Stroke and skin rash: A rare case of Henoch-Schonlein purpura

Kaushik Ghosh, Atri Chatterjee, Jyoti Tanmay Sau, Sandip Dey

Department of Medicine, Nilratan Sircar Medical College, Kolkata, West Bengal, India

Abstract

Henoch-Schonlein purpura (HSP) is a small vessel vasculitis that is rare in adults. Here, we present a case of a woman who presented with palpable purpura, abdominal pain, arthritis and ischemic stroke. The patient met the diagnostic criteria of HSP. However, cerebrovascular disease is reported as an uncommon, yet fatal, complication of HSP. The patient responded to aggressive immunosuppression with pulses of corticosteroids and cyclophosphamide. In the absence of an established protocol of treatment of such neurologic emergency in HSP patients, this report demonstrates a successful outcome.

Key Words

Cyclophosphamide, Henoch-Schonlein purpura, ischemic stroke

For correspondence:

Dr. Atri Chatterjee, 18/1E, D.P.P. Road, Kolkata, WB - 700 047, India. E-mail: chatterjee.atri@gmail.com

Ann Indian Acad Neurol 2012;15:307-9

Introduction

Henoch-Schonlein (HSP) is a small vessel vasculitis that involves joints, skin, kidneys and gastrointestinal tract.[1] It commonly affects children and young adults and presents classically with palpable purpura, abdominal pain and joint symptoms. Adults are less affected by HSP, although complications are more common in this age group.[2] The central nervous system (CNS) is rarely involved in HSP.[3] As CNS affection is more likely in other vasculitis syndromes, it is necessary to exclude these causes. CNS manifestations include seizures,[4] stroke, visual abnormalities as well as speech disturbances.[3] Patients who develop stroke can have either multiple infarcts or hemorrhage, and may have raised anti-phosphatidylethanolamine antibody in serum.[5] Usually, these patients do not have hypertension. On the other hand, hypertensive patients are reported to have a reversible CNS dysfunction.[6] Neuroimaging studies in patients of HSP with CNS features show a predominant pattern of infarcts affecting two vessel territories, and these patients require aggressive immunosuppression.[3]

Access this article online	
Quick Response Code:	Website: www.annalsofian.org
	DOI: 10.4103/0972-2327.104343

Case Report

A 46-year-old lady was admitted for sudden onset of weakness of the right side of the body and facial deviation without any history of unconsciousness, headache, vomiting or seizure. She was diagnosed as a case of hypertension 15 days back, for which she was given metoprolol 25 mg/day. In the emergency room, examination revealed stable vitals, blood pressure 170/110 mmHg, Glasgow coma score 12, right-sided hemiparesis and left-sided upper motor neuron facial nerve palsy. She also had palpable purpura over both the shins and buttocks and a digital infarct of the left thumb. Systemic examination was otherwise normal.

The patient had developed swelling and redness of both the knees and ankles 3 weeks prior to admission, which was associated with severe abdominal pain. She was prescribed corticosteroids for 1 week, which she stopped after relief of symptoms. Soon after withdrawal of the drug, she developed gradual darkening of the tip of the left thumb and painful crops of reddish lesions appeared over her legs and buttocks [Figure 1].

On admission, she underwent routine hematologic and biochemical testing and a noncontrast computed tomography (CT) scan of the brain. There was normochromic normocytic anemia, leukocytosis, raised erythrocyte sedimentation rate (ESR) and normal platelet count [Table 1]. Her blood urea nitrogen and creatinine was mildly elevated, while other biochemical parameters remained within normal limits. CT brain showed multiple infarcts in the left internal capsule, basal ganglia and right thalamus [Figure 2]. A routine urinalysis revealed presence of albumin and active urine sediment of red

blood cells (RBC) and RBC casts. Estimation of 24-h urinary protein showed nonnephrotic range proteinuria [Table 1]. Ultrasonography of the kidneys showed normal kidney size with increased cortico-medullary echogenecity. Tests for antinuclear factor, anti-neutrophil cytoplasmic antibody (p- and c-ANCA) and anti-phospholipid antibody were negative. A punch biopsy was taken from the skin lesions over the legs, which revealed leucocytoclastic vasculitis [Figure 3].

The patient was started on low-dose aspirin, metoprolol and ramipril. She remained conscious, oriented and afebrile, without new neurological deficits, and maintained normal urine output. However, on the fourth day of hospital stay, there was onset of abdominal pain and inflammation of both knee joints.

The clinical features and biopsy evidence supported the diagnosis of HSP, although age and presentation of the patient were not typical of the disease. Considering the presence of both renal and cerebrovascular involvement, the patient was administered intravenous cyclophosphamide 500 mg/sq meter on Day 1 combined with methylprednisolone 1 gm/day for Days 1–3. This was followed by a maintenance dose of prednisolone 0.5 mg/kg/day.

The patient was readmitted after 1 month for follow-up and a repeat pulse of cyclophosphamide and methylprednisolone. On clinical examination, her paresis had improved. Routine investigations of blood and urine showed an improvement of anemia and absence of active urinary sediment. She is scheduled to receive four more monthly pulses of cyclophosphamide and methylprednisolone along with maintenance dose of prednisolone.

Table 1: Summary of laboratory tests

Blood tests	On admission	On follow-up
Hemoglobin (gm/dL)	7.6	10.2
ESR (mm)	110	30
Leucocyte count (/cmm)	14,500	11,200
Platelet count (/cmm)	300,000	140,000
MCV (fl)	79.9	85.8
MCH (pg)	23.5	25.5
MCHC (gm%)	29.5	29.7
Urea (mg/dL)	30	38
Creatinine	0.7	0.7
Sodium (meq/L)	128	134
Potassium (meq/L)	4.2	4.4
Fasting blood glucose (mg/dL)	72	80
Anti-nuclear antibody	Negative	-
p-ANCA	Negative	-
c-ANCA	Negative	-
Anti-phospholipid antibody	Negative	-
Urine examination:		
Albumin	++	-
Red blood cells	14–16	-
Pus cells	4–6	4–6
Casts	RBC casts	-
24-h urinary protein (gm)	0.6	0.1

MCV: Mean corpuscular volume ESR: Erythrocyte sedimentation rate MCH: Mean corpuscular hemoglobin MCHC: Mean corpuscular hemoglobin concentration p-ANCA: Anti neutrophin cytoplasmic antibody c-ANCA: Anti neutrophin cytoplasmic antibody

Discussion

According to the American College of Rheumatology criteria

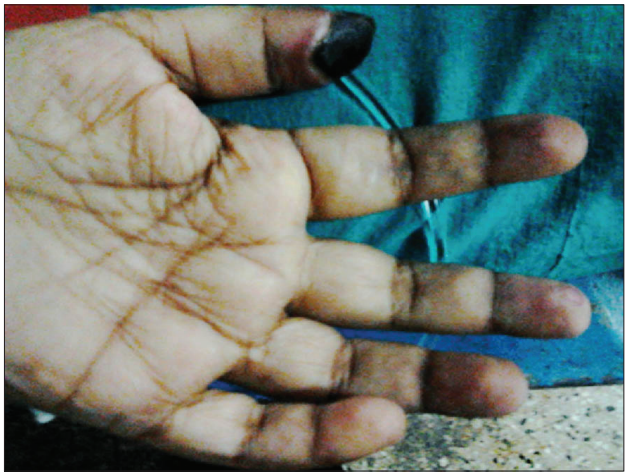

Figure 1: Dry gangrene of the left thumb due to vasculitis

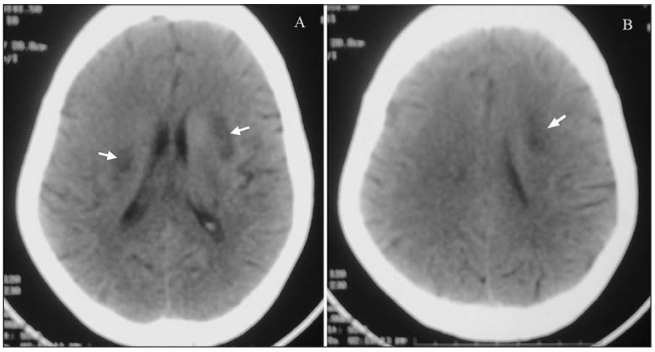

Figure 2: Noncontrast computed tomography scan of the brain. Arrowheads show bilateral basal ganglia infarcts

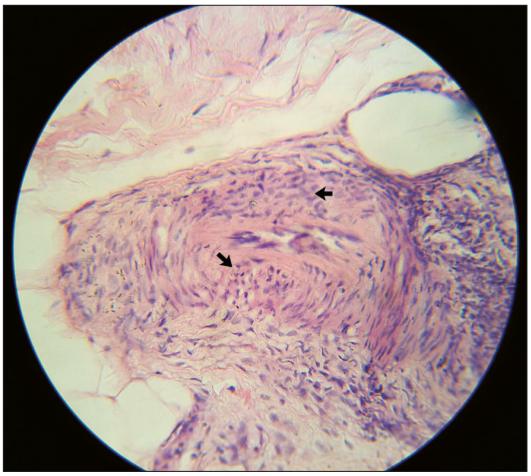

Figure 3: Skin biopsy from the palpable purpura site, stained with hematoxylin and eosin, x10 magnification, and showing leucocytoclastic vasculitis. Arrowheads indicate neutrophils infiltrating the vessel wall

(1990), diagnosis of HSP requires presence of two or more of the following criteria: age less than 20 years, palpable purpura, acute abdominal pain and presence of granulocytes in the walls of small vessels. Although our patient is 46 years old, she meets two of these criteria. She was therefore diagnosed as a case of HSP.

Apart from classical involvement of skin, kidney, gut and joints, complications affecting the CNS have rarely been reported. Cerebrovascular accident is a rare complication HSP, which is mainly described in children and young adults.[7,8] But, adult patients with HSP suffering from cerebrovascular accident is extremely rare. Karamadoukis *et al*.,[9] and Lévaif F *et al*.[10] described adult-onset HSP with intracerebral hemorrhage as a rare complication. To the best of our knowledge, there is only one other published report of cerebral infarction in a patient of HSP.[11] Acute rise of blood pressure has been considered to be the primary mechanism of intracerebral hemorrhage.[12] Other possible causes include the presence of cerebral vasculitis and reduced levels of factor XIII[13] and prothrombin.[14] However, the etiology behind cerebral infarction remains obscured till date. The management of cerebral infarction in older HSP patient lacks proper guideline as the reported incidence is low. Intravenous pulse methylprednisolone followed by oral steroids has been shown to be effective in the management of severe HSP nephritis.[15] In our patient, both HP and its neurological symptoms were successfully treated with intravenous cyclophosphamide and methylprednisolone, followed by oral prednisolone. In conclusion, we present a case of HSP with an atypical presentation of stroke in an adult female that responded to immunosuppression with cyclophosphamide in addition to steroid therapy.

References

1. Mills JA, Michel BA, Bloch DA, Calabrese LH, Hunder GG, Arend WP, *et al*. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura. Arthritis Rheum 1990;33:1114-21.
2. Ricardo B, Victor MM, Vicente RV, Miguel GF, Miguel AG. Henoch-Schönlein purpura in adulthood and childhood: Two different expressions of the same syndrome. Arthritis Rheum 1997;40:859-64.
3. Garzoni L, Vanoni F, Rizzi M, Simonetti GD, Goeggel Simonetti B, Ramelli GP, *et al*. Nervous system dysfunction in Henoch–Schönlein syndrome: Systematic review of the literature. Rheumatology 2009;48:1524-9.
4. Fielding RE, Hawkins CP, Hand MF, Heath PD, Davies SJ. Seizures complicating adult Henoch-Schonlein purpura. Nephrol Dial Transplant 1998;13:761-2.
5. Sokol DK, McIntyre JA, Short RA, Gutt J, Wagenknecht DR, Biller J, *et al*. Henoch–Schönlein purpura and stroke: Antiphosphatidyl-ethanolamine antibody in CSF and serum. Neurology 2000;55:1379-81.
6. Sasayama D, Shimojima Y, Gono T, Kaneko K, Matsuda M, Ikeda S. Henoch–Schönlein purpura nephritis complicated by reversible posterior leukoencephalopathy syndrome. Clin Rheumatol 2007;26:1761-3.
7. Clark JH, Fitzgerald JF. Hemorrhagic complications of Henoch-Schönlein syndrome. J Pediatr Gastroenterol Nutr 1985;4:311-5.
8. Chiaretti A, Caresta E, Piastra M, Pulitano S, Di Rocco C. Cerebral hemorrhage in Henoch-Schönlein syndrome. Childs Nerv Syst 2002;18:365-7.
9. Lazarus K, Linmarie L, Williams A. Henoch-Schönlein purpura with intracerebral haemorrhage in an adult patient: A case report**.** J Med Case Reports 2008;2:200.
10. Lévaif F, Szücs LH, Jászai Z. Cerebral hemorrhage and acute glomerulonephritis in Schonlein-Henoch syndrome in old age. Z Gesamte Inn Med 1971;26:309-11.
11. Zhang S, Liu GS. Report of a case with cerebral infarction in Henoch-Schonlein purpura. Zhonghua Er Ke Za Zhi 2007;45: 147-8.
12. Sang-Wuk J, Keun-Hwa J, Kon C, Hee-Joon B, Seung-Hoon L, Jae-Kyu R: Clinical and radiologic differences between primary intracerebral hemorrhage with and without microbleeds on gradient-echo magnetic resonance images. Arch Neurol 2004;61:905-9.
13. Imai T, Okada H, Nanba M, Kawada K, Kusaka T, Itoh S. Henoch-Schönlein purpura with intracerebral hemorrhage. Brain Dev 2002;24:115-7.
14. Clark JH, Fitzgerald JF. Hemorrhagic complications of Henoch-Schönlein syndrome. J Pediatr Gastroenterol Nutr 1985;4:311-5.
15. Niaudet P, Habib R. Methylprednisolone pulse therapy in the treatment of severe forms of Schonlein-Henoch purpura nephritis. Pediatr Nephrol 1998;12:238-43.

How to cite this article: Ghosh K, Chatterjee A, Sau JT, Dey S. Stroke and skin rash: A rare case of Henoch-Schonlein purpura. Ann Indian Acad Neurol 2012;15:307-9.
Received: 14-12-11, **Revised:** 14-01-12, **Accepted:** 24-01-12
Source of Support: Nil, **Conflict of Interest:** Nil