Testicular Vasculitis – Literature Review and Case Report in Queensland

Narelle Lintern  Nigel R. Johnson  Ian Mckenzie  Ben Martin
Urology Department, Royal Brisbane and Women's Hospital, Brisbane, Queensland, Australia

Key Words
Testicular vasculitis

Abstract
A 21-year-old male presented with acute onset, sharp right sided testicular pain. The testicle was removed with a histological diagnosis of testicular vasculitis. Anti-neutrophil cytoplasmic antibodies were negative. Although rare, males who present with acute onset pain should be screened for testicular vasculitis with a scrotal ultrasound and blood investigations including tumor markers and anti-neutrophil cytoplasmic antibodies.

Introduction
Vasculitis is defined as inflammation of the blood vessels, with the pathological consequence being destruction of the vessel wall, seen histologically as fibrinoid necrosis [1–4]. This disease process may be localized to a single organ or more commonly is generalised to the whole body. Muscular arteries may develop focal lesions leading to aneurysm formation and rupture of the vessel or stenosis with distal infarction [1–3].

The causes of many systemic vasculitis are unknown and therefore the current classification tools are based on clinical features and pathology [1, 3]. The most widely accepted classification system is based on dominant vessel size and association with anti-neutrophil cytoplasmic antibodies (ANCAs). This classification also reflects the therapeutic approach in treating the various diseases. Small to medium vessel disease responds well to immunosuppression with cyclophosphamide and corticosteroids whereas large vessel disease requires high dose steroids [1].

The pathogenesis of vasculitis is poorly understood. Three of the most common mechanisms of vascular damage are immune complexes, ANCAs and cell mediated T lymphocyte response [3]. An infectious agent can also act as the trigger. This includes direct microbial invasion as seen with rickettsia infection for example, causing a raised transendothelial migration and induction of inflammatory cytokines and chemokines. Immune complex deposition and propagation of the vasculitic process along with lymphocyte proliferation is seen with hepatitis C virus infection [4].

Single organ vasculitis (SOV) has been described as vascular inflammation of an isolated organ with no signs of vasculitis extending beyond this focus for at least 6 months [2]. SOV may present in areas that are common with systemic vasculitis, therefore it is difficult to assume that the initial surgical specimen or biopsy is not part of a systemic condition. The pathogenesis of isolated organ vasculitis is unknown, similarly why only one organ may be affected [5]. The majority of SOV is cured by surgical resection of the affected site. However, when this is not
possible, systemic management is required with a less favourable prognosis [2].

Often the diagnosis of testicular vasculitis is not immediately apparent as it often though to be a testicular neoplasm, torsion or infection; this is as the symptoms mimic these other conditions [2]. In the vast majority of cases local symptoms were seen in isolation with nil other haematological markers of vasculitis. In the study performed by Hernandez-Rodriguez et al. [2], it has been shown that testicular SOV is seen in middle aged men and presents with painful singular testicle and associated swelling.

Testicular vasculitis may be a SOV or part of a systemic vasculitis. Hernandez-Rodriguez et al. [6] retrospectively collected 7 cases over 22 years, combining these with 65 other cases reported in the literature. This study found that testicular vasculitis occurred in 0.003% of all testicular surgeries and could present as a testicular or epididymal mass that may be painful or painless. The most common sites affected are a single testicle, followed by the epididymis and spermatic cord.

Brimo et al. [7] retrospectively reviewed 19 cases between 1986 and 2009 and found that testicular vasculitis caused localized infarction, clinically mimicking a testicular tumor.

Generally orchidectomy is the diagnostic procedure of choice as suspected neoplasm is the most frequent preoperative diagnosis [6]. Polyrteritis nodosa is seen in almost two-thirds of patients with testicular vasculitis who have a systemic vasculitis concurrently [2, 6–8].

Case Report

A 21-year-old man of Asian descent presented to the emergency department with a five-hour history of acute onset, sharp right-sided testicular pain. This was on the background of intermittent pain over the preceding week. He was afebrile with no other systemic symptoms.

On physical examination he had a swollen, tender right testicle, with his right hemiscrotum being erythematous and warm to touch. No obvious lesion was felt in the scrotum.

Laboratory findings demonstrated a white cell count of 12.1 \times 10^9/l and neutrophils of 9.4 \times 10^9/l, with these results returning to normal range within 24 hours. Serum tumor markers in the form of beta human chorionic gonadotropin, alpha feta protein and lactate dehydrogenase were normal as was the full blood count, urea, creatinine and electrolytes, and liver function tests. Urine microscopy and culture was negative. The differential diagnosis at this stage included testicular torsion, infection or testicular tumor.

A scrotal ultrasound scan was performed demonstrating an oval-shaped lesion within the right testis. The oval lesion had an echogenic rim, with mixed echogenicity components within it. There was a well-defined lobulated outline, with several anechoic components at the periphery and extending to the rim of the testis. No internal vascularity to the lesion was demonstrated with the surrounding testis having normal vascularity. There was normal venous and arterial flow within the spermatic cord and the epididymis had a normal appearance. The impression from the ultrasound was a right testis lesion with mixed echogenicity internally, likely representing hemorrhage into a neoplastic lesion.

The patient was discharged once the pain had resolved, with a follow-up appointment in one week. On review, the swelling and erythema had resolved, while the right testicle was still tender. A p-ANCA and c-ANCA blood investigation was performed which were negative.

Due to the ultrasound findings being highly suspicious for neoplasm it was decided to proceed to a right inguinal orchidectomy and testicular prosthesis insertion. At the time of operation there was a discrete lesion palpable within the testicle, having developed since initial presentation.

Histological Findings

The testicle measured 40 x 20 x 25 mm and within the parenchyma there was a poorly circumscribed, hemorrhagic lesion measuring 20 x 16 mm, with a further area of discolouration measuring 12 x 6 mm. The epididymis and spermatic cord were both histologically normal. Throughout the testis there were multiple areas of hemorrhagic infarction, of varying duration. Vessels within the parenchyma showed fibrinoid necrosis associated with inflammatory infiltrate. The affected vessels were medium-sized muscular arteries with thrombi of varying ages seen in the specimens with re-canilisation of these vessels. Through-out the unaffected parenchyma, there was a decrease or absent spermatogenesis with increasing in stromal Leydig cells. There was no evidence of malignant cells.

Follow-up

The postoperative course was uneventful. An initial contrasted computed tomography scan of the chest, abdomen and pelvis performed for staging of presumed testicular malignancy.

This demonstrated no evidence to suggest distant changes of systemic vasculitis. Serum inflammatory markers in the form of C-reactive protein and erythrocyte sedimentation rate were both within normal limits. The patient was referred to a general physician for full evaluation to exclude systemic vasculitis.

Discussion

In evaluating this patient for a suspected scrotal neoplasm, the normal proforma of history, examination and investigations was conducted. The differential diagnosis for testicular neoplasms include seminomas, teratomas, germ cell tumours, dermoid cysts and Sertoli cell tumors [9].

Having a raised erythrocyte sedimentation rate or anaemia are more in keeping with systemic vasculitis and are not generally seen in SOV as in this case [6].
C-reactive protein or von Willebrand factor are potential markers for endothelial injury in systemic vasculitis, but may not be reflective in SOV [10].

This patient had negative ANCA’s which is in keeping with SOV. He had normal serum inflammatory markers. The systemic vasculitis such as Wegener granulomatosis, microscopic polyangitis and Churg-Strauss Syndrome are generally positive for ANCA [3].

Ultrasound of the testis is useful to exclude testicular abscess, but may fail to show any abnormality other than an existence of an echogenic mass [10]. The ultrasound from this patient was helpful in strengthening the case to undertake an orchidectomy.

There are no histological characteristics to distinguish between SOV and a systemic vasculitis [6]. The medium-sized vascular involvement is in keeping with the most common systemic vasculitis (polyarteritis nodosa) that is seen with testicular vasculitis [1, 6].

Conclusion

This case of testicular vasculitis appears to be the first reported in Queensland. Vasculitis, although rare, should be a differential diagnosis for a suspected testicular neoplasm. Once confirmed with histology postoperatively the patient should then be investigated for systemic vasculitis as there is no histological variation between SOV and the systemic form.

References