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Case Report

Unusual presentation of immunoglobulin A vasculitis with acute scrotal swelling and epididymo orchitis in a 5-year-old boy

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ABSTRACT

Immunoglobulin A (IgA) vasculitis, formerly known as Henoch-Schönlein purpura (HSP), is a small vessel vasculitis primarily affecting children. It commonly presents with palpable purpura, arthralgia, abdominal pain, and renal involvement. However, atypical presentations can occur, posing diagnostic challenges. Here, we present a case of IgA vasculitis in a 5-year-old boy with a unique manifestation of epididymitis as the primary presenting symptom. Scrotal ultrasound demonstrated Epididymo-orchitis with funiculitis. Skin examination revealed scattered palpable purpura over the lower extremities. Thus, a clinical diagnosis of IgA Vasculitis is made. The patient recovered with corticosteroids and supportive care.

Keywords: IgA vasculitis, Henoch-Schönlein purpura, Epididymitis, Orchitis, Purpura

INTRODUCTION

Immunoglobulin A (IgA) vasculitis, previously termed HSP, is a systemic vasculitis characterized by IgA immune complex deposition in small blood vessels. It predominantly affects children. Purpura, arthritis and abdominal pain are known as the "classic triad" of HSP.[1] However, a wide spectrum of clinical presentations exists, and atypical manifestations can pose diagnostic challenges. Here, we report an unusual case of IgA vasculitis presenting with epididymitis in a 5-year-old boy.

CASE REPORT

A 5-year-old previously healthy boy presented with a two-day history of acute scrotal pain and swelling. The pain was localized to the left hemiscrotum, exacerbated by movement. Further history revealed the patient had an episode of resolved sore throat with intermittent fever 8 days before this presentation which resolved 2 days back. There was no history of trauma or urinary symptoms. On examination, his left hemiscrotum was edematous, tender, and warm to the touch, with mild erythema on the overlying skin. The cremasteric reflex was present bilaterally. A review of systems was notable for a pruritic rash on his ankles, which the patient presumed to be due to recent mosquito bites, with no other exposures being identified. Patient also complained of mild ankle pain bilaterally.

Given the clinical suspicion of epididymitis, laboratory investigations were performed, revealing elevated inflammatory markers with a white blood cell count of 12,000/mm^3 (reference range: 4,500-11,000/mm^3) and a C-reactive protein level of 64 mg/l (reference range: <5 mg/l). The urinalysis result was normal, showing a specific gravity level of 1.014 and pH of 6.0, and being negative for leukocyte esterase, nitrates, blood, and protein.

Scrotal ultrasound demonstrated the following: The tail of the right epididymis is enlarged in size and shows

heterogeneous echotexture with increased vascularity and the Left testis is normal in size, measures 1.4×0.7×0.8cm, and shows minimal alteration of echotexture and mildly increased vascularity. The left epididymis is diffusely enlarged in size and shows heterogeneous echotexture with moderately increased vascularity. The left spermatic cord is diffusely thickened and shows heterogeneous echotexture with increased vascularity. Diffuse edema of the left hemiscrotal wall is noted. Mildly enlarged inguinal lymph nodes are noted on the left side. Minimal fluid is seen in the left Tunica vaginalis. Penis shows diffuse severe wall edema with increased venous flow, hyperemia consistent with epididymitis.

Further investigations including serological tests for autoimmune and infectious diseases were pursued. IgA levels were elevated at 480 mg/dL (reference range: 33-165 mg/dl), and serological tests for Streptococcus species, Mycoplasma pneumonia, and viral pathogens were negative. Skin examination revealed scattered palpable purpura over the lower extremities. Since the clinical presentation was atypical, a skin biopsy was performed which demonstrated leukocytoclastic vasculitis with IgA deposition on immunofluorescence, confirming the diagnosis of IgA vasculitis.

DISCUSSION

HSP is a condition characterized by the classic triad of symptoms, including purpura, arthritis, and abdominal pain.[1] However, urological involvement, in particular penile involvement, is exceedingly rare.[2]

The 90% of the children initially developed skin symptoms. Scrotal involvement in IgA vasculitis is rare, with few cases reported in the literature. However, the involvement of the scrotum may be the first symptom of HSP, and thus, a delayed appearance of the rash may impact the correct diagnosis.[3]

A scrotal or penile swelling with erythema or pain occurs in approximately 20% of males with HSP and is usually self-limited.[4]

Patients with HSP disease should undergo a comprehensive evaluation if they experience any scrotal swelling, erythema, or pain, which should include a thorough physical examination of the genital area, including palpation of the scrotal skin and its contents. Additionally, scrotal ultrasound and high-resolution color (and pulsed) Doppler imaging are necessary for proper diagnosis and treatment.[4]

Testicular torsion must be ruled out in cases with acute testicular or scrotal pain and swelling during the course of HSP. However, testicular torsion is unlikely to be bilateral or to have normal vascularity.[5] Therefore differential diagnosis of acute scrotum is essential to void urgent surgical exploration of the testis.[6]

The exact pathogenesis of epididymitis in IgA vasculitis remains unclear, but it may result from vasculitic involvement of the epididymal blood vessels.

Diagnosis of IgA vasculitis relies on clinical features supported by histopathological evidence demonstrating leukocytoclastic vasculitis with IgA deposition on immunofluorescence. Treatment is primarily supportive, focusing on symptomatic relief and management of complications, with NSAIDs being the mainstay for mild disease. Severe cases with renal involvement may require glucocorticoids or other immunosuppressive agents.

Patient course

The patient was administered corticosteroids orally at a dose of approximately 1 mg/kg/day. The patient was managed conservatively with NSAIDs for pain relief and supportive measures. His scrotal pain gradually improved over the following days, and repeat ultrasound showed resolution of epididymal changes. He remained asymptomatic during follow-up appointments, with no evidence of renal involvement.

CONCLUSION

We report a rare case of IgA vasculitis presenting with epididymitis as the primary symptom in a 5-year-old boy. Clinicians should be aware of the diverse clinical presentations of IgA vasculitis to facilitate prompt diagnosis and appropriate management. Usually, patients are managed with corticosteroids and supportive care. Long-term follow-up is often needed to monitor for potential relapses or complications.

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