to date has had a follow-up of only medium duration, is an example of the above approach.



(Figure-1)
Figure 1. CT of the abdomen.



(Figure- 2)
Figure 2. gross pathology photograph of the tumor.



(Figure-3)
Figure 3. Microscopic photograph of the tumor

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