

Recurrent Unilateral Orchitis as a Presenting Symptom of Polyarteritis Nodosa

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KEY WORDS: vasculitis, orchitis, polyarteritis nodosa (PAN), testicle, fibrinoid necrosis

IMAJ 2015; 17: 585–586

Vasculitic lesions of the testicle often mimic solid tumors or infections and may lead to testicular infarction and hemorrhage. Testicular vasculitis can be part of a systemic vasculitis but may present as single-organ vasculitis. Some of these cases may later develop systemic involvement. Among systemic vasculitides, testicular vasculitis has been associated most frequently with polyarteritis nodosa (PAN). In fact, testicular pain or tenderness is included in the 1990 American College of Rheumatology (ACR) classification criteria of PAN [1]. Testicular vasculitis occurs in almost all PAN cases, but in most of them it is clinically silent.

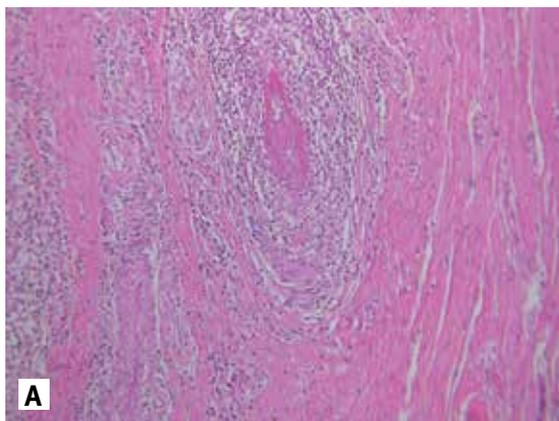
PAN presenting with longstanding recurrent orchitis as an isolated phenomenon is uncommon. We present a patient with isolated recurrent unilateral orchitis spanning more than 8 years, who later developed additional systemic features of PAN.

PATIENT DESCRIPTION

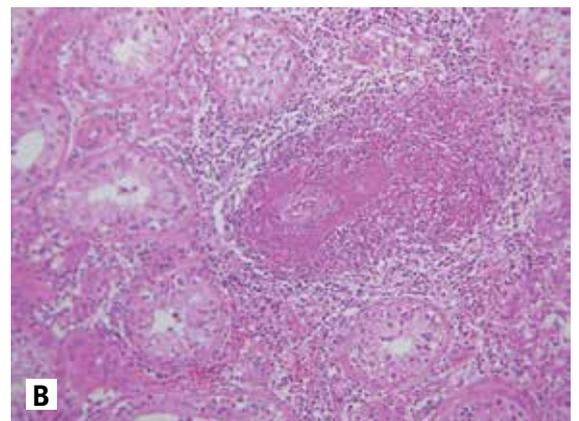
A 19 year old white male presented to the urology service with a 3 day history of painful swelling of the left testis. The rest of the physical examination was unremarkable. Following blood tests and imaging studies, culture-negative epididymo-orchitis was diagnosed; the patient was treated with antibiotics and he recovered fully. Six years later he was admitted with painful swelling of the same testis lasting almost a month. Epididymo-orchitis was diagnosed again and broad-spectrum antibiotics were administered, resulting in a slow recovery. Another two episodes recurred 2 and 2½ years later in the same testicle.

Due to these recurrent episodes left orchiectomy was performed. The pathological report revealed a 116 g testicle with necrotizing vasculitis in the fibrotic layer surrounding the testis and foci of vasculitis with fibrinoid necrosis within the testis [Figure 1A and B]. A rheumatology consultation at that time revealed an obese person, with no weight loss, gastrointestinal symptoms, myalgia or arthralgia. Blood pressure was normal. There was no livedo reticularis, but erythema nodosum lesions were noticed on both legs. There were no symptoms or signs of peripheral neuropathy. Blood tests showed elevated inflammatory markers (C-reactive protein and erythrocyte sedimentation rate). Blood counts and chemistry panel were within the normal range. Complement levels were normal and serological studies for hepatitis viruses were negative. A computerized tomography with angiography of the chest and abdomen was performed, but no vascular involvement was found.

Figure 1. [A] Active necrotizing vasculitis of an artery in epitesticular tissue, accompanied by mixed inflammatory infiltrate and thrombosis



[B] Active vasculitis with prominent fibrinoid necrosis and mixed inflammatory infiltrate in testicular tissues



A diagnosis of isolated (single-organ) PAN, involving one testicle, was made. The lesions of erythema nodosum did not respond to treatment with non-steroidal anti-inflammatory drugs, so treatment with prednisone was started in combination with azathioprine. The patient experienced an episode of acute pancreatitis. It was not clear whether this was due to vasculitis involving the pancreas, or was an adverse effect of azathioprine. Azathioprine was discontinued, but abdominal pain, fever, weakness and high levels of C-reactive protein persisted. Abdominal angiography did not show any micro-aneurysms or other signs of vasculitis. The addition of cyclophosphamide did not have any beneficial effect on these manifestations. The patient then received two doses of 1000 mg rituximab, achieving almost complete remission. Prednisone was gradually discontinued. He continues to experience recurrent episodes of mild cutaneous vasculitis without additional signs of systemic disease.

COMMENT

This patient represents a case of isolated testicular PAN, with recurrent flares over several years prior to diagnosis. Although most cases of vasculitis have systemic involvement, there have been reports of necrotizing vasculitis involving single

organs. Isolated or single-organ PAN is the term often used to describe these cases. Isolated PAN has been reported in the intestines, gallbladder, pancreas, breasts and urogenital organs of adult and adolescent males and females [2]. Clinical manifestations of an inflammatory reaction or laboratory markers of inflammation are often missing or mild, compared to cases with widespread PAN [3]. Patients with isolated PAN typically present with focal symptoms without systemic manifestations, often suggestive of tumors or infectious conditions [3], and the vasculitic lesions are found “incidentally” following excision or biopsy of these organs.

Sometimes orchitis is the presenting symptom of PAN [4]. However, in the case described above, the duration of symptoms prior to diagnosis was unusual. Despite that, this patient did not develop any systemic involvement over 8 years of recurring isolated testicular vasculitis. Hernandez-Rodriguez and colleagues [5] recently presented an analysis of 72 cases with testicular vasculitis reported in the medical literature. Half of them had isolated testicular vasculitis. The mean duration of testicular symptoms was 4.4 weeks before diagnosis. The most frequent presenting feature was painful testicular swelling or mass, occurring in 74% of these patients. Orchiectomy was performed in 62% prior to the final diag-

nosis. Genital complaints were followed by systemic symptoms in only 7% of the cases.

When feasible, isolated PAN is often cured by resection of the involved organ or tissue, without the addition of immunosuppressive therapy. However, some of these cases may later develop multifocal or systemic involvement [5]. Patients with single-organ PAN should be carefully evaluated initially and followed to exclude systemic involvement.

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