Ann Med Res 2020;27(4):1275-9

Annals of Medical Research

DOI: 10.5455/annalsmedres.2019.10.663

Case Report

Systemic steroid treatment of rarely seen genitourinary system manifestation in children with Henoch Schönlein purpura: Case report and literature analysis

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Abstract

Henoch-Schönlein Purpura (HSP) is the most commonly seen systemic leukocytoclastic vasculitis in children. Although it frequently presents with skin, joint, and gastrointestinal system involvement, genitourinary system involvement is rarely seen as well. We present a child with HSP-associated penile and scrotum involvement and a literature review was performed. An eight-years-old male patient who underwent five days systemic, five days oral steroid therapy in hospital and had a total recovery from the HSP-associated penile and scrotum involvement that is rarely reported in the literature. Scrotum and penile involvement are rare in patients with HSP; acceptable with a good prognosis. Disease-related genitourinary system involvement may be associated with findings such as penis and scrotum involvement, balanitis, urethritis, hematoma in the bladder wall, epididymo-orchitis and priapism. In the literature, most commonly scrotal edema or only penile involvement is mentioned. However, treatment for HSP is controversial, steroid and analgesic treatment can be applied in patients with HSP and penile/scrotum involvement.

Keywords: Balanitis; child; Henoch-Schönlein purpura; penile-scrotal involvement; treatment

INTRODUCTION

Henoch-Schönlein Purpura (HSP) (newly name: Ig A vasculitis) is a systemic small-vessel vasculitis and is well described in the paediatric literature. Its estimated incidence is about 13.5-20.4/100.000 and it occurs most frequently in two-year-old boys (1). Although the etiology of the disease is not known exactly, reasons such as infections (e.g. group A β–hemolytic streptococcus, Staphylococcus aureus, influenza, parainfluenza, Epstein-Barr virus, adenovirus, parvovirus, and mycoplasma) and vaccination (pandemic influenza A (H1N1) vaccine), are blamed for the emergence of the disease (2). There is often gastrointestinal system, kidney and joint involvement accompanying skin lesion (non-thrombocytopenic palpable purpura). However, other organ and system manifestations may occur depending on the location of the involved small vessels. Rarely genital, respiratory and central nervous system and pancreas involvement can be seen. Henoch-Schönlein Purpura associated genital system involvement is a rare condition and has been reported in 2-38 % of cases. Patients may present with complaints such as swelling in the penis, redness and scrotal pain that may be confused with acute scrotum (3).

In this paper, an eight-year-old male patient having systemic steroid therapy due to HSP-associated penile and scrotum involvement in addition to gastrointestinal system, skin and joint involvement, which is rarely reported in the literature, has been presented.

CASE REPORT

A previously healthy eight-year-old male patient presented to the Pediatric Emergency Department (PED) with complaints of swelling and pain of wrists and ankles, inability to walk, rash and abdominal pain. His medical history was reported that he had an upper respiratory tract infection and took oral penicillin-V suspension totally four doses two weeks ago. All complaints began on the day of application. There was no accompanying trauma, fever, diarrhea, nausea or vomiting. He was admitted to the PED with normal vital signs. In physical examination he had

Received: 23.10.2019 **Accepted:** 28.02.2020 **Available online:** 02.04.2020
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tonsillar hyperemia, hypertrophy. He had bilateral palpable, purple-colored rash, swelling, tenderness, limited mobility on the wrist and ankles. Other physical examinations were normal. No abnormal pathology was found on hemogram, biochemical and urinalysis parameters. Fecal occult blood (FOB) test was positive. Throat swab culture was normal. The patient was treated with intravenous fluids as conservative treatment in the PED Observation Unit. After admission, he complained of abdominal pain in the left lower quadrant. Because of the pain and the positivity of FOB test, abdominal ultrasound (US) was performed and reported as normal. Abdominal pain regressed in the third hour of the patient's follow-up. The patient whose clinical condition improved in follow-up and the patient did not have any abnormality in laboratory results, discharged by prescribing anti-inflammatory and proton pump inhibitors due to joint and skin involvement and antihistamine due to itching.

Over the next 24 h, he was re-admitted to the PED with vomiting and penile pain, swelling and redness. There was no history of penile trauma or dysfunction with micturition. Vomiting was twice with small amount and was not bile or bloody. Vital signs normal. On physical examination, it was detected that the rashes were more concentrated than the first examination in both lower extremities and they had spread to thigh and upper extremities. There was no swelling or pain on the joints. There was mild tenderness in the lower quadrants in the abdominal examination; there was no defense and rebound. In examination of the genitourinary system; bilateral testes were in the scrotum, the skin on both hemiscrotum was erythematous and sensitive to palpation, the penis was hyperemic and edematous (Figure 1).

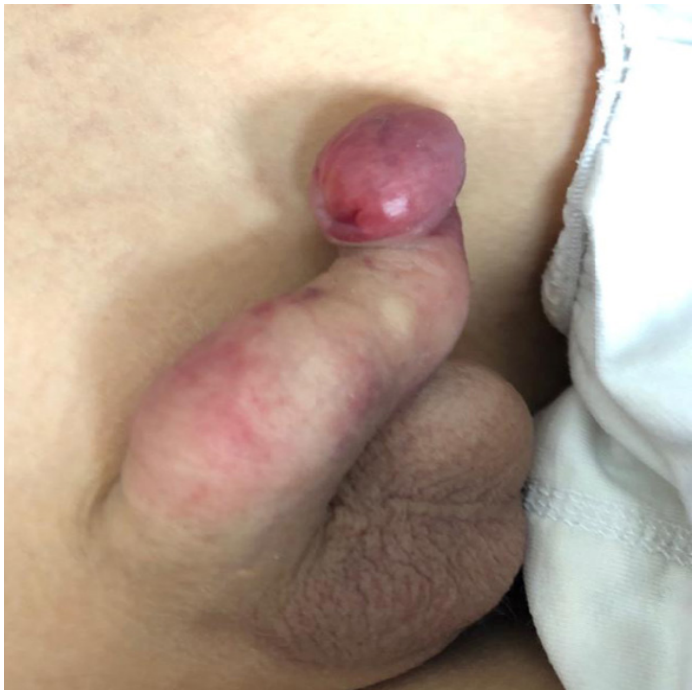

Figure1. Purpuric, erythematous and edematous penile involvement of glans penis and penile shaft.

In laboratory WBC count of 18.980/mL, hemoglobin level 13.8 g/dL, hematocrit level 43.4% and platelet count of 358.000/uL were detected. No abnormal pathology was found on biochemical and urinalysis parameters. CRP level was 18.5 mg/L and FOB was positive again. The abdominal and scrotal Doppler US reported as "increased thickness and heterogeneity were observed in the right epididymis head and trunk section and vascularization was increased (epididymitis). Some free fluid was detected in the right scrotal sac. There was no sign of intussusception". With these findings, in addition to the gastrointestinal system, skin and joint involvement, HSP-associated penile and scrotum involvement was considered and he was consulted to the Department of Pediatric Rheumatology and Pediatric Surgery. The patient was admitted to the Pediatric Health Care Unit to be treated with pulse steroid (30 mg/kg/day single dose methylprednisolone) for five days. Penile symptoms regressed on second day of treatment (Figure 2). Because of persistent scrotal erythema and edema, he was re-evaluated by the Department of Pediatric Surgery. Scrotal Doppler US was repeated and it was reported as "significant increase in left epididymis blood supply and minimal fluid within the left scrotal sac".

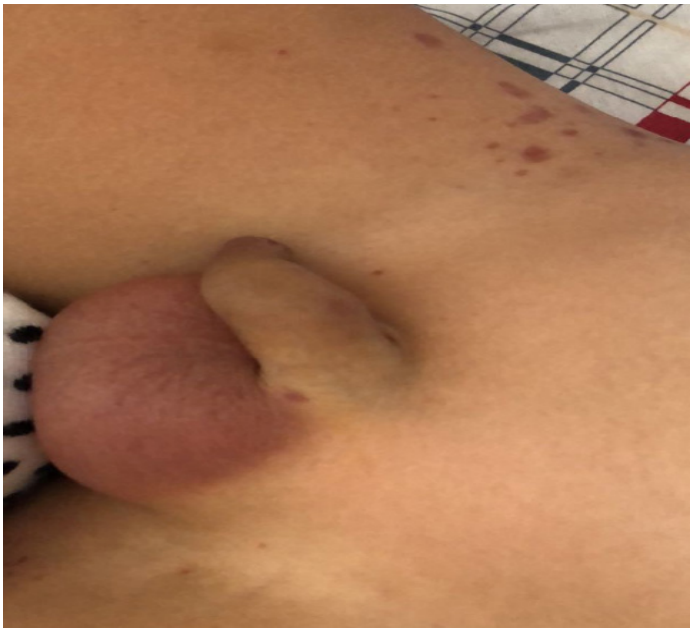

Figure 2. Second day of pulse methylprednisolone treatment; redness and edema of the penis reduced, yet scrotal edema and erythema persist

Pulse steroid therapy was terminated on patient whose signs of scrotal involvement regressed on the fifth day of hospitalization (Figure 3). It was switched to oral methylprednisolone (2 mg/kg/day, 2 doses) treatment due to continued edema and pain in the joints. Signs of joint and scrotal involvement were regressed on the 10th day and the patient was discharged.

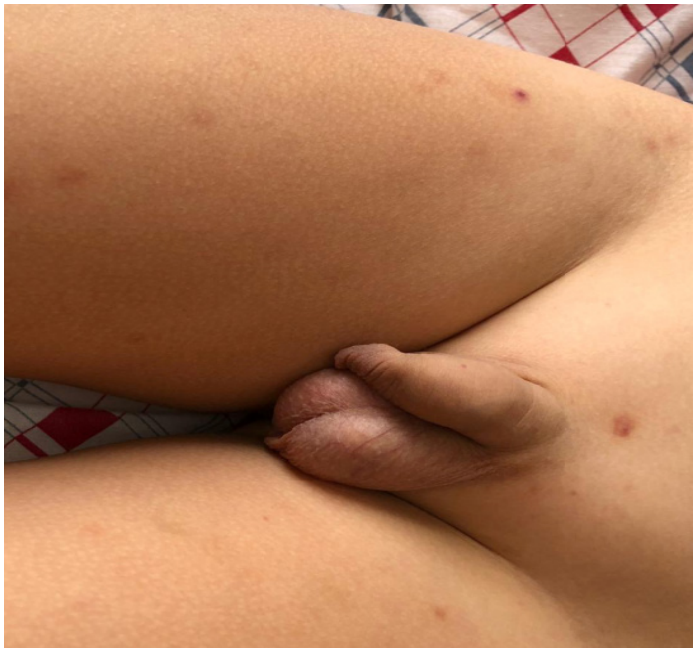

Figure 3. After pulse steroid, first day of oral methylprednisolone treatment, penis and skin findings

RESULTS and DISCUSSION

Henoch-schönlein purpura associated genitourinary system involvement is one of the rare cases and treatment is still controversial. Disease-related genitourinary system involvement may be associated with findings such as penile and scrotum involvement, balanitis, urethritis, hematoma in the bladder wall, epididymo-orchitis and priapism (4). In the literature, scrotal edema or only penile involvement is mentioned most commonly. The incidence of scrotal involvement in children with HSP has been reported 2-38% . Involvement may be seen at the time of initial admission, as it may also occur during follow-up after skin involvement (3).

Our case had developed skin and joint findings, followed by penile and scrotum involvement, in addition to gastrointestinal as well as skin involvement. According to our knowledge, there are fourteen cases with HSP-associated penile and/or scrotal involvement in the literature and different follow-up and treatment protocols have been applied for each case (Table 1) (3-14).

Table 1. Characteristic of HSP patients with genitourinary system manifestation (3-14)

Ref	Age (Yr)	Penile retention time after rash	Renal involvement	Arthritis/ Arthralgia	Scrotal involvement/ Orchitis	GIS involvement	Clinically Important Presentations During Disease Course	Treatment Regimen	Recovery Time of Penile Lesions	FollowUp Visits/ Complications
3	4,5	30 days	No	Yes	None	No		P	5 days	1 year / No
4	9	1 day	Proteinuria Hematuria	Yes	None	No	Priapism 1 month later	No	1 day after priapism	6 months/ Priapism
5	19	60 days	No	No	None	No		No	30 days	6 weeks/ Recurrence
6	5	3 days	No	No	None	No		P	2 days	Unfollowed
7	3	8 days	No	No	None	No	Ipsilateral otomastoiditis advanced on the 6th day of rash	MPZ and P	2 days	6 months/ No
8	5	14 days	No	No	S	No		No	unknown	Unfollowed
9	4	5 days	No	No	None	No		MPZ and P	12 hours	3 months / No
10	2	14 days	No	No	None	No	Balanopostitis	No	4 days	Unfollowed
11	4	Same day	Sterile Pyuria	No	None	No		P	1 day	6 months / No
12	3,5	7 days	No	Yes	S , O	No		P	4 days	4 months / No
13	5	4 days	No	Yes	S	No		P	3 days	Unfollowed
14	7	Same day	No	No	O	No		P	2 days	3 weeks/ No
Our Case	9	1 day	No	Yes	S	Yes	Epididymitis	MPZ and P	10 days	6 months/ No

MPZ: methylprednisolone, P: Prednisolone S: Scrotal involvement, O: Orchitis

In the literature it was observed that the scrotal involvement in a case other than ours also started one day after the rash (4), and two cases received methylprednisolone treatment in which there were no signs of systemic involvement as well (7,9). In our case, genitourinary system involvement occurred after other HSP-associated organ system manifestations and, in contrast to other cases in literature, having a positive FOB test he took a long-term steroid treatment.

Paydary at al. (15) followed up 168 cases with HSP ; nine cases of these were with penile involvement and six of with penil involvement were accompanied by abdominal pain and their abdominal US were normal. Additionally, it was reported that seven cases with penile manifestation were treated with prednisolone, and in only one of the cases crescentic glomerulonephritis developed as a complication in the second month after discharge.

In patients presenting with scrotal pain, edema and redness, acute scrotum condition causing testicular torsion, epididymitis, orchitis, balanitis and balanoposthitis should be considered in the differential diagnosis. Trauma and sexual abuse should be questioned (11). It should also be kept in mind that vasculitis, such as HSP, may lead to genital tract involvement as well. HSP-associated scrotal involvement may occur in 3% of patients presenting with acute scrotum. As mentioned in the literature, patients may present with involvement of the penis and/or scrotum without having skin manifestations (3).

In literature, accompanying testicular torsion has also been reported for patients with HSP (16). In our case, typical skin findings before genital system involvement facilitated the diagnosis of the patient. In the differential diagnosis of acute scrotal cases scrotal US is necessary.

Epididymis expansion and thickening of the scrotal skin are typical for US findings of HSP (12). In our case, "increase in thickness in the right epididymal head-trunk section" was determined and it was interpreted in favor of epididymis. HSP treatment, together with close follow-up of the patient, is a supportive treatment in terms of complications.

The use of steroids for treatment is controversial. In some cases, high-dose pulse steroid (9) and in some cases, oral/iv prednisolone was used (3,6,11-14). Complete recovery was observed in cases followed without treatment as well (4,5,8,10). As a conclusion, it has been reported that in steroid treatment the findings recover faster. It is recommended that steroid treatment should be used in cases where the signs of penis and scrotum are severe or systemic complications are seen with genitourinary system involvement (3,9). There is no study in the literature comparing the healing time of HSP patients who used steroids and the ones who did not, for genital involvement. However, when all cases were evaluated in general, it was seen that patients taking steroids recovered faster than those who did not (Table 1). Our patient received pulse methylprednisolone because of the emergence of penile and scrotum findings shortly after skin and joint involvement. He received oral prednisolone after pulse methylprednisolone for five days due to skin and penile involvement in addition to scrotal involvement and the persistence of the arthritis/ arthralgia findings for a long period of time. On the fifth day of the treatment, penile involvement was recovered which was similar to those in the literature.

CONCLUSION

As a conclusion, scrotum and penile involvement are rare in patients with HSP; acceptable with a good prognosis. The place of steroid therapy in these cases is controversial. In case of HSP cases with penile and/or scrotum involvement in addition to systemic involvement, steroid therapy should be considered for administration intravenously or orally.

Competing interests: The authors declare that they have no competing interest.
Financial Disclosure: There are no financial supports.

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