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Management and particular ultrasonographic finding in childhood acute scrotal swelling and pain associated with Henoch-Schönlein purpura: A case report and review of the literature

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ABSTRACT

Henoch–Schönlein purpura (HSP), also known as immunoglobulin A (IgA) vasculitis, is a systemic vasculitis of unknown cause, mostly affecting children, in which complexes of IgA and components of complement are deposited on arterioles, capillaries, and venules. It is characterized by nonthrombocytopenic palpable purpura, abdominal pain, and arthritis. While renal involvement is well-known and frequent, scrotal involvement is uncommon and, therefore, not easy to be interpreted. Here, we report a case of a 3-year-old boy hospitalized for HSP, presenting with bilateral acute scrotum, and discuss diagnosis and treatment.

Key Words: Henoch-Schönlein purpura; acute scrotum; scrotal pain; ultrasonography; children.

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Introduction

Henoch-Schönlein purpura (HSP) is the most common childhood acute systemic vasculitis involving the skin, gastrointestinal tract, joint and kidneys, due to circulating immune complexes including IgA [1,2]. At the admission to the hospital, patients usually show rash, arthralgia, abdominal pain and urinary complaints [3]. HSP generally heals without complications, except for the case of renal failure which represent an uncommon event. The first case of male genital involvement related to this syndrome, was reported in 1960 by Allen et al [4]. The incidence of scrotal involvement is estimated to be between 2% and 38%. When this condition represents the first sign of HSP, the diagnosis becomes more difficult [2]. In this report, we describe a particular ultrasound (US) characteristics and the management of bilateral scrotal acute swelling and pain occurred in a 3-year-old child previously admitted during the treatment of to the hospital for HSP.

Case report

A 3-year-old caucasian children, with a history of pharyngitis in the previous 10 days, was admitted to a tertiary pediatric centre of L'Aquila with symptomatology suggestive of HSP. On physical examination, he showed a

palpable purpura that starting on the ankles and dorsum of the feet and extending up to the thighs, with associated joint pain, periocular edema and diffuse aching abdominal pain. His vital signs were stable, with a blood pressure of 127/96 mmHg (high for age and size), a pulse rate of 70 beats/min. Urinalysis showed proteinuria of 2.35 g/day without hematuria and creatinine clearance of 103.0 L/day. Laboratory test showed a moderate elevation of erythrocyte sedimentation rate and C-reactive protein, while Rheumatoid factor, antinuclear antibody, p-ANCA, c-ANCA, blood cultures, urethral cultures, hepatitis B and hepatitis C serology were negative. Full blood count and biochemical profile were unremarkable. The working diagnosis of HSP was adopted and intravenous hydration was started. After Two days the patient showed tender bilateral erythematous scrotal swelling which was greater on the left side. For the suspicion of an acute scrotum, a testicular US was urgently carried out showing the presence of bilateral marked edema of the skin and Dartos fascia with hypervascularity, without involvement of the deeper layers, testes, or epididymis and mild reactive hydrocele, without signs of torsion [Fig. 1].

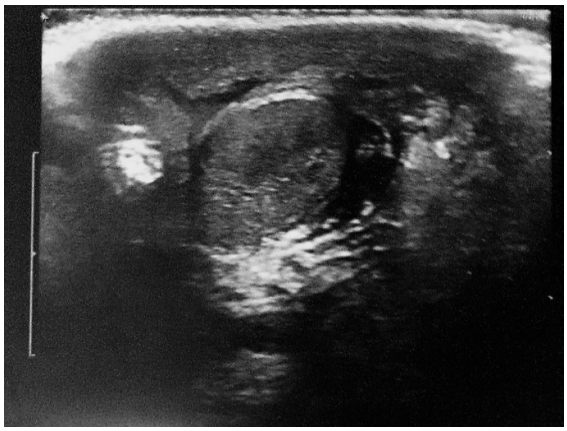

Fig. 1. Left scrotal ultrasound with evidence of very pronounced thickening of skin and Dartos fascia.

Once a surgical cause of pain was excluded, he was treated with a 6-week course of oral prednisone (starting by 2mg/kg/daily and later climbing), with a considerable clinical reduction of scrotal and abdominal pain with complete resolution of proteinuria. Ultrasound check showed normal finding after the first 10 days of treatment [Fig. 2]. At the end of treatment with prednisone, the patient was free from symptoms.

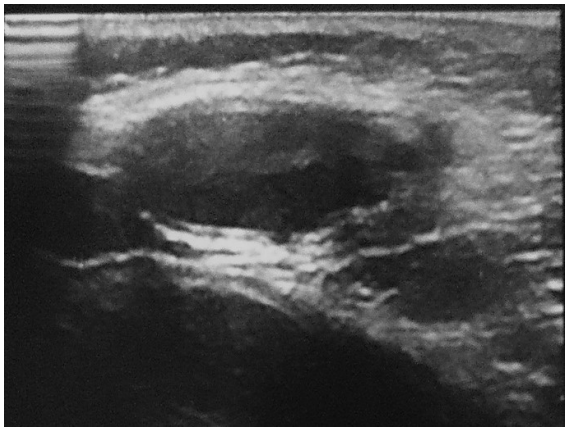

Fig. 2. Control after 10 days of steroidal therapy.

Discussion

Several studies conducted in patients with HSP, have highlighted the incidence of testicular and scrotal involvement. The reported incidence of scrotal involvement during HSP cases ranges from 2% to 38% and HSP with scrotal manifestation account for about 3% of all cases of acute scrotum [3-5]. Soreide et al, indicated that 80/603 patients (13%) with a diagnosis of HSP, had scrotal symptoms, and 16% of these, performed surgery; effective torsion was not identified in any patients [6]. Furthermore, Hara et al. [7] performed surgical exploration in 11/25 patients with HSP and did not identify testicular torsion in any of these patients. Ha and Lee reported that neurologic symptoms, localized edema, and high serum C3 levels

were significantly related to scrotal involvement in male patients with HSP [8]. In the literature it is advisable that cases of scrotal involvement in HSP should be managed conservatively, with a short-term administration of steroid therapy and/or antibiotics, without surgical treatment. US findings in HSP include scrotal wall thickening, epidydimal enlargement, and reactive hydrocele. Bilateral involvement is found in most cases; hence, HSP should be considered when the bilateral US findings are similar to those of inflammatory epididymitis [9]. Ben-Sira and Laor reported that in 13/87 boys with HSP the sonographic findings consistently included an enlarged, rounded epididymis, thickened scrotal skin, and hydrocele while testes were normal [10]. Another case report of HSP described a patient with acute scrotal pain and sonographic features included swollen, hypoechoic testes with hyperemia, which is suggestive of orchitis [11-12]. US features is essential to carry out a correct diagnosis excluding acute urological emergencies as the testicular torsion, characterized by twisting of the spermatic cord, altered blood flow based on the degree of torsion, increase in the size of the testis and epididymis, with early homogeneous echotexture, reactive hydrocele and reactive thickening of the scrotal skin. Regarding therapy, since HSP is a small vessel vasculitis, the rational use of prednisone is its ability in decreasing edema and the release of arachidonic acid, prostaglandins and leukotrienes produced during inflammation. For all these reasons, the role of corticosteroids was assessed in several studies evaluated in a meta-analysis of Weiss P.F. et al. which emphasizes the benefit of corticosteroid for both acute and chronic complications in the HSP [13].

Conclusion

In the light of the recent studies mentioned above, acute testicular pain in HSP is not a rare occurrence, for this reason the ultrasound finding of scrotum swelling in absence of testicular involvement is an alert sign and it must be recognized to avoid surgical interventions in the suspicion of a twist. Oral steroid therapy is a valid therapeutic approach that allows complete pain regression.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

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