BRITISH JOURNAL OF HOSPITAL MEDICINE

CASE REPORT

Male angiomyofibroblastoma-like tumour: a paratesticular tumour and the importance of a second opinion

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How to cite this article:
Neale A, Warner R, Miller M. Male angiomyofibroblastoma-like tumour: a paratesticular tumour and the importance of a second opinion. Br J Hosp Med. 2021. https://doi.org/10.12968/hmed.2021.0058

Introduction

The majority of masses found in the scrotum are neoplastic tumours occurring within the testis, but a subset arise from the paratesticular tissue (Khoubehi et al, 2002). Angiomyofibroblastoma-like tumours of the male genital tract are rare, with only 37 reported in the literature (Bouhajja et al, 2017; Msakni et al, 2017; da Silva et al, 2019). Clinical and sonographic findings are non-specific, with a number of differentials to consider that can dramatically affect patient outcomes. This article reports one example and highlights the challenges posed on diagnosis, emphasising the importance of expert and repeated reviews.

Discussion

Angiomyofibroblastoma-like tumours of the male genital tract generally present as 2.5–14 cm, superficial, mobile and well delineated masses in the scrotum or inguinal region

Case report

A 48-year-old man presented to his GP in August 2017 with a 4–5-month history of a slowly enlarging, painless left scrotal mass. His only comorbidity was hypertension.

His GP organised an ultrasound of the testes, which reported a left-sided inguino-scrotal hernia. No further follow up was arranged. The patient re-presented to his GP 8 months later with lower abdominal pain and was referred for an outpatient surgical review.

Owing to the uncertainty of diagnosis, the patient was then referred to a urologist. Clinical examination demonstrated a normal left-sided testis, with a 3 cm, soft, non-tender 'cystic feeling' inferior scrotal lesion. Repeat testicular ultrasound described the absence of an inguinal hernia and, instead, the presence of a 4 × 3.5 cm, extratesticular, solid lesion adjacent to the lower pole of the left testis.

Routine laboratory investigations were within normal limits, and contrast computed tomography showed no evidence of malignancy or metastatic spread. The patient then underwent scrotal exploration (via a midline scrotal incision) and excision of the left scrotal mass under general anaesthetic. During the procedure a mixed solid and cystic lesion was visible adjacent and separate to the left testis. The mass was separated from the testis and excised, leaving the testis in situ.

Initial histology described a tumour composed of bland spindle cells within a myxoid stroma. No mitotic activity was seen, and prominent vessels were noted. Immunocytochemistry revealed cells as positive for vimentin, CD34 and desmin. Smooth muscle actin, S100, EMA, MNF116 and H-caldesmon were all negative. STAT6 was initially found to be positive. Initial proposed differentials included angiomyofibroblastoma, angiomyxoma and myxoma, but appearances were most suggestive of a solitary fibrous tumour. The case was sent to the regional sarcoma unit for further review.

The final report described delicate spindle stellate cells set among a light myxoid stroma, with a prominent vascular component (Figure 1), diffuse positivity for ER but negative for PR, STAT6 and MDM2 amplification in FISH, in addition to the above.

Discussion at the regional sarcoma multidisciplinary team meeting confirmed the diagnosis of a paratesticular male angiomyofibroblastoma-like tumour. Given the benign nature of the disease, local clinical follow up was advised.

On subsequent 6-monthly reviews the patient has appeared well, with no evidence of scrotal mass recurrence on examination.

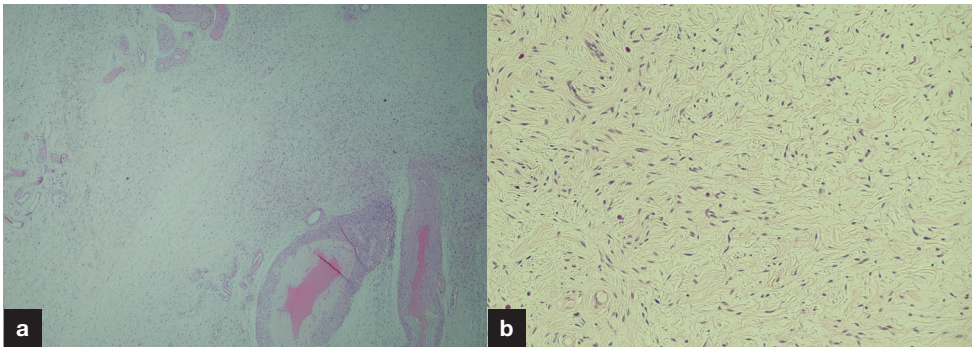

Figure 1. Micrographs of the specimen at (a) ×2 magnification and (b) ×10 magnification with haematoxylin and eosin stain, demonstrating spindle stellate cells proliferating around prominent vessels within a light myxoid stroma.

(presenting age ranging from 39 to 88 years) (de Souza et al, 2009). They are benign in nature, with complete local excision the treatment of choice, and have an extremely low risk of recurrence (Iwasa and Fletcher, 2004; de Souza et al, 2009).

During initial histological analysis, two of the main differentials were solitary fibrous tumour and angiomyxoma. The central histological distinctions are that angiomyofibroblastoma-like tumours lack the 'keloid-type collagen and hemangiopericytoma-like areas that are distinctive of solitary fibrous tumours' (Canales et al, 2006; Aytac et al, 2012), and contain 'high cellularity and more pronounced vascularisation' than angiomyxomas (da Silva et al, 2019). Angiomyxomas are aggressive and invasive, recurring frequently (da Silva et al, 2019). Most solitary fibrous tumours are benign and treated adequately by resection; however, up to 15% are aggressive and demonstrate risks of local recurrence or metastasis, requiring long-term follow-up (Gold et al, 2002; Lee et al, 2011). Had the above patient not had his histology repeatedly analysed and reviewed by a specialist sarcoma multidisciplinary team, he may have been given an incorrect diagnosis, and subjected to more intensive follow up unnecessarily.

The patient was diagnosed as having an inguino-scrotal hernia on his initial ultrasound. When used alongside appropriate clinical information, ultrasound can be helpful in distinguishing paratesticular tumours from other pathology (Khoubehi et al, 2002). However, as demonstrated by this case, it has limitations. It is unclear whether this patient had two separate pathologies at different time points or was misdiagnosed initially. This highlights the importance of an expert clinical examination, the usefulness, on occasion, of repeat imaging and the value of a second opinion.

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Learning points

- Paratesticular angiomyofibroblastoma-like tumours are rare, but important differentials to consider when presented with a scrotal lesion.
- Paratesticular tumours can be difficult to diagnose both clinically and radiologically. A multidisciplinary approach should be taken, and histological analysis remains the formal method of diagnosis.
- It is important for GPs and urologists to remember that paratesticular tumours can be malignant, and therefore require urgent investigation.
- At the time of surgery patients should be made aware of the uncertainty of the diagnosis and potential need for further surgery or other treatment. Likewise, the tumour may also be benign as in this case.
- Urgent referral to a sarcoma unit should be advised. Multiple analyses of specimens, notably by specialist units, is of irreplaceable value. Second or third opinions are invaluable to ensure the patient receives an accurate diagnosis.

Acknowledgement

Following acceptance of this article, Professor Marek Miller, supervising author sadly passed away. He was a wonderful mentor, educator, colleague and friend. The authors would like to thank Dr Antonia Barbieri and the histopathology team at Northampton General Hospital NHS Trust for providing the images for this article.

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