

# Henoch-Schönlein Purpura Presenting With Orchitis: A Case Report and Review of the Literature

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## GOAL

To understand Henoch-Schönlein purpura (HSP) to better treat patients with the condition

## OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Discuss the clinical presentation of HSP.
2. Explain the presentation and diagnosis of HSP orchitis.
3. Describe the pathogenesis of HSP.

**CME** Test on page 88.

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*Henoch-Schönlein purpura (HSP) is the most common cause of nonthrombocytopenic purpura in children. The clinical picture is classically a cutaneous purpuric eruption of the legs and buttocks and infrequently the upper torso and extremities. Arthritis, gastrointestinal tract*

*symptoms, and nephritis are other common findings typically associated with the cutaneous findings. We present an unusual case of HSP with scrotal swelling and orchitis.*

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## Case Report

A 7-year-old boy presented to the Emergency Department with a 1-day history of bilateral scrotal rash, swelling, and pain of insidious onset. The pain had worsened progressively to the point that ambulation was difficult. The patient's only medical history was a circumcision as a newborn. There was no

history of hematuria, urinary tract infections, or trauma. The family reported the boy had been ill 10 days prior to presentation with intermittent fevers, nausea, emesis, and diarrhea, the latter of which had been bloody on at least one occasion. These symptoms had abated several days prior to his presentation to the Emergency Department.

Results of a physical examination revealed an anxious boy in no acute distress. He was afebrile with a blood pressure level and respiratory rate within reference range. The abdomen was soft and without masses, rebound tenderness, or guarding. On the genitalia, there was an extensive beefy-red rash confined to the scrotal skin, with several areas of petechia overlying the suprapubic fat pad. Both testis were tender to palpation but were descended bilaterally and positioned normally. No inguinal hernias were detected. Results of a cutaneous examination revealed a diffuse purpuric rash over the lower extremities (Figure). The rash did not extend to the buttocks or above the waist. There were no bullae or vesicles. On further questioning, the patient and his parents admitted noticing the rash 2 days prior to presentation to the Emergency Department.

Results of a urinalysis were within reference range. Results of a complete blood count revealed a hemoglobin level within reference range, but an elevated platelet count of  $500 \times 10^3/\mu\text{L}$ . The patient was sent for a scrotal ultrasound with Doppler flow studies, the results of which showed blood flow to both testes, edema of the scrotal wall, and hyperemia of the epididymides bilaterally.

The diagnosis of Henoch-Schönlein purpura (HSP) orchitis was made, and the patient was discharged on a therapeutic regimen of prednisolone 2 mg/kg per day, as well as instructions to elevate and apply ice packs to the scrotum. His pain improved dramatically over the next 24 hours; by 48 hours, the swelling and erythema had started to improve. He was seen in follow-up 2.5 weeks later, at which time the results of a repeat Doppler ultrasound of the testes showed no abnormalities. Results of an examination revealed his scrotal rash and tenderness had resolved completely, as did his lower extremity rash.

### Comment

HSP, a vasculitic syndrome of the small blood vessels, is the most common cause of nonthrombocytopenic purpura in children. The etiology of HSP remains unknown; however, its occurrence has been reported following upper respiratory tract infections. Elevated levels of antistreptolysin O antibodies have been associated anecdotally with HSP in a minority of cases. Several other infectious processes have been



Cutaneous purpuric eruption on the lower extremity.

reported in association with HSP; however, no specific organism appears to be found causative in HSP.<sup>1</sup>

The overall incidence of HSP is estimated to be 9/100,000 people, and males are affected more often than females.<sup>1,2</sup> HSP has been reported in patients aged 6 months to 86 years, but most cases occur in children (mean age, 6 years). Two thirds of affected children are younger than 8 years.<sup>1</sup> There appears to be some seasonal variation, with more cases occurring in fall through spring and fewer cases reported in summer.<sup>1,3</sup>

The most common clinical presentation of HSP is the cutaneous purpuric eruption that typically affects the legs and buttocks and, less frequently, the upper extremities. Bullae and vesicles are seen less commonly. This eruption is the initial presenting finding in more than half of patients with HSP.<sup>4</sup> The second most common clinical symptom found in HSP is arthritis, which can occur in 60% to 82% of

patients.<sup>3,4</sup> Involvement of the gastrointestinal tract with bleeding and abdominal pain occurs in 50% to 75% of patients and can be life threatening if the gastrointestinal bleeding is severe.<sup>3,5</sup> Intussusception is an uncommon manifestation of HSP.<sup>3,5</sup> Renal involvement occurs in 20% to 50% of patients with HSP. Nephritis is the only manifestation that can result in a chronic disease, with 1% to 5% of patients developing end-stage renal disease. Uncommon manifestations of HSP include scrotal disease, pulmonary disease, carditis, and central nervous system involvement.<sup>5</sup>

Scrotal involvement is an uncommon manifestation of HSP but has been reported to occur in 3% to 38% of cases. It may include scrotal rash, swelling, and either bilateral or unilateral pain. Allen and colleagues<sup>6</sup> were the first to describe scrotal involvement in HSP in a 1960 review that focused on renal complications of the disease. In the case series of 131 patients, 5 boys had scrotal involvement: 2 with "swelling and hemorrhage of the testes" and 3 with "marked scrotal hematomas." Clark and Kramer<sup>7</sup> reviewed the Mayo Clinic experience in a 1986 report; in a series of 87 males diagnosed with HSP, scrotal involvement was seen in 3.4%. Interestingly, orchitis has been the presenting symptom of HSP in only a handful of reported cases.<sup>7</sup> More commonly, the development of scrotal signs and symptoms parallels the development of the classic purpuric rash.

The clinical presentation of HSP orchitis may mimic testicular torsion, which can often be a difficult distinction to make based on clinical examination findings alone. Several reports have described patients diagnosed with HSP who also presented with signs of testicular torsion and were explored surgically. In almost all cases, the patients were found not to have torsion but rather to have vasculitic changes in the spermatic cord, testes, and necrotic hydatids of Morgagni.<sup>7-9</sup> There does exist one reported case in the literature of concomitant testicular torsion in the HSP syndrome; however, it occurred in a patient with physical examination findings that were classic for torsion (unilateral tenderness with a testis that was "drawn up and tense") and whose pain had been of sudden onset.<sup>10</sup> Results of exploration revealed the testis to be torsed and found HSP orchitis (evidence of vasculitis and hemorrhage of the morgagnian cyst).<sup>10</sup>

Most reported cases of HSP orchitis in the literature occurred before the widespread availability of Doppler ultrasound. Since that time, the ability to differentiate torsion from HSP orchitis based on imaging has matured. Ben-Sira and Laor<sup>11</sup> viewed the sonographic findings in a series of boys with

acute scrotal pain and HSP and found that most had epididymal enlargement, scrotal skin thickening, hydrocele, and normal-appearing testes with blood flow that was either within reference range or increased. In addition, other authors have described gray-scale ultrasonographic findings of hypoechogenicity and hypovascularity of the testicular parenchyma, which may represent areas of necrotic foci.<sup>12,13</sup> In these reports, patients found to have intact testicular blood flow did well with observation and, in some instances, with a therapeutic regimen of oral or intravenous steroids.<sup>11-13</sup>

Other uncommon genitourinary findings of HSP include priapism, purpura of the penile shaft, hemorrhagic cystitis, renal colic, ureteral calcifications, bladder hematomas, and stenotic ureteritis.<sup>14-17</sup>

HSP typically is acute and self-limited, running its course in about 4 weeks; however, 30% to 40% of patients can experience recurrences.<sup>5</sup> Short-term morbidity depends on the presence and severity of gastrointestinal tract involvement, and long-term prognosis primarily is dependent on the extent of renal involvement.<sup>5</sup> Supportive therapy is all that typically is necessary, though systemic steroids often are used to treat the associated arthritis, skin findings, and orchitis, when present. However, studies have shown that these symptoms typically resolve with or without the use of corticosteroids; additionally, the use of corticosteroids does not have a significant effect on the duration of the disease or the frequency of recurrences.<sup>3,5,18,19</sup> The use of high-dose steroids in severe nephritis and gastrointestinal tract hemorrhage is more established.<sup>3,5,18,19</sup>

The pathogenesis of HSP results from a leukocytoclastic vasculitis due to immunoglobulin A (IgA) deposition in vessel walls. IgA also has been found to be elevated in the serum of patients with HSP; additionally, IgA circulating immune complexes have been found. IgA1 and IgA2 are the 2 subclasses of IgA, and IgA1 is the predominant subclass of serum IgA; secretory IgA is made up of equal parts of IgA1 and IgA2. The pathology of HSP involves only IgA1 and may be a result of aberrant glycosylation in the hinge region of IgA1.<sup>1,20</sup> Even though HSP is the most common vasculitis in children, the pathogenesis still is not completely understood.

The typical clinical picture of a patient with HSP is a purpuric eruption on the lower extremities and buttocks with arthritis, gastrointestinal tract symptoms, and varying amounts of renal disease. We report the presentation of HSP with an acute scrotum. In our case, based on the presence of bilateral scrotal pain, classic cutaneous findings, and Doppler and gray-scale ultrasound findings consistent with

orchitis, a diagnosis of HSP orchitis was made and unnecessary surgery was avoided. Furthermore, the use of a short course of oral steroid therapy seemed to hasten the relief of the patient's scrotal and cutaneous symptoms. This report illustrates an unusual presentation of HSP.

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