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Research Article

A RARE CASE OF PARATESTICULAR ANGIOFIBROMA

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ABSTRACT

Cellular angiofibroma, also referred to as angiomyofibroblastoma-like tumor, is a rare type of extratesticular mesenchymal tumor found in the scrotum and perineum. Initially identified predominantly in women's vulva, perineum, and pelvic region, it represents a distinct entity within this spectrum of tumors. Angiofibroma in male population is very rare. We hereby present the case of a 48 year old male patient with gradually progressive, painless swelling in the right scrotum, where USG and MR revealed paratesticular mass. Transingunal orchidectomy was done and histopathological and immunohistochemical analysis confirmed the diagnosis of cellular angiofibroma.

INTRODUCTION

Paratesticular tumors, a rarity within the scrotum, originate from structures such as the epididymis, spermatic cord, and their coverings. Among these tumors are cellular angiofibromas (CAFs), benign growths uncommon in soft tissue, often observed in the genitourinary region. While both genders can be affected, in women, CAFs are predominantly found in the vulvo-vaginal region, while in men, they more frequently manifest in the inguino-scrotal area. these tumors typically emerge around the fifth decade of life. We present the case of a 48-year-old man who presented with a painless testicular mass with no significant medical history.

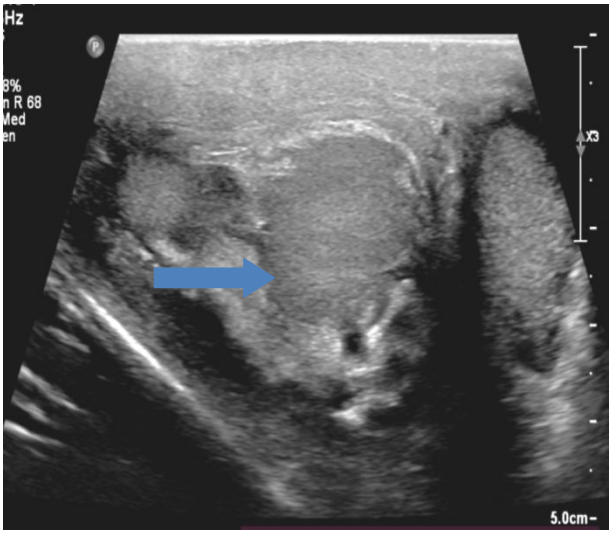
CASE REPORT

A 48-year old male presented with a gradually growing painless mass in the Right scrotum since 10yrs. He had no significant family history, trauma, infections, or other risk factors that could have contributed to his condition. On genital examination, a large oval shaped nontender mass was noted in the right scrotum. The right testis was not seperately palpable. The left testicle, however, was of normal size, and lymphadenopathy was not observed.

Imaging Features

Ultrasonography

A solitary oval shaped, encapsulated heteroechoic lesion in the right scrotum adjacent to the right testis with significant internal vascularity measuring approximately 4.8x3.5cms.
Both the testes, epididymis and spermatic cords were visualised seperately.



Ultrasound transverse section shows heteroechoic mass ( ) seperate from both testes.

Provisional diganosis of extratesticular benign mass was made and the patient was suggested to undergo MRI for further evaluation.

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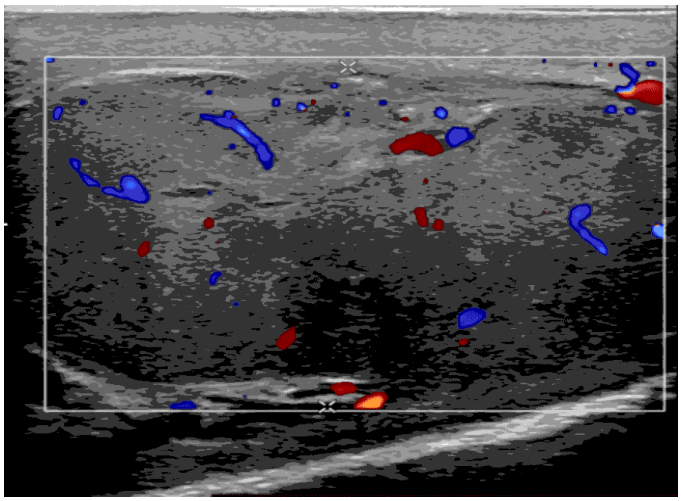
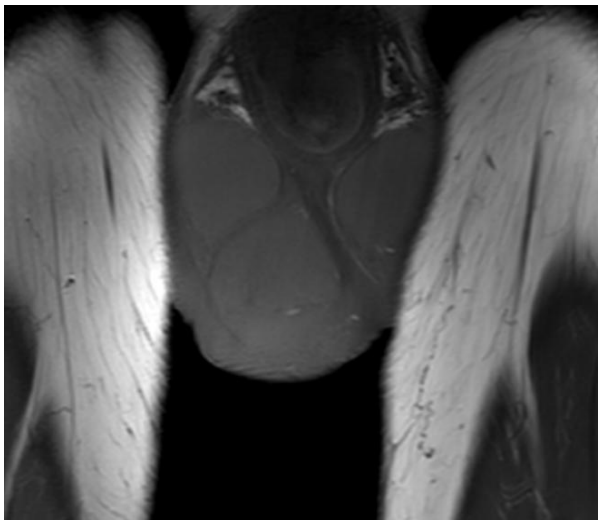
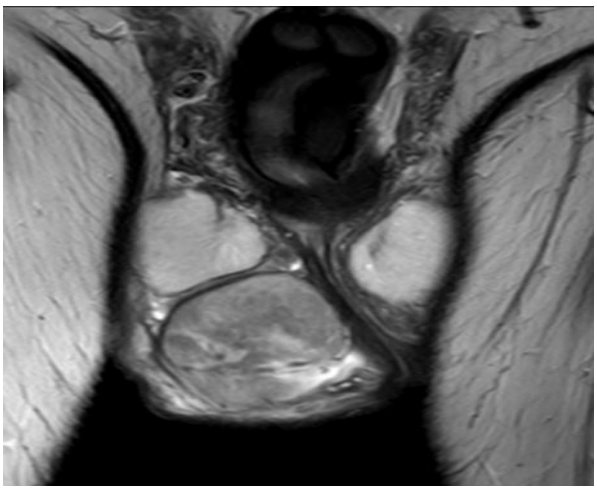


Ultrasound longitudinal section shows heteroechoic mass with internal vascularity

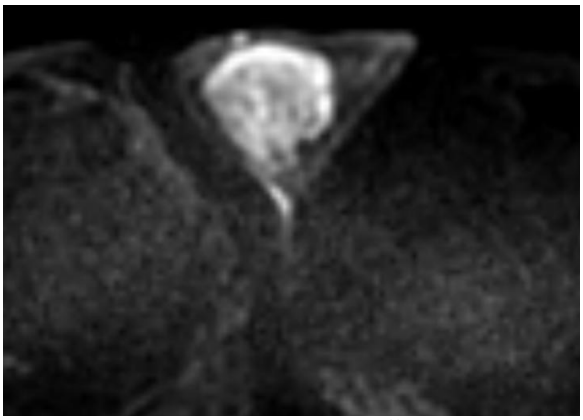
MRI

Plain MRI was performed. It showed an intra scrotal, paratesticular mass lesion which was seperate from both testes measuring 60x40mm. T1-weighted image shows a tumor with homogeneous low signal intensity (arrow) secondary to fibrous content. T2-weighted image shows a high-signal-intensity tumor with internal heterogeneity (arrow). There was no restriction on diffusion weighted imaging. Few intralesional bloomings on GRE.

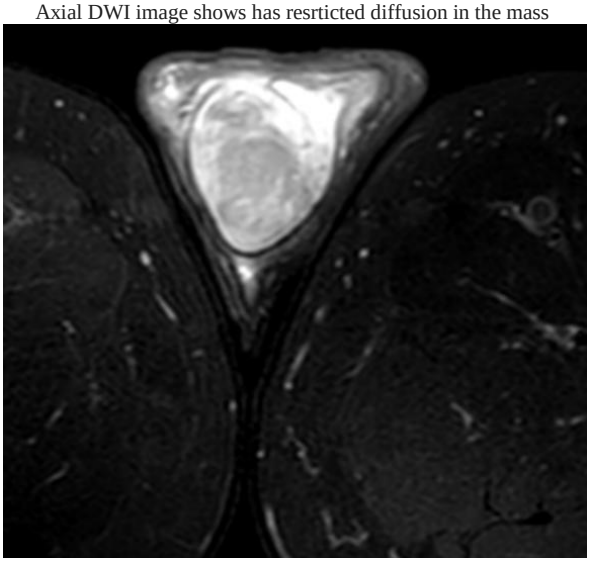




MR coronal images show extratesticular mass seperate from right testis which is hypointense on T1W and heterogenously hyperintense on T2W images



Axial DWI image shows has resrticted diffusion in the mass



The mass does not show supression on T2W Fat saturated axial section, which eliminates the possibilities of fat within lesion.

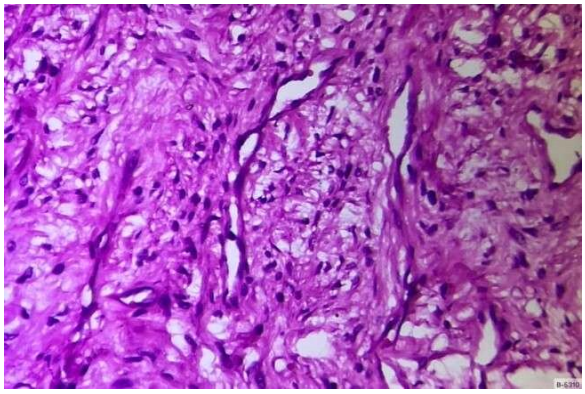
Tumor markers including beta-human chorionic gonadotropin, alpha-fetoprotein, and lactate dehydrogenase were tested, and all yielded negative results. A provisional diagnosis of benign paratesticular tumor was made, and the patient was sent for surgery.

Histopathological Findings

A transinguinal orchidectomy was done. There were no specific findings of adhesion to surrounding tissue or intraoperative complications. On gross examination, the tumor measured 6x3.7x4cm and was heterogeneous grayish-white in color. The tumor was encapsulated by fibrous tissue and detached from the testis.

Gross examination of postorchidectomy specimen shows a paratesticular mass.

Microscopically, the tumor showed proliferation of fibroblastic spindle cells in a diffuse pattern accompanied by collagenous fibers and dilated vascular channels with perivascular fibrinoid hyalinization. Immuno histochemical markers were positive for Vimentin, CD34 & Desmin and negative for SMA & S100.



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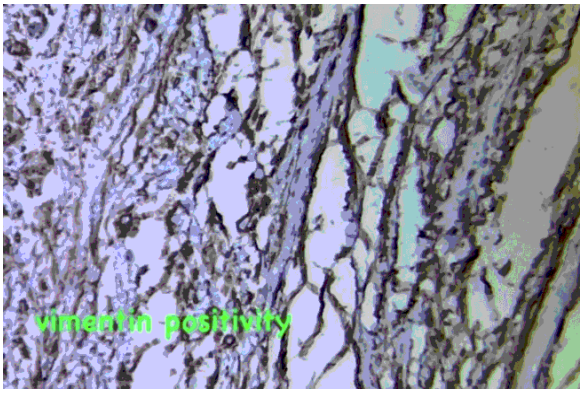

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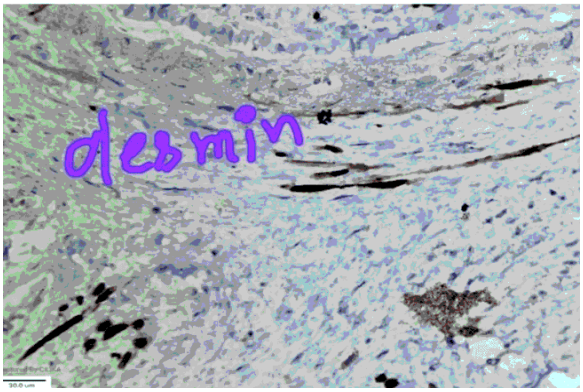

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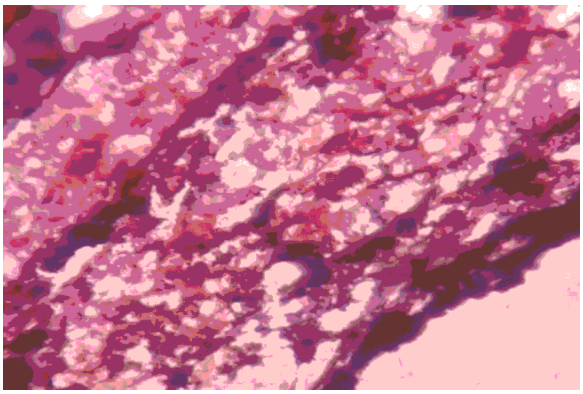

D

(A)Microscopically, tumor lesion shows proliferation of short spindle cells in a diffuse or perivascular pattern, accompanied by collagenous stromal fibers and dilated vascular channels (B-D)Immunohistochemical findings (B)CD34 (C)Vimentin (D) Desmin Histopathological diagnosis of Paratesticular cellular angiofibroma was made.

DISCUSSION

Cellular angiofibroma is a rare mesenchymal tumor and is often recognized by its well-defined shape. It is usually characterised by painless mass developing slowly over about 5 months on average. Although some individuals might experience mild to moderate discomfort. Typically, the tumor measures around 2.8 cm in women and 7 cm in men, with a cut surface that commonly exhibits shades of gray, pink, and brown.

Diagnostic imaging methods for CAFs encompass ultrasonography with color Doppler, CT scans, and MRI. The presentation of cellular angiofibroma on imaging studies may vary, with certain tumors displaying hyperintense signals on T2-weighted MRI images or density alterations on CT scans attributable to their vascularity. Nevertheless, radiological observations lack high specificity and do not substantially aid in distinguishing CAFs from other mesenchymal tumors.

Histologically, cellular angiofibromas (CAFs) are well-defined, cellular, and consist of spindle-shaped cells with short collagen bundles, fusiform nuclei, and pale eosinophilic cytoplasm. Importantly, they often contain numerous thick-walled and hyalinized vascular structures. Cellular atypia or necrosis is rarely observed.

Immunohistochemically, CAFs typically show positive staining for vimentin, CD34, and estrogen receptor (ER), while they are negative for desmin, smooth muscle actin (SMA), S100 protein, and epithelial markers. The presence of CD34 and ER immunopositivity further supports the vascular and hormonal characteristics of CAFs. A minority of cases are positive for SMA and desmin. These distinctive histological and immunohistochemical features aid in the accurate diagnosis and differentiation of CAFs from other spindle cell tumors and help guide appropriate clinical management.

The pathogenesis is still unknown; however, multiple hypotheses have been proposed, including the involvement of a monoallelic deletion of *RB1 (RB transcriptional corepressor 1)* and *FOX1 (RNA Binding FOX-1 Homolog-1).* Both genes reside within chromosome 13q14, which is highly linked to the development of the disease.

The treatment of choice is local excision with negative margins. The probability of recurrence is very low.

CONCLUSION

In conclusion, this case report highlights a rare instance of CAF. Diagnosing this tumor type poses challenges because of its vague clinical and radiological characteristics, often resulting in misdiagnosis or unwarranted invasive interventions. Although surgical excision remains the primary treatment approach for CAFs, monitoring is crucial for detecting any potential malignant transformation, recurrence or related complications.

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