

## Case for diagnosis <sup>\*</sup>

### Caso para diagnóstico

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#### CASE REPORT

5 year-old boy, of previous good health, was sent by his family doctor to the hospital's emergency department complaining of testicular pain which had developed over the previous few hours. Initial examination by the Pediatric Surgeon revealed violet-colored staining of both scrotal bags, more pronounced on the right, and with pain on manipulation of the right testis (Figure 1). Despite the presence of cremasteric reflexes we performed a Doppler testicular ultrasound which showed normal testes with increased blood flow, but ruled out testicular torsion. Multiple erythematous, maculo-papular violaceous circular lesions, between 2-8 mm in diameter, were also observed on both lower limbs and buttocks (Figure

2). The child also experienced pain at the tibio-tarsal joint which affected his ability to walk. He had no fever or gastro-intestinal pain. The previous week the patient had developed unmedicated acute rhinosinusitis. Henoch-Schönlein purpura (HSP), with inflammation of the testes (orchitis), was diagnosed. All the additional tests (blood pressure, blood platelet count and urine analysis) were normal. The child was treated with ibuprofen and monitored regularly both clinically and analytically (weekly blood pressure and urine tests) until resolution of the clinical symptoms, and thereafter sporadically for 6 months post first treatment.



FIGURE 1: Violet-colored testicular lesions with purpura on the thighs



FIGURE 2: Skin lesions on the thighs

Received on 21.12.2010.

Approved by the Advisory Board and accepted for publication on 24.03.2011.

\* Work performed at the Coimbra Pediatric Hospital, Coimbra Hospital Center (CHC), Coimbra, Portugal.

Conflict of interest: None / *Conflito de interesse: Nenhum*

Financial funding: None / *Suporte financeiro: Nenhum*

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## DISCUSSION

HSP is a leukocytoclastic vasculitis that mainly affects children aged between 5 and 10. Its prevalence ranges from 3.3 to 26.7 per 100,000 inhabitants, with a characteristic seasonal variation (with the highest number of cases during the winter months).<sup>1,3</sup>

Although the underlying etiology is unknown, it is generally acknowledged that HSP is an IgA-mediated immune complex vasculitis, probably triggered by infectious agents (50% of cases are preceded by upper respiratory tract infection) and other antigenic stimuli such as vaccines, drugs or even insect bites.

The classic tetrad of HSP is palpable purpura, arthritis or arthralgias (60-84%), abdominal pain (35-85%) and nephritis (21-54%), generally manifesting as skin lesions and arthralgia. Palpable purpura, which develops in all patients, is a mandatory criterion.<sup>2,3</sup>

Kidney involvement, characterized by hematuria (usually microscopic), proteinuria and rarely oliguria, hypertension, and azotemia,<sup>2</sup> is the most severe complication and may present at any stage of the disease.

Scrotal involvement is variable (2-38%) and may mimic testicular torsion, which can be ruled out by clinical examination with a cremasteric reflex test and, where necessary, a Doppler ultrasound test.<sup>4,5</sup> Testicular torsion is rarely the first presenting symptom. Patients may have testicular inflammation, bleeding or epididymitis, but a real torsion is excep-

tional. In a study of 120 children with HSP<sup>4</sup> scrotal involvement was in fact present in only 26.

The diagnosis of HSP is mainly clinical, with no specific laboratory diagnostic tests recommended. A hemogram with platelets and coagulation helps to exclude other types of purpura. Renal function (creatinine and urea) should be tested if the urine test shows changes. Urine screening together with blood pressure monitoring should be continued beyond the acute phase, since nephritis may occur at any time up to 6 months later.<sup>6</sup>

A skin or kidney biopsy should be reserved for atypical cases or when kidney involvement justifies it (e.g. to reveal leukocytoclastic vasculitis accompanying classic IgA deposits).

HSP treatment is primarily supportive and symptomatic, involving adequate hydration, rest and pain relief. Treatment can also be targeted at lessening the risk of other complications (corticosteroid therapy in the event of severe abdominal pain, or corticosteroids, azathioprine, cyclophosphamide and plasmapheresis for use in cases of severe nephritis).<sup>7</sup>

HSP in children has an excellent prognosis, with spontaneous resolution within an average of one month, although the disorder can recur in about one third of patients. Only a minority develop long-term complications, mainly as the result of kidney disease.<sup>2,6</sup> □

**Abstract:** Henoch-Schönlein Purpura (HSP) is the most common vasculitis in children. In the absence of significant renal disease it has an excellent prognosis. In the case described, HSP initially presented together with orchitis. This infrequent event required the exclusion of testicular torsion.

**Keywords:** Orchitis; Purpura; Purpura, Schoenlein-Henoch; Testis; Vasculitis

**Resumo:** A Púrpura de Henoch-Schönlein é a vasculite mais comum na idade pediátrica, tendo um prognóstico excelente na ausência de doença renal significativa. No caso descrito, a apresentação inicial cursou com orquite, o que não é frequente, obrigando a exclusão de torção testicular.

**Palavras-chave:** Orquite; Púrpura; Púrpura de Schoenlein-Henoch; Testículo; Vasculite

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How to cite this article/Como citar este artigo: Januário G, Santiago F. Case for diagnosis. Henoch-Schönlein Purpura. *An Bras Dermatol*. 2012;87(1):153-4.