IgG4 paratesticular fibrous pseudotumor: Case presentation and literature review

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ABSTRACT

PFP with IgG-4 immunostaining is a rare paratesticular tumour. Pre-operative ultrasound scan and MRI usually confirm the benign nature of the paratesticular mass avoiding the need for radical orchiectomy. The final diagnosis is based on histology of the removed paratesticular tumour. FDG PET scan plays an important role in ruling out systematic IgG4-related disease (IgG4-RD). We describe a case of multiple paratesticular fibrous pseudotumors with IgG4 immunostaining, not associated with systemic IgG4 related disease.

1. Introduction

Paratesticular Fibrous Pseudotumours (PFPs) are benign lesions of paratesticular structures. PFPs, known as proliferative periorchitis and reactive periorchitis, are rare with fewer than 200 reported cases to date. PFPs can also be a manifestation of Immunoglobulin G4-related disease (IgG4-RD) which form a smaller proportion of PFPs. We present an uncommon case of IgG4-RD presenting as a Paratesticular Fibrous Pseudotumour.

2. Case description

A fit 28-year-old man presented in 2018 with left-sided testicular pain and mass. Scrotal ultrasound confirmed normal testis and epididymis bilaterally. There were two extra-testicular isoechoic nodules measuring 8 and 9 mm respectively with no increase in vascularity, in contact with the surface of the left testis with no invasive features. There was also presence of a varicocele on the left side. These lesions were thought to be granulomas and a repeat ultrasound in 6 months was planned. He defaulted from follow-up and re-presented in October 2021 with left testicular pain and a clinical increase in the number of left testicular lumps. Repeat ultrasound identified an 18 mm × 0.8 mm nodule in the anterolateral aspect of the left testis. It had soft tissue characteristics, was well-defined and separate from the testis, and showed minimal internal vascularity (Fig. 1). It had increased in size from the previous scan in 2018, and a possible second lesion may have been hidden within the enlarging left varicocele. A diagnosis of fibrous pseudotumor or extratesticular leiomyoma was considered at this stage. MRI of the pelvis and scrotum was performed which identified 4 conglomerate nodules, with the largest measured at 14mm. All these nodules were isointense on T1 weighted sequences and hypointense on T2 weighted sequences, with no restriction of diffusion. The appearances were suspicious for testicular adenomatoid tumour or paratesticular fibrous pseudotumor (Fig. 2).

The patient underwent left scrotal exploration and excision of the left paratesticular tumours. Intra-operatively there were 13 yellow/white hard nodules with narrow stalks scattered in the paratesticular area and epididymis with no invasion of the testis or epididymis. Histology confirmed paratesticular fibrous pseudotumour. Further immunostaining for IgG4 was positive, with 8–10 IgG4 positive cells per 400x high power field (Fig. 3).

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He was discharged from Urology and referred to regional Lupus and Rheumatology Centre to rule out systematic IgG4-RD. His autoantibody screen and protein electrophoresis were normal. FDG PET scan from base of skull to mid-thighs showed minor uptake in the tonsils (where he remains asymptomatic), with no evidence of IgG4-RD elsewhere.

3. Discussion

PFPs are benign, rare lesions of paratesticular structures accounting for 6% of paratesticular lesions. Majority of PFPs arise from tunica vaginalis with less than 10% originating from epididymis or spermatic cord. It can be stimulated by infection, trauma or inflammation. Recent literature has shown that PFPs can also be a manifestation of IgG4-RD.

Immunoglobulin IgG-related disease (IgG4-RD) is a multi-system fibro-inflammatory disease characterised by fibrosis and pseudotumours infiltrated with IgG4 plasma cells. This may or may not be associated with elevated plasma IgG4 levels with one-fourth of cases having normal blood IgG4 levels, as in our case. Our Medline search showed that approximately 11 cases were described in the English literature. The pathological staining of these tumours prior to 2003 did not commonly include IgG-4, suggesting this number could be higher.

Most IgG4 affected organs are salivary/lacrimal glands, pancreas, biliary tract and kidneys. However, any organ may be affected, and concurrent multi-system disease may also occur. Typical presentation is organ enlargement or mass, as in our case. It should be noted that the tissue infiltration/fibrosis can result in complications often seen in neoplasms such as obstruction/compression. In our case, all clinical investigations including FDG PET scan showed no systemic IgG-RD.

PFPs usually present with a unilateral painless mass or masses ranging from 0.5 to 8 cm. The peak incidence of PFP is the third decade although it may occur at any age. 50% of cases have a hydrocoele as an associated feature and 30% report a history of epididymo-orchitis-trauma. None of these features were present in our case.

Ultrasound scan typically reveals well-defined, homogenously hypoechoic lesions in an extra-testicular location. There may be small foci of calcification present. Appearance on MRI is low signal on T1, however, MRI is usually needed to rule out diagnoses of other paratesticular lesions such as malignancy, like in our case.

Management of PFPs is challenging due to their overall presentation mimicking malignancy in terms of location and consistency. Therefore, most patients undergo unnecessary radical orchietomies with the diagnosis of PFP made on postoperative histologic examination. In our case, PFP was clinically suspected as the multiple masses were not related to the testis/epididymis. Ultrasound and MRI scans were helpful in confirming the benign nature of paratesticular masses. However, the final diagnosis of PFP IgG-4 is made on postoperative histology. FDG half-body PET scan is essential in ruling out systematic IgG4-RD.

There is no consensus on a follow-up period in the literature. In this case, repeat scrotal US four months following excision showed no recurrence of previously seen pretesticular lesions. This patient has been discharged from urology and will be followed up by rheumatology for ongoing management of IgG4-RD.

4. Conclusion

Paratesticular Fibrous Pseudotumours are becoming increasingly recognised as a manifestation of IgG4-RD. PFP is a rare but important differential to consider in the pre and perioperative periods to avoid unnecessary radical orchietomy. Ultrasonography and MRI are imaging modalities that can be used in the preoperative period.

Informed consent

Obtained from patient.
Declaration of competing interest

None.

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