Bullous Henoch-Schönlein Purpura in 9-year old Filipino Male: A Case Report*

Camille Clarisse S. Mundin, MD[1]
Christene Pearl F. Arandia, MD, FPDS[2]
Mae Ramirez-Quizon, MD, FPDS[2]

ABSTRACT

Henoch- Schonlein purpura (IgA vasculitis) is the most common vasculitis in the pediatric population. It usually affects the skin, synovia, gastrointestinal tract, and kidneys. It usually presents as a palpable purpura. The occurrence of hemorrhagic bullae in children with HSP is an uncommon presentation. We present a case of an otherwise healthy 9-year-old male with a three-day history of erythematous maculopapular lesions over the lower extremities which progressed to violaceous plaques with central hemorrhagic bullae affecting the bilateral lower extremities, buttocks and arms. Odynophagia and intermittent abdominal pain were present. Histopathology revealed small vessel leukocytoclastic vasculitis and direct immunofluorescence (DIF) showed granular deposition of IgA and fibrinogen along the walls of the papillary dermal blood vessels. The patient was successfully treated with prednisone at 1mg/kg/day and showed resolution of lesions within 1 week of treatment with no recurrence at 1 month follow-up. We stress the importance of having a high index of suspicion in these atypical presentations in order to prevent delay in diagnosis and achieve maximal treatment gains.

Keywords: bullous henoch- schonlein purpura, pediatric, IgA vasculitis, rare, case report

INTRODUCTION

Henoch- Schonlein purpura (IgA vasculitis) is the most common vasculitis in the pediatric population. It accounts to 75% of cutaneous necrotizing vasculitis in children, and 25% in adults. It usually affects the skin, synovia, gastrointestinal tract, and kidneys. It usually presents as a palpable purpura (1). The occurrence of hemorrhagic bullae in children with HSP is an uncommon presentation which occurs in 2% of HSP cases (2). We present a case of a 9-year old Filipino male with bullous HSP successfully treated with oral corticosteroids.

CASE REPORT

An otherwise healthy 9-year-old male presented with a three-day history of erythematous maculopapular lesions over the lower extremities, which progressed to violaceous plaques with central hemorrhagic bullae. The lesions noted to gradually progress affecting the bilateral lower extremities, buttocks, and few on the arms. The lesions were noted to be pruritic and tender to touch. The patient denies manipulation of the lesions. It was accompanied by odynophagia and intermittent abdominal pain. No joint pains or urinary symptoms were noted. There was no family or personal history of skin disease. He consulted a pediatrician and was started on an unrecalled dose of Clindamycin which was subsequently shifted to Co-Amoxiclav which provided no relief.

Cutaneous examination revealed multiple erythematous macules, papules and plaques with hemorrhagic bullae over the bilateral lower legs, buttocks and few on the arms (Figure 1,2). Diascopy was positive.

Laboratory values on admission revealed leukocytosis (18.86 x 10^9/L [reference range: 4.00-10.00 x 10^9/L]) with neutrophilic predominance (0.82 [reference range: 0.350-0.850]), thrombocytopenia (502 x 10^9/L [reference range:

* Department of Dermatology, Rizal Medical Center
[1] Resident, Department of Dermatology, Rizal Medical Center
[2] Consultant, Department of Dermatology, Rizal Medical Center

150-450 x 10^9/L]), elevated erythrocyte sedimentation rate (40 mm/hr [reference range: 0-20 mm/hr]), normal blood urea nitrogen (4.00mmol/L [reference range: 2.5-6.4mmol/L]), low creatinine (45.50 umol/L [reference range: 62-115]), low sodium (133.0 mmol/L [135.0-148.0 mmol/L), normal potassium (4.01mmol/L [reference range: 3.5-53mmol/L]), low chloride (96.7 mmol/L [98.0-107 mmol/L]), and normal ionized calcium (1.24 mmol/L [1.13-1.32 mmol/L]). Urinalysis showed ketonuria (+1). No hematuria or proteinuria were noted. Fecal occult blood was weakly positive. Culture and sensitivity of the aspirate was done with negative results.

A skin biopsy was performed and histological examination revealed a small vessel leukocytoclastic vasculitis (Figure 5), and direct immunofluorescence analysis revealed granular deposition of IgA and fibrinogen along the walls of the papillary dermal blood vessels (Figure 6), consistent with Henoch- Schonlein Purpura.

The patient was started on Prednisone 20mg/5ml 7ml OD (1 mkD), Diphenhydramine 30mg/IV every 6 hours (1 mkd), and Omeprazole 30mg IV OD. The abdominal pain improved and the bullous lesions resolved within 1 week leaving only post inflammatory hyperpigmentation (Figure 3,4). No recurrence and adverse events were noted at 1 month follow up.

DISCUSSION

Henoch Schonlein Purpura (IgA vasculitis) is the most common vasculitis in children. It has an incidence of 20.4 per 100,000 per year. It is highest in children age 4 to 6 years old, and more common in males than female having a 1.2-1.0 ratio (3).

The diagnosis of HSP is based on the European League Against Rheumatism (EULAR) and the Paediatric Rheumatology European Society (PReS) consensus criteria for classification of childhood vasculitides. The criteria for diagnosing HSP would include a palpable purpura in the presence of at least one of the following features: diffuse abdominal pain, biopsy showing predominant IgA deposition, arthritis or arthralgia, and renal involvement (proteinuria or hematuria) (4).

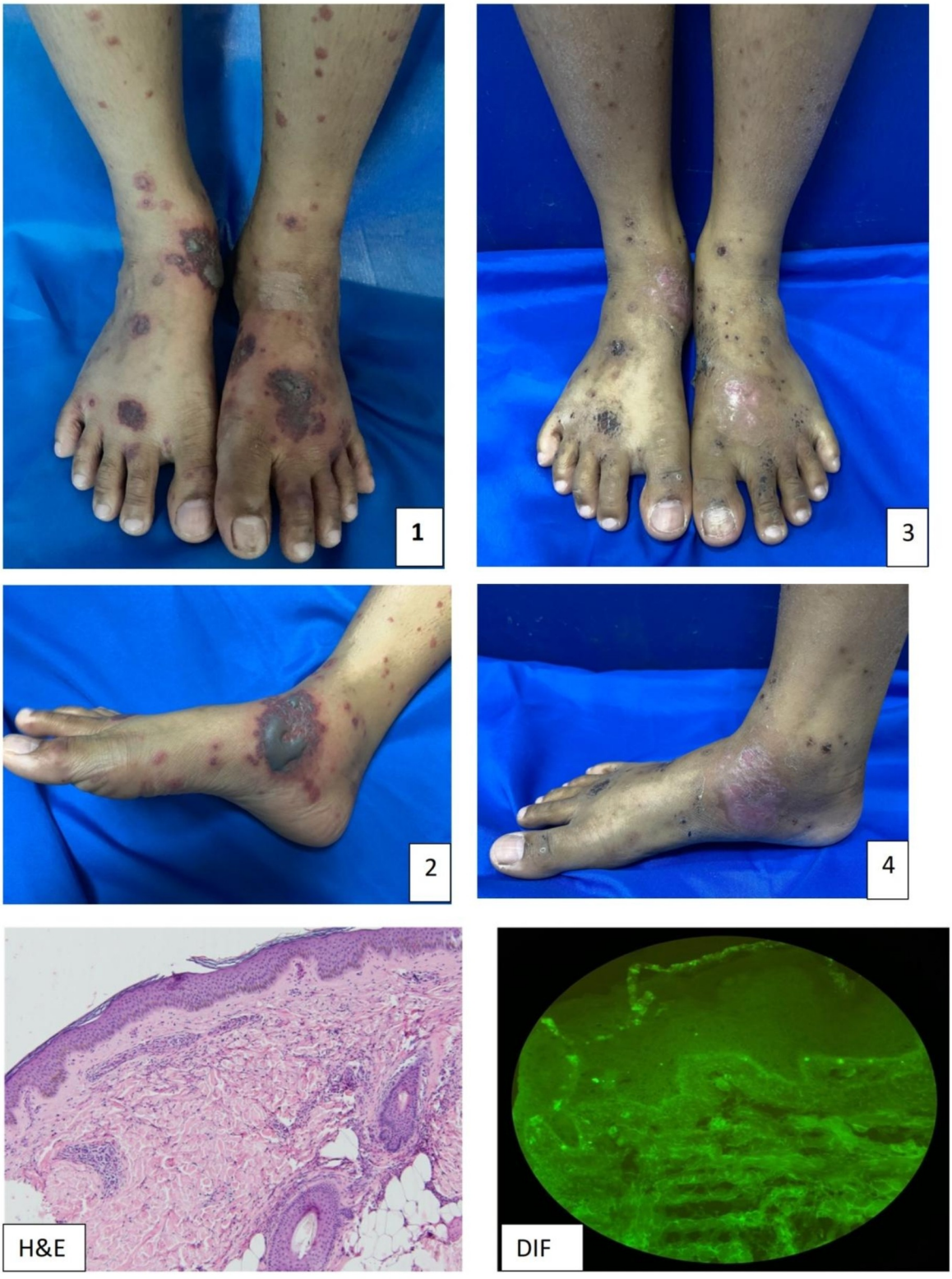
The occurrence of hemorrhagic bullae in children with HSP is an uncommon presentation which occurs in 2% of HSP cases (2). The pathogenesis of bullous lesions in HSP is unknown. One study in Japan reported elevated metalloproteinase-9 (MMP-9) in the blister fluid using zymography. MMP-9 which is secreted by polymorphonuclear leukocytes on the dermal side of the dermo- epidermal junction migrated from the lesions of intensive vasculitis. This could cause blister formation by degrading the basement membrane components such as type VII collagen (5). Early biopsy of the lesion is essential to make a diagnosis of bullous HSP. Ideally, biopsy must be done within 48 hours because immunoreactants such as IgA and C3 tend to destroyed during that timeframe (6). Histopathologic finding of leukocytoclastic vasculitis was reported on all cases, however IgA deposition are not positive in all cases (7). In a review of cases by Su et al, only 31.6% was positive for IgA which might be explained by the timing of the biopsy (8).

To date, there is still no guidelines regarding the management of hemorrhagic lesions in HSP. Several drugs has been used such as corticosteroids, anti-inflammatories, and immunosuppressants (9). Of which, the most commonly used drug in the management of bullous HSP are oral corticosteroids. Several reports have shown that early prednisone therapy reduces severity, extent, and sequelae of bullous lesions (10,11).

CONCLUSION

In summary, we report a case of a 9-year old male who presented with an uncommon presentation of Bullous Henoch Schonlein Purpura. The patent was diagnosed early thru histopathology and direct immunofluorescence. and was then successfully treated with oral corticosteroids which led to the resolution of all lesions with post inflammatory hyperpigmentation and no recurrence.

In cases like this we stress the importance of having a high index of suspicion in these atypical presentations in order to prevent delay in diagnosis. Referral to a dermatologist is also recommended. We also stress the importance of early biopsy and initiation of treatment to achieve maximal treatment gains.


1
3
2
4
H&E
DIF

REFERENCES

1. Wolff K, Goldsmith L, Katz S, Gilchrest B, Paller AS, Leffell D. Fitzpatrick's Dermatology in General Medicine, 8th Edition. New York: McGraw-Hill, 2011.
2. Saulsbury, Frank T. M.D. Henoch-Schonlein Purpura in Children: Report of 100 Patients and Review of the Literature, Medicine: November 1999 - Volume 78 - Issue 6 - p 395-409
3. Gardner-Medwin J, Dolezalova P, Cummins C, Southwood T. Incidence of Henoch-Schonlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. The Lancet. 2002;360(9341):1197-1202.
4. Ozen S. EULAR/PReS endorsed consensus criteria for the classification of childhood vasculitides. Annals of the Rheumatic Diseases. 2005;65(7):936-941.
5. Kobayashi T, Sakuraoka K, Iwamoto M, Kurihara S. A Case of Anaphylactoid Purpura with Multiple Blister Formation: Possible Pathophysiological Role of Gelatinase (MMP-9). Dermatology. 1998;197(1):62-64.
6. Chen, MD C, Garlapati, MD S, Lancaster, MD J, Zinn, MD Z, Bacaj, MD P, Patra, MD K. Bullous Henoch-Schonlein Purpura in Children. Pediatric Dermatology. 2015;96.
7. Nothhaft M, Klepper J, Kneitz H, Meyer T, Hamm H, Morbach H. Hemorrhagic Bullous Henoch-Schonlein Purpura: Case Report and Review of the Literature. Frontiers in Pediatrics. 2019;6.
8. Su H, Chen C, Chiou Y. Hemorrhagic bullous lesions in Henoch-Schonlein purpura: a case report and review of the literature. BMC Pediatrics. 2018;18(1).
9. Kausar S, Yalamanchili A. Management of haemorrhagic bullous lesions in Henoch–Schonlein purpura: Is there any consensus?. Journal of Dermatological Treatment. 2009;20(2):88-90.
10. Den Boer S, Pasmans S, Wulffraat N, Ramakers-Van Woerden N, Bousema M. Bullous lesions in Henoch Schonlein Purpura as indication to start systemic prednisone. Acta Paediatrica. 2010;99(5):781-783.
11. Kocaoglu C, Ozturk R, Unlu Y, Akyurek F, Arslan S. Successful Treatment of Hemorrhagic Bullous Henoch-Schonlein Purpura with Oral Corticosteroid: A Case Report. Case Reports in Pediatrics. 2013;2013:1-4.