Diagnosis of Henoch-Schonlein purpura in a child presenting with bilateral acute scrotum

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Abstract. Schonlein-Henoch syndrome (HSP) is one of the manifestations of acute systemic vasculitis related to circulating immune complexes including IgA. It usually involves the kidney, gastrointestinal tract, joints and skin. Except for rare cases that progress to renal failure, it is a disease which heals without complications. The first case of male genital involvement in this syndrome was reported by Allen et al in 1960. Since then, several reports of this condition with an incidence of scrotal involvement varying from 2 to 38% have been described. The involvement of the male genitalia presenting as the only initial manifestation of SHS is so unusual that the diagnosis can easily be missed. In literature rare cases have been reported in which acute scrotum was the initial presenting symptom in patients affected by HSP. We report a case of HSP presenting as acute scrotum in a 5-year-old boy hospitalized for fever and viral bronchopneumonia. (www.actabiomedica.it)

Key words: Purpura, acute scrotum, Schonlein-Henoch syndrome

Schonlein-Henoch syndrome (SHS), also known as anaphylactoid purpura, one of the manifestations of systemic vasculitis, usually involves the skin, gastrointestinal tract, joints and kidney (1). The involvement of the male genitalia presenting as the only initial manifestation of SHS is so unusual that the diagnosis can easily be missed. In literature rare cases have been reported in which acute scrotum was the initial presenting symptom in patients affected by HSP (2-4). We report a case of HSP presenting as acute scrotum in a 5-year-old boy hospitalized for fever and viral bronchopneumonia.

Case report

A five year old boy presented with a six-day history of cough and rinorrea. Three days before he had presented fever (39°C), with a successive spontaneous regression, without the use of medications, in association with bilateral cervical lymphoadenopathy. At admission on physical examination he showed a temperature of 36°C, bilateral cervical lymphoadenopathy and coarse breath sounds bilaterally. No hepatomegaly or splenomagaly were present. Laboratory test revealed a leukocyte count of 8.9 x 109 per liter (8,800 per μ l), with 0.24 neutrophilis, 0.63 lymphocytes, 0.06 monocytes, 0.04 eosinophilis and 0.01 basophilis. The haemoglobin level was 13.6 grams per dl and platelet count was 317 x109 per liter (317,000 per µl). Blood chemistry values resulted in normal values, with the exception of lactate dehydrogenase (372 U per liter, normal value 100 to 260). C-reactive protein was 22 mg/dl (normal value 0-5) and erythrocyte sedimentation rate was 20 mm/hour (0-15). Chest-X ray showed increased parahilar peribronchial infiltration and a neck ultrasound examination evidenced diffuse cervical lymphoadenopathy with reactive characteristics. Abdominal ultrasound examination resulted negative. Serological tests for Brucella species, Salmonella typhi and paratyphi, Epstein Barr virus, cytomegalovirus and Toxoplasma gondii resulted negative. Mycoplasma

antibody and cold hemagglutinin titers were within the normal values. Urinalysis was normal. Bilateral painful scrotal swelling with ecchymosis was observed at the third night. Scrotal sonography was immediately performed and it evidenced a good blood supply to the testis. Scrotal nuclear scanning confirmed no testicular torsion and bilateral epididymoorchitis was considered. Four days later the patient presented purpura on his limbs and trunk and intermittent abdominal and knee pain developed thereafter. Hemogram was rechecked and the haemoglobin level was 13.8 grams per dl, hematocrit 36%, platelet count 389,000 per µl. Prothrombin time, and partial thromboplastin time were within the normal values. IgA serum concentration resulted normal. Urinary protein excretion and faecal occult blood were negative. HSP was diagnosed by clinical elements and without histological examination or immunoflorescence staining for IgA. Intravenous dexamethasone was administered (0.5 mg/Kg/day). The symptoms gradually subsided and 7 days later an oral prednisolone treatment (1 mg/Kg/die) was started. He was discharged in stable conditions and knee pain and purpura flared up a few times during the 2-month period after discharge. Corticosteroid treatment was continued for two months.

Discussion

HSP is one of the manifestations of acute systemic vasculitis related to circulating immune complexes including IgA. It usually involves the kidney, gastrointestinal tract, joints and skin. Except for rare cases that progress to renal failure, it is a disease which heals without complications. The first case of male genital involvement in this syndrome was reported by Allen et al in 1960 (5). Since then, several reports of this condition with an incidence of scrotal involvement varying from 2 to 38% have been described. The disease is generally self-limited and responsive to steroid treatment, without infertility thereafter (6-11). In literature rare cases have been reported in which an acute scrotum was the initial presenting symptom in patients affected by HSP, while in previous reports the majority of boys had the diagnosis of HSP established before scrotal complaints developed. Our patient was

admitted to our hospital for fever, bronchopneumonia and bilateral cervical lymphoadenopathy and the initial diagnosis was focused on infectious disease. After three days from admission bilateral ecchymotic painful scrotal swelling without purpuric lesions was observed, while typical purpuric lesions of HSP were evidenced after five days from admission. Scrotal nuclear scanning was performed. It is extremely accurate in differentianting hyperemia from impaired blood flow encountered in the torsion of the spermatic cord (12). The demonstration of increased radionuclear activity of the affected scrotum evidences hyperemia. Since vasculitis of the scrotum and testicle is the basis for the swelling in HSP and the testicular blood supply is intact, the images are expected to show increased activity on the affected side. In our case scrotal nuclear scanning confirmed no testicular torsion and bilateral epididymoorchitis was considered. Our patient did not have renal complications and the lack of renal involvement (absence of micro/macrohematuria and proteinuria) was correlated with the age. In fact in children the renal complications are infrequent, while for adult patients are more frequent. Among previous cases of HSP that showed acute scrotal swelling as the only initial presentation, no one was found to have testicular torsion (13). The patient was treated with corticosteroids with a complete regression of purupura and scrotal involvement. This case report evidences that an acute scrotum may be the only initial manifestation of HPS and that this disease must be included in the differential diagnosis. The precise diagnosis is fundamental to prevent an unnecessary surgery.

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Accepted: December 21th 2009

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