

Acute scrotum in Henoch-Schönlein purpura: fact or fiction?

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Henoch-Schönlein purpura (HSP) is the most common systemic vasculitis of childhood. It is usually seen following upper respiratory tract infections. It rarely involves the genital system or causes scrotal edema. With this report, we wanted to bring a different perspective to this clinic of acute scrotum. Herein, we present two HSP patients admitted to our clinic with scrotal involvement, with a review of the literature.

Key words: Henoch-Schönlein purpura, scrotal involvement, testicular torsion.

Henoch-Schönlein purpura (HSP) is a leukocytoclastic vasculitis with unknown etiology that affects primarily the skin and gastrointestinal system, joints and kidneys. It is the most common vasculitis of childhood, and the incidence between 4-6 years of age is 20/100,000¹. Acute scrotal pathologies are rarely seen in HSP, and only a few cases have been reported in the literature. Two boys who were admitted to the emergency service with the complaint of scrotal pain and diagnosed with HSP are discussed herein with a review of the literature.

Case Reports

Case 1

A seven-year-old boy admitted to our emergency service with sudden-onset pain, edema and erythema of the scrotum. It was learned from his parent that he had been prescribed prednisolone treatment due to an upper respiratory tract infection with a diagnosis of HSP one month before. Diagnosis of HSP was made on the basis of severe abdominal pain, purpuric lesions on his lower limbs and microscopic hematuria. Prednisolone was used for two weeks at a dose of 2 mg/kg/day, and it was ceased following a tapering over a week. The patient's pain was reported to occur approximately 6 hours before admission, with gradually increasing severity.

The physical examination was normal except for hyperemic, edematous and tense scrotum (Fig. 1). Additionally, he had purpuric skin rashes on the gluteal region, feet and back of the legs, which did not blanch when pressed. Laboratory findings were normal and there was no leukocytosis. The scrotal color Doppler ultrasound (CDUS) revealed that arterial flows could not be evaluated clearly in each testicular parenchyma.

Case 2

A six-year-old boy was admitted to the emergency service with pain and erythema of the scrotum that began 8 hours before. The



Fig. 1. Hyperemic and edematous scrotum at the time of admission of the child (Case 1).



Fig. 2. Both the epididymis and the spermatic cord were slightly edematous, but no testicular torsion was found (Case 1).

patient also had purpuric skin rashes on the gluteal region. Diagnosis of HSP was made by palpable purpura on his lower limbs and gluteal region and abdominal pain. Histopathology of the skin biopsy specimen had proven leukocytoclastic vasculitis; however, steroid treatment was not started for this patient. The scrotum was hyperemic and edematous on the physical examination. Vital signs and biochemical tests were normal. In DUS, arterial flow was not seen in the left testicle and was reported to be minimal in the right testicle.

One month before, both patients had been diagnosed as HSP with clinical findings and skin biopsies. Testis torsion could not be excluded in each case based on the presented laboratory and physical examination findings. Scrotal exploration was determined after informed consent was obtained from the children's parents. After performing a vertical incision from the scrotal raphe in each patient, edema was noticeable when the skin and subdermal tissues were passed through. Upon exploration, the testes were normal in macroscopic appearance. The epididymis and spermatic cord were slightly edematous (Fig. 2). Although there were no observed testicular torsion findings, bilateral testicular fixation was done in both cases. These two patients were discharged 24 hours after the surgery, with no complications. At the first month, Case 1 had no symptoms and his scrotal examination was normal (Fig. 3). CDUS revealed that arterial flow was within the physiological range.

Discussion

Henoch-Schönlein purpura (HSP) is the most common systemic vasculitis of childhood. Although many related factors have been defined, the underlying etiological mechanism is still unclear. Though the histopathological diagnosis of leukocytoclastic vasculitis is mandatory, at least two of the criteria established by the American Rheumatology Association must be met for the clinical diagnosis. These criteria are hemorrhagic skin lesions not accompanied by thrombocytopenia, age below 20 years, intestinal angina, and visualization of granulocyte accumulation in the arterial and venule walls in tissue biopsies (leukocytoclastic vasculitis)². Purpura or petechiae (mandatory) with lower limb predominance and at least one of the four following criteria -- abdominal pain, histopathology, arthritis or arthralgia, and renal involvement -- should be included for the definition of HSP as described by EULAR\PRINTO\PRES³.

The differential diagnosis of cases with acute scrotum in childhood includes testicular torsion, appendix testicle/epididymis torsion, epididymitis/orchitis, hernia/hydrocele, trauma/sexual abuse, testis tumors, idiopathic scrotal edema, cellulitis, and vasculitis. One of the other vasculitides mimicking acute scrotum is polyarteritis nodosa (PAN). The clinical presentation of testicular PAN may include pain, swelling or local mass, leading to a preoperative diagnosis of acute orchitis, torsion or tumor⁴. In the case of orchitis, scrotal pain and swelling start slowly and progression occurs in days. Nausea and vomiting usually accompany these symptoms⁵. In our cases, testicular pain and edema had an immediate



Fig. 3. Physical appearance of the scrotum at the first month follow-up (Case 1).

onset. However, neither of our cases had nausea or vomiting.

It is considered that there is a maximum 4-8 hour span in order to save testicular function in testicular torsion⁵. Only 3% of patients attending with acute scrotum have HSP-related scrotal involvement⁶. The history of patients presenting with scrotal involvement should be assessed well, and the physical examination should be performed carefully. Testicular pain that is accompanied by nausea and vomiting and has a sudden onset favors torsion at first consideration. However, nausea and vomiting also exist among half of the patients diagnosed with HSP⁷. Tight scrotum, spermatic cord shortening and cremasteric muscle spasm are the manifestations of torsion, and surgical exploration is recommended in these situations⁸. Scrotal pain may occur before HSP rash or long after the disappearance of the rash. It is hard to relate scrotal involvement to HSP in patients who do not present with rash or arthralgia. Most of these cases undergo surgical exploration⁹.

Sonographic imaging (gray scale or color Doppler) should be done in patients with suspected testicular torsion. Widening of the epididymis, thickening of scrotal skin and hydrocele are sonographic findings in symptomatic patients with HSP. Most of the patients may have minimal intratesticular flow alterations. Tc-99m radionuclide evaluation may be useful for confirmation of suspicious cases. Katz et al.⁷ reported one child with severe scrotal pain, hemorrhage and swelling who underwent unnecessary scrotal exploration; they avoided operation in four additional patients with similar symptoms after a testicular scintiscan demonstrated good blood flow.

According to the literature, a majority of the patients with HSP presenting with scrotal involvement undergo surgical exploration. This may stem from the insufficient arterial flow showed by CDUS preoperatively. However, the reliability of CDUS for the differential diagnosis of torsion is debatable, when it is considered that almost all of the explored patients had no torsion. Only one case of real torsion was reported in HSP in 1974. Fukuda et al.¹⁰ reported an idiopathic testicular infarct case that may have been related with HSP and mycoplasma infection. Since both HSP and

mycoplasma infection may cause testicular torsion and infarct, testicular torsion could not be excluded in that case; hence, orchiectomy was performed. Søreide¹ indicated that 80 of 603 cases (13%) presenting with a diagnosis of HSP had scrotal symptoms, and 16% of these patients underwent surgical exploration due to scrotal symptoms. Real torsion was not identified in any of these patients. Hara et al.¹¹ performed surgical exploration in 11 of their 25 HSP cases and did not identify testicular torsion in any of these patients. Scrotal symptoms were found to recover within one month in most of the patients who were treated conservatively.

Since both clinical and ultrasonographic evaluations were deemed insufficient to determine the exact diagnosis of acute scrotum in the presented children with HSP preoperatively, scrotal exploration with intraoperative evaluation of the testes was performed in our cases. In conjunction with the literature, we recommend avoiding surgical exploration in HSP patients with scrotal involvement. Additionally, the spectrum of these scrotal signs might be redefined from acute scrotum to scrotal involvement.

REFERENCES

1. Søreide K. Surgical management of nonrenal genitourinary manifestations in children with Henoch-Schönlein purpura. *J Pediatr Surg* 2005; 40: 1243-1257.
2. Mills JA, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura. *Arthritis Rheum* 1990; 33: 1114-1121.
3. Ozen S, Pistorio A, Mlusan S, et al. EULAR/PRINTO/PRES criteria for Henoch Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria. *Ann Rheum Dis* 2010; 69: 798-806.
4. Rubin MF, Ergas D, Sthoeger ZM. Limited polyarteritis nodosa of the male and female reproductive systems: diagnostic and therapeutic approach. *Ann Rheum Dis* 2002; 61: 362-364.
5. Gatti JM, Patrick Murphy J. Current management of the acute scrotum. *Semin Pediatr Surg* 2007; 16: 58-63.
6. Clark WR, Kramer SA. Henoch-Schönlein purpura and the acute scrotum. *J Pediatr Surg* 1986; 21: 991-992.
7. Katz S, Borst M, Seekri I, Grosfeld JL. Surgical evaluation of Henoch-Schönlein purpura. Experience with 110 children. *Arch Surg* 1991; 126: 849-853.

8. Caldamone AA, Valvo JR, Altebarmakian VK, Rabinowitz R. Acute scrotal swelling in children. *J Pediatr Surg* 1984; 19: 581-584.
9. Singer J, Kissoon N, Gloor J. Acute testicular pain: Henoch-Schönlein purpura versus testicular torsion. *Pediatr Emerg Care* 1992; 8: 51-53.
10. Fukuda S, Takahashi T, Kumori K, et al. Idiopathic testicular infarction in a boy initially suspected to have acute epididymo-orchitis associated with mycoplasma infection and Henoch-Schönlein purpura. *J Pediatr Urol* 2009; 5: 68-71.
11. Hara Y, Tajiri T, Matsuura K, Hasegawa A. Acute scrotum caused by Henoch-Schönlein purpura. *Int J Urol* 2004; 11: 578-580.