Case Report

A rare presentation of Henoch-Schonlein Purpura with facial involvement in a teenager

Basnayake BMDB[1], Kannangara T[1], Dharmadasa DSP[1], Janappriya GHDC[2]

[1]Medical unit, Teaching Hospital, Kandy
[2]Dermatology unit, Teaching Hospital, Kandy

Abstract
Henoch-Schonlein purpura is a small vessel vasculitis characterized by cutaneous, joint, gastrointestinal and renal involvement. Vasculitic purpura is mainly seen on lower limbs & buttocks and rarely on face & upper limbs. We report a 13 year old girl with Henoch Schonlein purpura, who presented with purpuric rash predominantly involving the face, ears and upper limbs.

Keywords: Henoch schonlein purpura; Atypical presentation; Facial involvement; Duodenitis; Gastropathy

Funding: None

Competing interests: None

Received: 13 November 2015 **Accepted revised version:** 22 December 2015 **Published:** 26 December 2015

***Correspondence:** bmdbbasnayake@yahoo.com

Cite this article as: Basnayake BMDB, Kannangara T, Dharmadasa DSP, Janappriya GHDC. A rare presentation of Henoch-Schonlein Purpura with facial involvement in a teenager. *Anuradhapura Medical Journal* 2015; **9**: 41-43.
DOI: http://dx.doi.org/10.4038/amj.v9i2.7595

Introduction

Henoch-Schonlein purpura (HSP) is an acute, immune complex mediated leukocytoclastic vasculitis. Cutaneous involvement is typically seen on the lower limbs and buttocks but may also affect the face, upper limbs and trunk (1). We report a girl with HSP involving the face, ears and upper limbs.

Case presentation

A 13 year old girl presented with abdominal pain, fever and bilateral elbow joint pain for 3 days duration. On admission she developed passage of tarry black coloured loose stools. Then she developed painless echymotic rash on face and ears, which gradually involved both lower extremities & left upper limb, over the next 2-3 days. On examination she was febrile and ill looking. There were non-blanching palpable purpuric lesions on face, mainly on bilateral maxillary and left mandibular regions, and ears (Figure 1). Petechial and palpable purpuric lesions were symmetrically distributed over the feet (Figure 2).

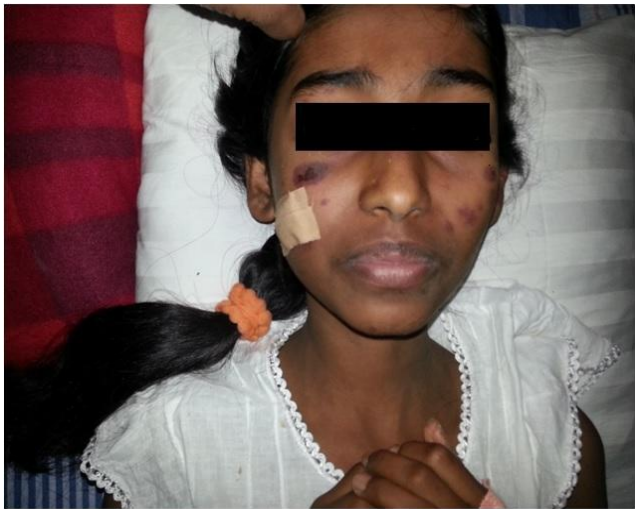

Figure 1 **Purpura involving the face & ears**

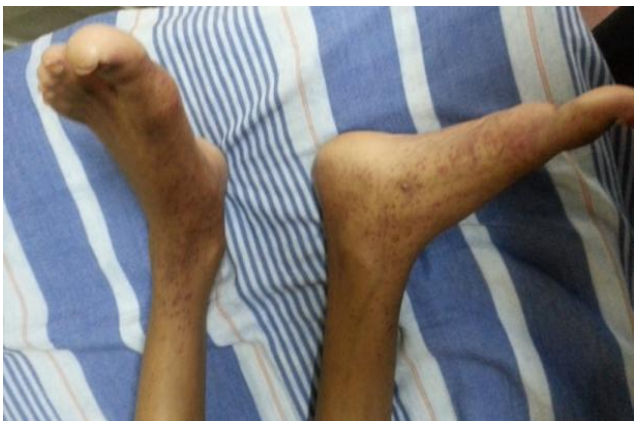

Figure 2 **Bilateral lower limb petechiae and palpable purpuric rash**

Abdominal examination revealed generalized tenderness, but no hepatosplenomegaly or free fluid was observed.

Digital rectal examination confirmed the presence of malena. Investigations showed haemoglobin 13.8 g/dl, WBC 9.8×10^9/l and Platelet 274×10^9/l. BU was 4.9 mmol/l with S. Cr 0.36 mg/dl. Na^+ 138 mmol/l and K^+ 3.8mmol/l.

On admission UFR was normal but 1 week later there were red cells, moderately field full with 80% dysmorphic cells. ESR was 12mm/1st hour with normal CRP. upper gastrointestinal endoscopy showed grossly oedematous gastric mucosa, erosions with gastropathy and duodenitis. ANA, rheumatoid factor, P ANCA and C ANCA were negative. Skin biopsy suggested the appearances are of leukocytoclastic vasculitis, compatible with HSP. Renal biopsy confirmed the diagnosis of HSP. She made a complete recovery with one week of treatment after intravenous steroids followed by two weeks of oral prednisolone therapy. Total hospital stay was three weeks and she was reviewed twice in the medical clinic. No complications were encountered.

Discussion

Henoch-Schonlein purpura is characterized by small vessel vasculitis involving the skin, GI tract, kidneys, joints, and rarely lungs and CNS. It is an acute IgA mediated disorder (2). The classic lesion typically affects extremities of the lower limbs, gluteal region but may involve upper limbs, trunk, face and mucous membranes (3). In this patient lesions were predominantly seen on face, ears and upper limbs with lesser involvement of lower limbs. The reason for the low incidence of facial involvement is not known. Some suggest that the location of the lesions depends on gravity. It would cause increased intravascular pressure in dependent areas such as the lower limbs and gluteal region, which stimulates extravasation of plasma in those regions resulting in oedema and hemorrhage (4, 5). In children under the age of two years with HSP, the purpuric skin rash frequently involves the face (6). An atypical facial manifestation might be mistaken for systemic lupus erythematosus, papular urticaria and meningococcemia (7). Although the facial involvement is unusual in the presentation of HSP, the diagnosis should therefore be considered in atypical cases. Clinical diagnosis based on a multidisciplinary approach and appropriate management will lead to a better clinical outcome.

Conclusion

Henoch-Schonlein purpura which classically affects extremities of lower limbs and gluteal region, can also present atypically with facial involvement. Thus better outcome relies on clinical acumen and appropriate management.

Consent

Informed written consent was obtained from the patient for publication of this case report and for all accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References

1. Kraft DM, Mckee D, Scott C. Henoch-Schönlein purpura: a review. *American Family Physician* 1998; **58**: 405–8.
2. Szer IS. Henoch-Schonlein purpura. *Current Opinion in Rheumatology* 1994; **6**: 25-31.
3. Azevedo CB de, Villella GMS, Nicolato AM, Carmo VMP do, Schlaucher SH, Bonfante H de L. Unusual face involvement in Henoch-Schonlein purpura. *Revista brasileira de reumatologia* 2009; **49**: 735-740. DOI: http://dx.doi.org/10.1590/S0482-50042009000600010
4. Nussinovitch M, Prais D, Finkelstein Y, Varsano I. Cutaneous manifestations of Henoch-Schonlein purpura in young children. *Pediatric Dermatology* 1998; **15**:426-8. DOI: http://dx.doi.org/10.1111/j.1525-1470.1998.tb01387.x
5. Sohagia AB, Gunturu SG, Tong TR, Hertan HI. Henoch-Schonlein Purpura - A case Report and review of the literature. *Gastroenterology Research and Practice* 2010; **2010**: Article ID 597648. DOI: http://dx.doi.org/10.1155/2010/597648
6. Al-Sheyyab M, El-Shanti H, Ajlouni S, Sawalha D, Daoud A. The clinical spectrum of Henoch-Schonlein purpura in infants and young children. *European Journal of Pediatrics* 1995; **154**:969-72. DOI: http://dx.doi.org/10.1007/BF01958639
7. Freiman A. Dermacase. Henoch-Schonlein purpura. *Canadian Family Physician* 2005; **51**:511-2.