# **Case Report**

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# Henoch-Schonlein purpura masquerading as vesiculobullous lesions in a child

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#### **ABSTRACT**

Henoch-Schönlein purpura (HSP) is a systemic small vessel vasculitis occurring commonly in children presenting with palpable non-thrombocytopenic purpura, arthralgia and abdominal pain. Haemorrhagic bullous and ulcerative lesions are rare in HSP in children and can be a diagnostic challenge. We present a case of 6-year-old boy child who presented with bullous purpuric lesions in lower limbs, arthralgia and increased serum IgA. Histopathology of skin lesion revealed leukocytoclastic vasculitis. However, the direct immunofluorescence was negative for IgA deposits.

Keywords: HSP, Haemorrhagic bullae, Direct immunofluorescence, Leukocytoclastic vasculitis, IgA

## **INTRODUCTION**

Henoch-Schönlein purpura (HSP) is the most common childhood vasculitis with over 90% of patients under 10 years of age and a mean age of 6 years. The incidence of HSP ranges from 6.7-22 per 100,000 children. HSP typically presents with palpable purpura and petechiae. While erythematous maculopapular and urticarial lesions have also been reported, bullous HSP is a rarer presentation of HSP, exhibited by only 2% of affected children. The presence of atypical symptoms such as bullae and painful lesions can complicate the diagnosis of HSP.

We report this case as haemorrhagic bullous lesions are very rare in childhood HSP and hence can be a challenge in clinical diagnosis.

# **CASE REPORT**

A 6-year-old boy presented with palpable purpuric rash on extremities, back of trunk and buttocks for 9 days. The

rash first appeared in lower limb, spreading upwards. Some lesions were vesiculo-bullous (Figure 1 and 2).



Figure 1: Palpable purpuric rashes, vesico- bullous lesions in the patient.



Figure 2: Hemorrhagic bullous lesions in the patient.

He had swelling and pain of small joints of both hands and wrists for two days. He had fever, cough and sore throat with poor oral intake 4 days before the onset of rashes and was treated with oral erythromycin for the same elsewhere. On arrival, his vitals were stable. Examination revealed numerous petechiae, palpable purpura, bullae with surrounding erythematous to violaceous induration, and arthritis of small joints of both hands and wrists. The palms, soles, face were spared.

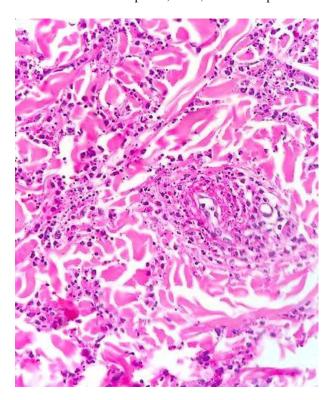


Figure 3: Histopathology of the skin lesions demonstrating leukocytoclastic vasculitis.

Blood investigations revealed neutrophilic leucocytosis with normal platelet counts. Renal function tests and urinalysis were normal. CRP was elevated (20.8). ESR was normal. C3, C4 were low. ANA, ASO titre and RA

factor were negative. Suspecting vasculitis, rheumatologist and dermatologist opinion was obtained. A clinical diagnosis of HSP was made. In view of atypical bullous presentation and extensive involvement, a skin punch biopsy was done (11 days after onset of rashes) and child was started on prednisolone. Serum IgA was elevated. Histopathology of skin showed typical features of leukocytoclastic vasculitis (Figure 3 and 4).

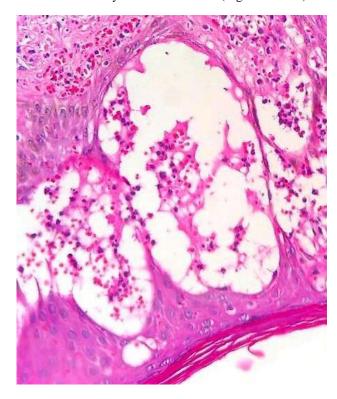


Figure 4: Histopathology showing spongiotic bullae.



Figure 5: Resolution of lesions with no new purpuric or bullous lesions but with mild residual pigmentation.

However, direct immunofluorescence did not show IgA deposits. The child was discharged post biopsy on

prednisolone and was followed up in OPD after one week. There was resolution of lesions with no new purpuric or bullous lesions but with mild residual pigmentation (Figure 5). Joint pain and swelling also subsided.

#### **DISCUSSION**

HSP is an IgA mediated autoimmune hypersensitivity vasculitis characterised by the classical triad of non-thrombocytopenic palpable purpura, abdominal pain and arthritis. It is named after two German physicians Henoch and his teacher Schonlein. Schonlein found the correlation between purpura, arthritis and urinary sediments in 1837, while Henoch discovered the association of purpura with abdominal pain, bloody diarrhoea and renal involvement in 1874.

HSP has been associated with a history of preceding infections, especially upper respiratory tract infection.<sup>6</sup> Most common symptoms include palpable purpura, joint pain, gastrointestinal complaints, and renal involvement. Lesions typically occur on pressure prone surfaces like ankles, feet and buttocks. Bullous lesions in paediatric HSP has been exhibited by only 2 percent of affected children.<sup>3</sup> Conversely, haemorrhagic bullous rash is present in 16-60 percent of adult cases of HSP.<sup>7,8</sup> In a case report followed by literature research by Hung-Wen et al in 2016 at Taiwan concluded that so far only 39 paediatric patients have been reported with haemorrhagic bullae in HSP.9 Nothhaft et al in Germany conducted an extensive literature research in 2018 and observed that only 41 paediatric patients with bullous HSP were reported so far excluding the patient that he reported in his article.10

Other causes of bullae in children include erythema multiforme, toxic epidermal necrolysis, epidermolysis bullosa, bullous mastocytosis, pemphigus, bullous pemphigoid, dermatitis herpetiformis, bullous impetigo.

Kobayshi et al reported that matrix metallopeptidase-9 (MMP-9, gelatinase) is secreted by polymorphonuclear neutrophils, which can cause formation of blisters by degrading type VII collagen in basement membranes thus contributing to bulla formation.<sup>11</sup>

In 2010, the EULAR/PRINTO/PRES criteria for HSP was formally published. The criteria include palpable purpura as a mandatory criterion, together with at least one of the following findings: diffuse abdominal pain, Leukocytoclastic vasculitis (LCV) with predominant IgA deposits on skin biopsy or proliferative glomerulonephritis with predominant IgA deposits, acute arthritis or arthralgias in any joint, and renal involvement as evidenced by proteinuria and/or haematuria. The sensitivity and specificity of these criteria were 100 and 87% respectively. 12

LCV is a small vessel vasculitis characterized histopathologically by immune complex-mediated vasculitis of the dermal capillaries and venules, classically demonstrated by immune complex deposition, neutrophil infiltration, leucocytoclasis with nuclear dust (karyorrhexis) in the vessel walls. <sup>13</sup> The primary immunoglobulin deposited in HSP in both the skin and the kidney is IgA. <sup>3,14</sup> Elevated IgA levels support diagnosis of Henoch-Schönlein purpura over other forms of LCV, but is nonspecific. <sup>15</sup>

IgA deposition is demonstrated in the epidermis or dermo-epidermal junction using immunofluorescence staining. IgA deposition is found in 75% to close to 100% of clinically confirmed cases of HSP. However, it has been demonstrated that in some cases of HSP vascular IgA deposition in the skin appears to be negative. If In a study conducted by Nandeesh et al it was observed that a biopsy specimen taken too late (i.e., after 48 hrs) may show more of the pathology of repair than of the initial injury and may have negative DIF because immune deposits are degraded rapidly. It was concluded in this study that DIF is negative in a significant number of cases, especially when samples are taken more than seven days after onset of lesions.

Biopsy should be obtained from the border of a fresh non-bullous and non-necrotized lesion where proteolytic degradation of IgA is less advanced. <sup>10</sup> Taking multiple punch biopsies including the lesional and normal skin might increase the chances of obtaining positive IgA deposits.

Corticosteroids are beneficial in HSP patients with severe bullous and ulcerative lesions due to their potent anti-inflammatory effect by inhibition of AP-1 binding activity in the nucleus, reduction of nuclear factor-kappa B (NF-kB) and decreasing plasma levels of MMP-2 and MMP-9.<sup>18</sup>

## **CONCLUSION**

Conclusion of the study is hemorrhagic vesicles and bullae are rare in children with HSP, they can pose a challenge for clinical diagnosis. Early biopsy within 48 hours of appearance of lesions is essential to demonstrate Ig A deposits in immunofluorescence. Bullae in HSP have no prognostic value. Systemic steroids reduce the severity of bullae in HSP.

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