Case Report

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Scrotal Masson's tumour masquerading as dysplastic polyorchidism – a case report

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Abstract
This case describes a rare genitourinary presentation of a Masson's tumour, preoperatively assumed to be a dysplastic third testis.

Keywords
Urological surgery, urology, histopathology

Case description

A 34-year-old man presented with a new scrotal lump. He described a history of a small and painless right scrotal mass which had been present for 2 years. In the preceding 4 weeks it had rapidly increased in size. On examination, there was a 3 cm smooth, firm, non-tender mass, separate from the right testes, with no associated ecchymosis or skin changes. The mass was inferior to the right testicle and not adherent to the skin. Posterior to the mass there was a palpable pedicle.

He was initially investigated with an ultrasound of his testes which demonstrated an extra-testicular mass measuring 29 mm × 29 mm × 19 mm in size, posterior and separate to the right testis. The mass had a heterogeneous echotexture, and appeared vascularised on colour Doppler. The nature of this lesion remained uncertain.

He then underwent an MRI of his pelvis which demonstrated a solitary ovoid mass in the right hemiscrotum posterior and separate to the testes and epididymis, measuring 36 × 21 × 21 mm. On the rostral margin it appeared to drain into the ipsilateral pampiniform plexus. Bilateral testes, epididymi and the spermatic cords were normal. On review of the images, the lesion had radiological features consistent with a testicle, it appeared to have the same density, and hence raised suspicion of polyorchidism with a dysplastic third testes (Figure 1).

He subsequently underwent surgical excision of the lump via a perineal incision. Intraoperatively, a discrete lesion was found at the base of the scrotum, however it was external to the hemiscrotum tunica vaginalis and completely separate from the testes (Figure 2).

Preoperative testicular tumour markers were normal (LDH 184, AFP 1, HCG <1), as were all other blood tests.

Histology of the lesion revealed an encapsulated haemorrhagic structure. Microscopic examination revealed circumscribed vascular proliferation. Interconnecting small vascular channels were seen within an expanded vessel. Many of these contained fibrinoid material/thrombus and they were lined with endothelial cells. The endothelial cells were cytologically bland in appearance and varied from flattened to plump but did not demonstrate cellular atypia or mitotic activity. This lesion was characterised as intravascular papillary endothelial hyperplasia (also known as Masson's tumour), with no evidence of malignancy nor of any testicular tissue.

Discussion

Masson's tumour or intravascular papillary endothelial hyperplasia (IPEH) is a rare benign vascular lesion, representing 2% of all vascular tumours.[1] It was first described by Pierre Masson in 1923 and has been variably described as Masson's haemangioma, pseudoangiosarcoma and vegetant intravascular haemangioendothelioma.[2] Latterly, Clearkin and Erzinger reported a large case series observing the causative association with thrombosis and suggested the more descriptive term IPEH.[3]

IPEH is most commonly described on the head and neck, hands and trunk, although may occur in any blood vessel.[3,4] Uropathological presentations of IPEH are extremely rare and to our knowledge has not previously been described within the scrotum. There have been a very limited case reports detailing IPEH within the urethra,[1,5] kidney[6] and urinary bladder.[7]

IPEH is thought to be a reactive proliferation of vascular cells secondary to a thrombus.[7] Therefore, it appears microscopically as a network of vessels containing thrombus with a surrounding network of proliferative papillary endothelial tissue. It may be characterised further depending on where the thrombus arises: primary or pure IPEH is formed around a normal blood vessel, secondary in association with a pre-existing vascular malformation and

Figure 1. Preoperative magnetic resonance imaging of pelvis and scrotum demonstrating testicle-like lesion. Left: sagittal slice showing the location of the mass in the right hemiscrotum with apparent drainage into the ipsilateral pampiniform plexus. Right: axial slice at the level of the perineum.

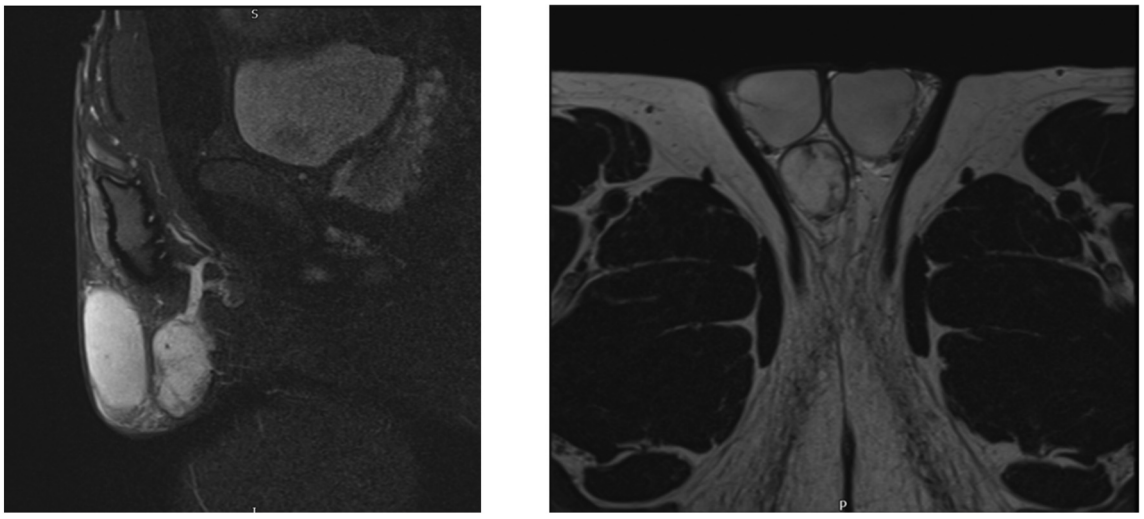
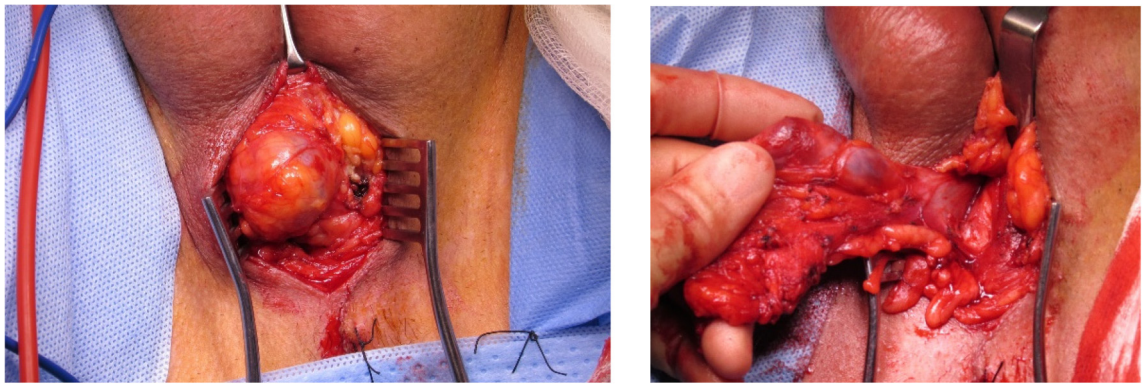


Figure 2. Photographic images demonstrating intraoperative findings of the mass via perineal approach. Note that it is discrete in nature and external to the right hemiscrotum tunica vaginalis.



more rarely it may be found extravascularly in association with a haematoma.[8]

Clinically, IPEH presents with a small firm mass often palpable beneath the skin and subcuticular tissues.[4] IPEH may cause painless visible and non-visible haematuria when associated with the urinary tract.[1,5]

IPEH may present significant diagnostic challenges as some aspects of its clinical and histological presentation mimics that of a malignant neoplasm. Differential diagnoses include sarcoma, granuloma and haemangioma. In this case report, the main differential was a dysplastic third testes.

Ultimately diagnosis is histological. Microscopically the fine vascular network of IPEH may closely mimic haemangiosarcoma but may be differentiated by its mesenchymal origin, reduced atypia, size and its close relation to a thrombus.[7] IPEH stains positive CD31, CD34 and SMA.[9]

Management of IPEH is usually via complete excision biopsy to exclude malignancy. Complete resection may be considered curative.[7] There have been some reports of recurrence in relation to incomplete resection or underlying vascular tumours.[10] However, IPEH in itself is not a malignant process and therefore does not warrant ongoing follow-up.

Conclusion

In this case, the mass radiologically mimicked testicular tissue and dysplasia of the presumed tertiary testis. The

intraoperative findings demonstrated that the mass was extra-scrotal and was amenable to excision via the perineum. Histology confirmed the diagnosis of IPEH and noted the presence of thrombus in the small vascular channels of the tumour. This raises the suspicion of an intratumoral bleed, which would explain the sudden increase in size in the preceding four weeks. At his six-week post-operative outpatient review, the patient's wound was well healed, and the histological findings were explained to him. As this lesion did not have any malignant potential, local resection was considered curative, and he was discharged.

Declarations

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