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A CASE OF HENOCH-SCHÖNLEIN PURPURA WITH PENILE SKIN INVOLVEMENT

PENİS DERİSİ TUTULUMU İLE SEYREDEN HENOCH-SCHÖNLEIN PURPURALI BİR OLGU

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ABSTRACT

Henoch-Schönlein purpura (HSP); the most common leukocytoclastic vasculitis of childhood; may presents with a wide range of signs and symptoms. Testicular and scrotal involvement may be seen among genitourinary manifestations of HSP, whereas penile involvement is very rare. Here, we present a 5-year-old boy with-penile skin involvement of HSP.

Key Words: Henoch-Schoönlein purpura, penile skin involvement.

ÖZET

Henoch-Schönlein purpura (HSP) çocukluk çağının en sık görülen lökositoklastik vaskülitidir ve çok değişken semptom ve bulgularla ortaya çıkabilir. HSP genitoüriner bulguları arasında testiküler ve skrotal tutulum görülebilir, ancak penil tutulum çok nadirdir. Bu makalede HSP ve penis derisi tutulumu olan 5 yaşında bir olgu sunulmuştur.

Anahtar Sözcükler: Henoch-Schönlein purpura, penis derisi tutulumu.

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INTRODUCTION

Henoch-Schönlein purpura (HSP) is the most common vasculitis in children. It is characterized by a systemic leukocytoclastic angiitis of the small vessels most commonly affecting the skin, joints, gastro-intestinal tract and kidneys. A wide range of manifestations involving the genitourinary tract have been reported in children with HSP (1). Among nonrenal genitourinary manifestations, testicular and scrotal involvement have been reported, but penile involvement is very rare (2- 5). Here we present a five-year-old boy who had HSP with penile involvement with dramatic response to corticosteroid treatment.

CASE REPORT

A 5-year-old boy was admitted with pain and swelling on his wrists and right knee and rash on the lower extremities and in the palms. He suffered from an upper respiratory tract infection two weeks ago. He was a circumcised boy with normal anthropometric development. Extensive purpuric rash were present on the lower extremities and in the palms. The rest of the physical examination was normal. Laboratory investigations revealed normal urinalysis, complete blood count, hepatic and liver functions, C3 and C4 levels. Erythrocyte sedimentation rate (25 mm/h) and C-reactive protein (8 mg/L) were slightly elevated; antinuclear antibody, antineurophile cytoplasmic antibody, anti double-stranded DNA antibody, romatoid factor and fecal blood were negative. Nonthrombocytopenic palpable purpura particularly on lower extremities along with arthritis/arthralgia were compatible with the diagnosis of HSP (6). At the second day of the follow-up, proteinuria (2+) and hematuria (1+) were detected on the urinalysis. 24-hour urine protein excretion was in the non-nephrotic range (21 mg/m2/h). At the 4th day of follow-up palpable purpura developed on the scrotum and penile skin in addition to edema of the penis (figures 1 and 2). Doppler USG of the scrotal contents was normal other than the thickening of scrotal skin. Prednisolone treatment (1 mg/kg/day) was started for severe cutaneous and penile involvement. Rash and penile edema regressed by the 3rd day of this treatment along with the resolution of hematuria and non-nephrotic proteinuria. Thus, prednisolone treatment was tapered over one week and stopped. He was discharged from clinic and has remained well.

Figure 1: Scrotal rash

DISCUSSION

Henoch-Schönlein purpura is a leukocytoclastic vasculitis characterized by inflammation and necrosis of arterioles, capillaries and postcapillary venules (7). It is a small vessel vasculitis with multiorgan involvement including skin, gastrointestinal tract, joints, and kidneys with variable clinical expression. Classification criteria for HSP include the presence of palpable purpura and at least one of the

Figure 2: Penile edema

following features: diffuse abdominal pain; any biopsy showing predominant IgA deposits; arthritis or arthralgia; renal involvement (any hematuria and/or proteinuria) (6).

Extrarenal genitourinary manifestations of HSP include acute scrotum, ureteritis with associated hydronephrosis, hematoma of the bladder wall, urethritis, hemorrhagic spermatic cord, acute scrotum, priapism, thrombosis of the spermatic veins and epididymo-orchitis (2, 4). However, the involvement of the glans, prepuce and penile shaft has rarely been reported as either the earliest presenting symptom or occurring after the first skin eruption of the lower extremities (5,8,9). In one study, Mintzer et al reported genital involvement in 10 out of 155 children with HSP (11.6%). The affected children had scrotal swelling and tenderness. Only 3 of the 10 had penile involvement with swelling being the major symptom (10). In our case there was scrotal purpura and a significant penile involvement with swelling and rash. However, no scrotal swelling and testicular involvement was noted.

Treatment of penile involvement is controversial. Ferrara et al reported that penile involvement did not confer a worse prognosis and should be treated with conservative management only (4). On the other hand, Sandell et al treated their patient with pulsed methylprednisolone (600 mg/m2) followed by prednisolone 2 mg/kg/day, while Penesi et al treated with prednisone of 25 mg/day (9,11). Furthermore, penis is an end organ with a higher risk of permanent damage (5). Thus, we treated our patient with a short course of prednisolone and he responded well to this treatment.

In conclusion, penile involvement is an unusual complication of HSP and response to corticosteroid treatment is dramatic.

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