

CASE REPORT

Bullous Henoch–Schonlein purpura with involvement of face

Devdeep Mukherjee, Ishita Majumdar, Priyankar Pal¹, Sandipan Dhar², Ritabrata Kundu

Departments of Pediatric Medicine, ¹Pediatric Rheumatology and ²Pediatric Dermatology, Institute of Child Health, Kolkata, West Bengal, India

ABSTRACT

Henoch–Schonlein purpura (HSP) with facial involvement with bullous rashes are extremely rare. A 12-year-old boy presented with abdominal pain and features of arthritis. He also had multiple purpuric rashes over his lower limbs. Gradually, he developed bullous rashes which were seen on his legs and hands and progressed to involve the face. He was confirmed to be suffering from HSP from clinical presentation and skin biopsy. The child responded well to oral steroids. Bullous lesions may be seen in HSP. However, there is neither prognostic significance of this nor does it alter the management. Other causes of bullous lesions should be ruled out. As facial involvement is associated with renal and gastrointestinal involvement, these children should be monitored for sequelae.

Key words: Bullous Henoch–Schonlein purpura, facial involvement, Henoch–Schonlein purpura

INTRODUCTION

Henoch Schonlein purpura is rarely noted to have skin lesions of the face. Our patient presented with abdominal pain, arthritis and bullous, non hemorrhagic lesions of the face

CASE REPORT

A 12-year-old boy was admitted with a history of pain, swelling of ankles, and difficulty in walking for the last 5 days. He was also having abdominal pain since 3 days and had developed a purpuric rash over his legs and trunk since the last 2 days.

On admission, he was afebrile with normal sensorium. Vitals were stable and blood pressure 112/78 (<90th centile for age and sex) of Hg. Abdomen was soft, without any tenderness, or organomegaly. Both his ankle and right knee joint was swollen, warm, and tender. He had multiple purpuric eruptions over his lower limbs and on his hands. He also had

a few bullous lesions on his legs [Figure 1]. Blood counts were normal with platelet – 3.2 lakhs/cumm. Sepsis markers were negative. He had normal liver and renal function and coagulation profile. Routine urine examination did not reveal any hematuria or proteinuria. Antinuclear antibody was negative and C3 and C4 were normal. Skin biopsy was suggestive of neutrophilic infiltrate in the epidermis. Fibrinous necrosis of wall of small dermal vessels associated with karyorrhexis and red cell extravasation was noted. Perivascular and interstitial neutrophilic dermal infiltrate admixed with nuclear fragments, and mononuclear cells were present. Sections also showed subepidermal blisters containing inflammatory cells and RBCs and involvement of subcutaneum in the form of lobar panniculitis. Immunofluorescence showed IgA and IgM deposition in the wall of small

ADDRESS FOR CORRESPONDENCE

Dr. Devdeep Mukherjee,

Flat No 6F, Uttara Cooperative Housing Society, 13, Broad Street,
Kolkata - 700 019, West Bengal, India.
E-mail: devdeep_dm@rediffmail.com

Access this article online

Quick Response Code



Website:
www.ijpd.in

DOI:
10.4103/2319-7250.188462

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Mukherjee D, Majumdar I, Pal P, Dhar S, Kundu R. Bullous Henoch-Schonlein purpura with involvement of face. Indian J Paediatr Dermatol; doi: 10.4103/2319-7250.188462.

dermal vessels. Staining of C3 in vessel wall was patchy and faint with the overall morphological features being suggestive of leukocytoclastic vasculitis, i.e., Henoch–Schonlein purpura (HSP).

Over the next 48 h of admission, his abdominal pain persisted. He developed purpuric rashes over his face and postauricular region. Bullous eruptions without any hemorrhage were noted on lower legs, hands, trunk, and even on face [Figure 1]. He was started on oral prednisolone at 60 mg/day. Over the course of next 5 days, his joint swelling and the abdominal pain subsided. Rashes also stopped progressing, and he was discharged on tapering doses of prednisolone for the next 6 weeks. During this period, his blood pressure increased to 132/84 mm of Hg (>95th centile), for which he was given amlodipine. Repeat urine examination was normal. He was gradually tapered off antihypertensive. At 1 year follow-up, he is clinically well with no evidence of renal involvement.

DISCUSSION

HSP – a leukocytoclastic vasculitis is characterized by the deposition of IgA in the wall of the small vessels, with the predominant involvement of the skin, renal, gastrointestinal system, and joints.^[1] The diagnosis of HSP is confirmed on the basis of palpable purpura (without any evidence of coagulopathy and thrombocytopenia) and along with 1 of the 4 following features: abdominal pain, features of arthritis or arthralgia, biopsy confirming IgA deposition, and kidney involvement (proteinuria, hematuria or red cell casts) as per EULAR criteria.^[2] Our patient had purpura, arthritis and positive biopsy findings suggestive of HSP.



Figure 1: Purpuric and bullous lesion affecting the legs, hands, and face in a child with Henoch–Schonlein purpura

Dermatological involvement is characterized by palpable purpura over the legs and buttocks. Bullous lesion is extremely rare in children. Only 29 children have been reported to have bullous lesion in literature.^[1] Two percent children have been reported to have bullous lesion in a study by Abdel-Al *et al.* compared to 60% in adults.^[3,4] Bullae are characteristically seen in the legs and buttocks (dependent areas). However, our patient had bullae on his leg, hands, trunk, and a few on the face as well. Lesions affecting the face are extremely uncommon in children. Only two children have been previously reported to have bullous lesion of the face.^[5,6] Bullous lesion is commonly seen in staphylococcal scalded skin syndrome, toxic epidermal necrolysis, bullous impetigo, and erythema multiforme in children, leading to a diagnostic dilemma. The presence of abdominal pain with articular involvement along with biopsy findings confirmed this to be HSP.

Bullous lesion and IgM deposition have been associated with renal involvement in an adult study. Our index case had abdominal pain without any renal involvement.^[7]

The child responded well to oral prednisolone. Joint inflammation and skin lesions gradually subsided. Steroids may have a role in extensive bullous HSP to reduce the severity of lesions and chance of ulcers and necrosis.^[8-10] We also gave steroids because of the persistent abdominal pain and arthritis.

Skin lesions are not known to influence the outcome in bullous HSP.^[1] Our patient responded well to treatment without any sequelae. We would like to highlight the fact that although rare, bullous lesions are seen in HSP, and a renal workup with skin biopsy should be considered to establish the diagnosis.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial Support and Sponsorship

Nil.

Conflicts of Interest

There are no conflicts of interest.

REFERENCES

1. Trapani S, Mariotti P, Resti M, Nappini L, de Martino M, Falcini F. Severe hemorrhagic bullous lesions in Henoch Schonlein purpura: Three pediatric cases and review of the literature. *Rheumatol Int* 2010;30:1355-9.
2. Ozen S, Ruperto N, Dillon MJ, Bagga A, Barron K, Davin JC, *et al.* EULAR/PReS endorsed consensus criteria for the classification of childhood vasculitides. *Ann Rheum Dis* 2006;65:936-41.
3. Abdel-Al YK, Hejazi Z, Majeed HA. Henoch Schönlein purpura in Arab children. Analysis of 52 cases. *Trop Geogr Med* 1990;42:52-7.
4. Tancrede-Bohin E, Ochonisky S, Vignon-Pennamen MD, Flageul B, Morel P, Rybojad M. Schönlein-Henoch purpura in adult patients. Predictive factors for IgA glomerulonephritis in a retrospective study of 57 cases. *Arch Dermatol* 1997;133:438-42.
5. Ishii Y, Takizawa T, Arakawa H, Saga R, Mochizuki H, Tokuyama K, *et al.* Hemorrhagic bullous lesions in Henoch-Schönlein purpura. *Pediatr Int* 2005;47:694-7.
6. Júnior CR, Yamaguti R, Ribeiro AM, Melo BA, Campos LA, Silva CA. Hemorrhagic vesicle-bullous lesions in Henoch-Schönlein purpura and review of literature. *Acta Reumatol Port* 2008;33:452-6.
7. Belli AA, Dervis E. The correlation between cutaneous IgM deposition and renal involvement in adult patients with Henoch-Schönlein purpura. *Eur J Dermatol* 2014;24:81-4.
8. den Boer SL, Pasmans SG, Wulffraat NM, Ramakers-Van Woerden NL, Bousema MT. Bullous lesions in Henoch Schönlein Purpura as indication to start systemic prednisone. *Acta Paediatr* 2010;99:781-3.
9. Rosato L, Chehade H, Cachat F. Re: Steroids in haemorrhagic bullous Henoch-Schoenlein purpura. *Acta Paediatr* 2011;100:319-20.
10. Park SJ, Kim JH, Ha TS, Shin JI. The role of corticosteroid in hemorrhagic bullous Henoch Schönlein purpura. *Acta Paediatr* 2011;100:e3-4.

