An Unusual Complication of Henoch-schönlein Vasculitis in an 11-year-old Boy: Global Testicular Necrosis Mimicking Testicular Torsion

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ABSTRACT

Henoch-schönlein purpura (HSP), an anaphylactic vasculitis, is characterized by palpable purpuric rash and collection of immunglobulin A (IgA) around small vessels. Its clinical triad is purpura, arthritis and abdominal pain. Henoch-schönlein purpura usually involves the kidney, gastrointestinal tract, joints and rarely, non-renal genitourinary organs. Except all rare cases that progress to renal failure, HSP is a disease which resolves without any sequelae. Henoch-schönlein purpura may mimic surgical conditions such as appendicitis and testicular torsion, leading to unnecessary laparotomy and explorations. Herein, we report a case of an 11-year-old boy, diagnosed on the 12th day of HSP and operated on with suspicion of testicular torsion, for which surgical exploration did not reveal any torsion but global necrosis of the testicle.

Keywords: Henoch-schönlein vasculitis, testicular necrosis, testicular torsion

Complicación Inusual de la Vasculitis de Schönlein-Henoch en un Niño de 11 Años: Una Necrosis Testicular Global que Semeja una Torsión Testicular

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RESUMEN

La púrpura de Henoch-Schönlein (PSH), una vasculitis anafiláctica, se caracteriza por erupción purpúrea palpable y acumulación de inmunoglobulina A (IgA) alrededor de los vasos pequeños. Su tríada clínica es púrpura, artritis y dolor abdominal. La púrpura de Henoch-Schönlein implica generalmente el riñón, el tracto gastrointestinal, las articulaciones, y en raras ocasiones, los órganos genitourinarios no renales. Con excepción de todos raros casos que progresan hacia la insuficiencia renal, PSH es una enfermedad que se resuelve sin ninguna secuela. La púrpura de Schönlein-Henoch puede semejar condiciones quirúrgicas tales como apendicitis y torsión testicular, dando lugar a que se realicen laparotomías y exploraciones innecesarias. Aquí reportamos el caso de un niño de 11 años de edad, diagnosticado al 12mo día de PSH, y operado por sospecha de torsión testicular, en que la exploración quirúrgica no reveló torsión alguna, sino una necrosis global del testículo.

Palabras claves: Vasculitis de Henoch-Schönlein, necrosis testicular, torsión testicular

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INTRODUCTION

Henoch-schönlein Purpura (HSP), is a leukocytoclastic (anaphylactic) vasculitis in organs such as kidneys and skin and it appears after an upper respiratory tract infection and is characterized with a palpable purpuric rush and collection of immunglobulin A (IgA) around small

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vessels. This may cause bleeding. It is more common in children than adults and males are more affected than females (1).

Symptoms of the disease vary depending on the location of the accumulation. The skin symptoms include purple colored rashes called purpura, on the lower legs and hips. These rashes do not fade when pressed. Affected joints may also have pain. In the case of renal involvement, there is haematuria and proteinuria. Although the exact cause of this disease is unknown, there is often a viral or bacterial upper respiratory tract infection in the patients' history. The clinical triads include purpura, which is present in all cases, arthritis and abdominal pain. Arthritis and abdominal pain have been reported in 74% and 51% of all cases, respectively (2).

Penile and testicular involvement are very rarely reported (3, 4). Testicular torsion tends to occur most commonly during puberty (5, 6).

CASE REPORT

An 11-year-old boy was admitted with complaints of swelling in his arms and legs while playing sports. Pink rash on his hips and legs accompanied by joint pain, occurred the next morning.

He had a five-day history of upper respiratory tract infection. On his physical examination, purpuric rash was seen on hips as well as lower extremities; the rash was less petechial on the arms. Bilateral knee swelling was present and there was tenderness and warmth in the right-elbow. The patient reported pain in the left-ankle and there was also warmth.

Later, he developed tenderness on the abdomen. There was no organomegaly. The tonsils were hyperaemic and hypertrophic. Rales on auscultation of both lungs were present at baseline. Arterial blood pressure was 100/60 mmHg, body temperature was 36 C° and pulse was 106/minutes. Complet blood count, biochemistry, C-reactive protein (CRP), antistreptolysin O (ASO), erythrocyte sedimentation rate (ESR), urogram, urine culture, abdominal X-ray, prothrombin time (PT), partial thromboplastin time (PTT), immunoglobulins (IgA), C3, C4 and auto-antibodies were investigated.

White blood cells: (WBC): 16.2/mm^3, haemoglobin (Hb):15.4 g/dL, platelet count (PLT): 368 000/mm^3, PT: 31.8, PTT: 15.9, sodium (Na): 137 mEq/L, potassium (K): 3.86 mEq/L, calcium (Ca): 10.29 mg/dL, glucose: 98 mg/dL, blood urea nitrogen (BUN): 12 mg/dL, creatinine: 0.47 mg/dL. Creatinine clearance was calculated as 88 mL/min/1.73 m^2 according to Schwartz Formula. There was a moderate proteinuria (38 mg/kg/day). Microscopic haematuria was present. C_3:139.4, C_4:35.6, IgA: 179, IgE: 64, IgG: 133.8, IgM: 157. Antinuclear antibodies were negative: ANA (-), AMA (-), anti dsDNA (-). The stool occult blood test was negative. Abdominal X-ray and ultrasonography showed no remarkable findings. The patient was started on methylprednisolone 2 mg/kg/day with the diagnosis of HSP nephritis.

The renal biopsy revealed basal membrane thickening, focal atrophic changes, presence of intense glomerular IgA and IgG deposition on immunofluorescence, IgM (-), C3 (-), C4 (-), C1q (-) was identified and the diagnosis of Henoch-schonlein nephritis was confirmed.

On the 12th day of the patient's admission the left testis became tender. On day 13, the patient reported severe scrotal pain. The temperature increased in the left testis, stiffness, redness and tenderness were present upon physical examination. There was no history of trauma. Scrotal Doppler US could not show any blood flow on the left. An emergent surgery was planned with the diagnosis of testicular torsion. Surgical exploration did not reveal any torsion but necrosis of the spermatic cord and epididymis. However, the testicle appeared swollen, darker in colour and necrotic. The incision was made in order to assess the viability of the testicle and bleeding did not occur (Fig. 2).

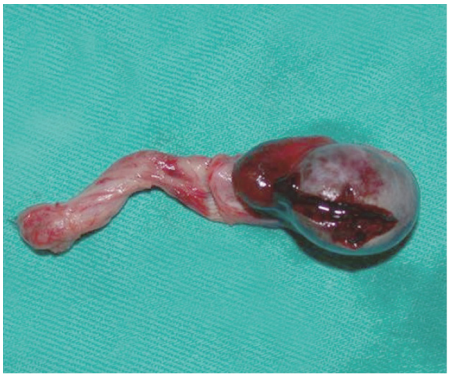

Fig. 2: Shows macroscopic findings of the necrotic left testis indicating no obvious torsion of the spermatic code and testis.

A radical orchiectomy was performed. Histological findings indicated that the affected testis and epididymis had haemorrhagic necrosis and infarction (Fig. 1A). Haematoxylin-eosin–stained sections showed necrotizing leukocytoclastic vasculitis of small vessels (Fig. 1B). Neutrophilic infiltration and nuclear debris were seen in and around the necrotic vessel.

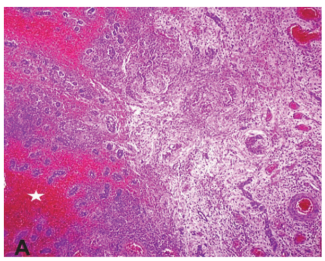

Fig 1 (a)

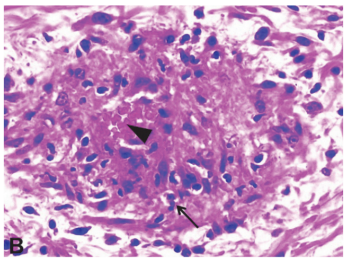

Fig 1 (b)

Fig.1 (A, B): (HE X 20) Haematoxylin-eosin–stained sections from testis and epididymis showing haemorrhagic necrosis and haemorrhagic infarction (star) (A).
(HE X 40) necrotizing leukocytoclastic vasculitis are seen of a small vessel. Neutrophilic infiltrate (black arrow) and nuclear debris (arrow head) was seen in and around necrotic vessel (B) .

DISCUSSION

Henoch-schönlein purpura, which is characterized by non-thrombocytopaenic purpura is the most common systemic vasculitis of unknown origin in children (7). Annual incidence in different countries is between 10 and 20 cases per 100 000 children (8). Henoch-schönlein purpura usually involves the kidney in 20–60% of the patients (9–11), the gastrointestinal tract in 38–75% (12, 13), joints 74% (2, 14) and non-renal genitourinary (NRG) manifestations in 6% of the patients (6).

Except all rare cases that progress to renal failure, HSP is a disease which resolves without any sequaela. Corticosteroids are used to treat abdominal pain, subcutaneous oedema and nephritis, even though there is no agreement about their effectiveness (15). Henoch-schönlein purpura may mimic a surgical condition such as appendicitis and testicular torsion, leading to unnecessary laparotomy and explorations (16–18). On the other hand, it sometimes causes surgical complications such as intussusception, small bowel perforation (19), gastrointestinal bleeding, gastric ulcer (19, 20) and some NRG problems. Complications in the NRG involvement have a cumulative incidence of 13.3% of scrotal symptoms (mainly, oedema and pain) in boys with HSP (6) including: impairment of the scrotum and testes together with painful swelling, epididymo-orchitis or orchitis, priapism and penile involvement (3), ureteritis with associated hydronephrosis, haematoma of the bladder wall and haemorrhagic spermatic cord, thrombosis of the spermatic veins (6) and testicular infarction.

Testicular infarction is a very rare condition and common causes are trauma, epididymitis, thrombosis and torsion (21, 22). Both global and segmental testicular infarctions are rare conditions in the testicles. Global testicular infarction may occur as a result of epididymo-orchitis (2, 22, 23). Segmental testicular infarction may be evident with sickle cell anaemia, after varicocelectomy, secondary to epididimo-orchitis or idiopatic (24).

Thrombosis of the spermatic veins are very rare in childhood and are mostly associated with malignancy (25). Testicular trauma is one of the causes of infarction and can be identified with recent history of trauma, especially in the abdominal or testicular area. Henoch-schonlein purpura epididymitis and orchitis may mimic testicular torsion and ultrasound is the gold standard for diagnosis (7). Only three cases of HSP with testicular or appendicular torsion, have been reported previously (7). Most explored scrotum in HSP show no testicular torsion, good blood supply and oedematous testis without necrosis. The blood flow of the testis on Color Doppler US shows normal or high levels in case of HSP, while it shows lower levels or none at all in case of testicular torsion. Although the Color Doppler US is a powerful tool to distinguish testicular torsion from other aetiologies of acute scrotum (26), there are some instances in which Doppler US may show inconsistent findings when compared with surgical exploration (16, 27). It frequently returns a false-positive result due to the oedema of the surrounding tissue and a correct diagnosis depends on the experience of the radiologist. Therefore, Soreide K *et al* suggest more conservative attitude toward acute scrotal symptoms in HSP (6).

There are some dilemmas about the testicular involvement. First, 20–36% of the patients with testicular involvement are operated on, however, the surgical exploration for testicular torsion usually turns out to be unproductive (6). Second, is the case of testicular torsion with necrosis. However, true torsion of the testis in HSP has only been reported three times since 1974 (28). Third is, as in our current case, testicular necrosis without torsion, which has only been reported once so far (21).

Some research indicates that HSP can cause ischaemia, necrosis and perforation in a lot of systems and this shows that, even rare, this disease can lead to life threatening complications as well as organ losses besides having a nature of healing without having a sequel. The aggressive course of vasculitis may be the cause of this clinical manifestation.

In contrast to popular belief, although scrotal involvement is high in HSP, testicular torsion is almost negligible or non-existent and it can appear unexpectedly as testicular necrosis without torsion as it did in our case.

According to the authors opinion, if testicular torsion symptoms occur during the clinical course of HSP in patients with an acute scrotum, Doppler US findings may

be misleading in regards to how much experience that the radiologists have. Although the possibility of scrotal involvement in HSP is very high, testicular torsion is less than expected upon surgical exploration. Therefore, it should be kept in mind that as a rare complication, it is possible to see testicular necrosis in HSP patients without the presence of torsion and despite sufficient blood supply.

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