CASE REPORT

Self-limiting recurrent bullous Henoch-Schonlein purpura with lupus anticoagulant

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SUMMARY

Henoch-Schonlein purpura (HSP) is the most common acute systemic vasculitis of childhood with an incidence of approximately 1:10 000. It commonly presents with skin, gastrointestinal tract, joints and renal system signs and symptoms. We present a case of recurrent self-limiting HSP with lupus anticoagulant presenting with haemorrhagic bullous skin lesions and scrotal swelling. The initial presentation resolved within 4 weeks but over the next 6 months, a few self-limiting but milder skin lesions occurred. Some institutions have advocated the use of steroids in these cases but our case showed that the use of steroids may not be required. This atypical presentation of HSP led to unnecessary investigations that this case report aims to prevent in the future by raising its awareness.

BACKGROUND

We present a case of a self-limiting recurrent Henoch-Schonlein purpura (HSP) with lupus anticoagulant presenting with haemorrhagic bullous skin lesions and scrotal swelling. This is a rare presentation of HSP and commonly leads to initial diagnostic uncertainties and unnecessary investigations and treatments. This report aims to increase awareness of this atypical form of HSP to avoid unnecessary investigations and treatments. The role of steroids to treat this form of HSP remains controversial. It is hoped that this report and other similar reports will encourage formal research on the role of steroids in bullous HSP.

CASE PRESENTATION

A previously healthy 3-year-old boy presented to a tertiary paediatric centre with a painful, swollen right scrotum, a 48 h history of a blistering rash on his right leg and general irritability. He had no significant medical history. He was on no medications and had no known allergies or a history of atopic conditions. He had been growing and developing normally with no history of weight loss. He was up to date with his vaccinations and had no recent trauma, travel or insect bites. His family was well with no history of significant illnesses such as connective tissue disorders or atopic illnesses.

On admission he was afebrile, normotensive and urinalysis was negative for protein and blood. In view of the unilateral scrotal pain and swelling, he underwent a surgical exploration. During surgery, his epididymis appeared inflamed, his hydatid was erythematous and testis was non-torted. These were confirmed by histopathology. By 2 days postoperation the rash had spread to both lower limbs and

the extensor surface of his left elbow. Infection was high on our differential diagnosis and this prompted a prescription for oral co-amoxiclav before discharge.

At review 5 days postoperation the rash had spread to his buttocks and both elbows and was accompanied by ankle swelling and an intermittent limp. The characteristics of the rash had also changed. In addition to several discrete bullae, it also consisted of several red non-blanching maculopapular lesions. Some of these were clearly purpuric (figure 1) and one possibly infected, indurated lesion on his right ankle. No signs of abdominal or renal involvement were present. The nature and distribution of the non-blanching component of the rash suggested a diagnosis of HSP. However, diagnostic uncertainty remained because of the bullous component of the rash. At this point, he was referred to a dermatologist for a second opinion. They were also uncertain of a diagnosis but they prescribed steroid and emollient creams pending the results of a skin biopsy. The purpuric lesions prompted clotting studies which demonstrated a prolonged activated partial thromboplastin time (APTT). This was subsequently attributed to the presence of lupus anticoagulants in the blood. The skin biopsy suggested that the most likely diagnosis was a leukocytoclastic vasculitis in keeping with a clinical diagnosis of HSP. By this stage, the rash had begun to resolve and therefore no treatment was administered. The entire episode had fully resolved by 4 weeks and subsequent follow-ups reported a few further self-resolving skin flare-ups. Our patient was back to

Figure 1 Discrete bullae on right thigh with small purpuric satellite lesions. Similar lesions also present on extensor surfaces of lower legs, buttocks and forearms. Lesions resolved without specific treatment within 4 weeks.

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normal after 4 weeks and continued to thrive and gain weight appropriately.

INVESTIGATIONS

- ▶ Urine culture: negative
- ▶ Urinalysis negative for blood and protein
- ▶ Blood culture: negative
- ▶ C3, C4 levels: normal
- ▶ Anticardiolipin and anti-β₂-glycoprotein antibodies: negative
- ▶ Hepatitis B serology: normal
- ▶ Ultrasound abdomen: normal
- ▶ Normal blood pressure
- ▶ Abnormal clotting on presentation (prothrombin time (PT) 15.2, APTT 62.8, thrombin time 19.0)
- ▶ Normal plasma stat mix does not correct (<50%), prolonged dilute Russells viper venom time.
- ▶ Serial follow-up. Last repeat 2 months after presentation (PT 13.5, APTT 29.4)
- ▶ Haemoglobin 11.1, white cell count 4.96, platelets 297
- ▶ Skin biopsy: Punch biopsy shows what appears to be ruptured intraepidermal blister and pandermal infiltrate of neutrophils and eosinophils, including abundant karyorrhectic debris. In some areas the cells were aggregating around blood vessels. Despite the absence of true vasculitis in the multiple sections examines, the most likely diagnosis is leukocytoclastic vasculitis in keeping with a clinical diagnosis of HSP

DIFFERENTIAL DIAGNOSIS

- ▶ Skin infection (initially)
- ▶ Causes of secondary bullous eruptions: drug reactions, neoplasia, inflammatory connective tissue diseases

TREATMENT

Supportive care only. This usually involves pain control, use of anti-inflammatory medics such as ibuprofen, oral steroids in some cases.

OUTCOME AND FOLLOW-UP

The initial presentation resolved within 4 weeks; however, he subsequently had a few further self-limiting flare-ups with similar rashes. He was completely well with no renal involvement when reviewed 6 months from initial presentation.

DISCUSSION

HSP is the most common acute vasculitis in children, with an incidence of approximately 1:10 000.^{1 2} Classical clinical features include a rash (100%), arthralgia (82%), abdominal pain (63%), nephritis (40%) and gastrointestinal bleeding (33%).³

The cutaneous features most commonly consist of erythematous papules, petechiae and purpura mainly affecting the lower extremities but can also be present on arms, face and neck. Pressure has been proposed as a factor in the pathogenesis as the severity of lesions are often greater at pressure points such as around the malleoli or on the dorsal aspect of the foot beneath tight shoelaces.⁴ Rarely, the cutaneous lesions can present as haemorrhagic bullae as was seen in this case. The first report of such bullous HSP was made in 1985 of a 5-year-old boy who developed blistering lesions on the extensor surfaces and perioral region.⁵

The diagnosis of bullous HSP can be made on clinical features only. The bullous lesions can cause diagnostic problems but as was shown in our case, the classical features of HSP such as classical rash and distribution, joint pains, renal involvement and

abdominal pains may become apparent later on. The presence of these features and the absence of other features suggesting alternative causes can be used to make a diagnosis. In retrospect, this could have been the scenario with our patient. A skin biopsy may be required to aid diagnosis but increased awareness of this rare form of HSP may avoid the need for skin biopsies.

Owing to the rare presentation of bullous HSP, there appears to be a lack of consensus on the preferred management. It has been theorised that anti-inflammatory effects of steroids could be beneficial. Indeed, in a case report published last year, Kocaoglu *et al* initiated prednisolone 2 mg/kg/day in a child with severe systemic symptoms and haemorrhagic bullous lesions in light of favourable reports previously published by Liu *et al*.⁶ The bullous lesions subsequently began to resolve within the week and were fully recovered by 3 weeks.⁴ By contrast, Trapani *et al*⁷ report a lack of success of steroid treatment and need for additional immunotherapies in their patients with bullous HSP. We considered the use of prednisolone to treat our patient, however by the time this decision was reached, our patient was clearly much improved and almost back to normal. Hence, it was never used. I believe that the fact that our patient got better without significant treatment and remained afebrile supports the diagnosis of bullous HSP and ruled out other rarer causes of bullous eruptions such as malignancy, connective tissue disorders and significant bacterial infection such as scalded skin syndrome.

This case report aims to raise awareness of bullous cutaneous lesions as a variant of HSP which like most cases of HSP can resolve without the need for steroids with no apparent long-term effects or prognosis. Episodes may reoccur and resolve without treatment within 12 months of initial presentation. A high level of suspicion could prevent a delay in diagnosis, avoid unnecessary investigations and prompt appropriate management. Furthermore, the presence of lupus anticoagulants is worth noting as a possible aid in diagnosis.

Learning points

- ▶ Haemorrhagic bullous Henoch-Schonlein purpura (HSP) is a rare variant of HSP which is self-limiting and can be recurrent.
- ▶ It is important to be aware of this rare variant of HSP in order to avoid unnecessary investigations and treatment.
- ▶ The role of steroids in the management of bullous HSP is not clear and is a topic that needs further clinical research.
- ▶ Cases can be associated with lupus anticoagulants.

Contributors This case report is based on a patient from EO's clinical workload as a consultant paediatrician. He collected all relevant information for the case report. BG wrote up the case report.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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