CASE REPORT

Guillain-Barre syndrome in an adult patient with Henoch-Schonlein purpura

Puneet Aggarwal,¹ Ratnakar Sahoo,¹ A K Gadpayle,¹ Gunjan Kumar²

¹Department of Internal Medicine, Post Graduate Institute of Medical Education & Research & Dr Ram Manohar Lohia Hospital, New Delhi, India

²Department of Internal Medicine, Dr Ram Manohar Lohia Hospital, New Delhi, India

Correspondence to

Dr Puneet Aggarwal,
puneetagarwal4u@gmail.com

SUMMARY

A 42-year-old man presented with pain in the abdomen, massive haematemesis and rashes over the body and development of bilateral lower limb weakness the next day. The patient was later diagnosed with Guillain-Barre syndrome with Henoch-Schonlein purpura. He was treated with intravenous immunoglobulins. At 3 months of follow-up and rehabilitation, patient showed complete recovery from weakness.

BACKGROUND

Henoch-Schonlein purpura (HSP) is an autoimmune disorder owing to small vessel vasculitis and preferentially affects children. Neurological manifestations are rare in patients with HSP. Involvement of peripheral nervous system is even rarer in HSP. In the present case, Guillain-Barre syndrome (GBS) occurred in association with HSP in an adult patient.

CASE PRESENTATION

A 42-year-old non-alcoholic male patient presented with complains of acute onset severe pain in the abdomen, two episodes of haematemesis and rashes over the body. Next day the patient developed weakness in both the lower limbs. There was no history of any fever, recent vaccinations, animal bite, loose stools, cough or sore throat. The patient had no history of similar episodes. There was no history of any joint pain or joint swelling.

On examination, patient had tachycardia (pulse rate 102/min), pallor and weak peripheral pulses. Patient had palpable purpuric rashes throughout the body except oral mucosa, palms, sole and face (figures 1 and 2). Abdominal examination revealed tenderness in the epigastric region. There was no

free fluid in the abdomen. Cardiovascular and respiratory system examination were unremarkable.

Neurological examination revealed flaccid areflexic paralysis of limbs. Abnormality on sensory examination was limited to involvement of posterior column sensations. Bowel and bladder were not involved. Cranial nerve examination was normal. There was no respiratory muscle weakness.

INVESTIGATIONS

Investigations revealed anaemia (haemoglobin 7.2 g/dl), polymorphonuclear leucocytosis (total leucocyte count 14 000/mm³) and normal platelet count. Liver function tests and renal function tests were within normal limits. Coagulation profile was within normal limit and fibrin degradation products were absent. Urinary examination was normal with no proteinuria or casts. The patient had increased C reactive protein, immunoglobulin A (IgA) level and normal complement levels and other immunoglobulin levels. Antinuclear antibody, perinuclear antineutrophil cytoplasmic antibodies (p-ANCA) and cytoplasmic ANCA (c-ANCA) were negative.

Lumbar puncture performed in second week of weakness revealed a clear cerebrospinal fluid (CSF) with an elevated CSF protein (120 mg/dl) without accompanying pleocytosis. There were no oligoclonal bands in the CSF.

MRI of the spine revealed no abnormality.

Motor nerve conduction studies demonstrated prolongation of the distal latencies along with reduced motor conduction velocities. Reduction of compound muscle action potential (CMAP) amplitude with temporal dispersion in the morphology of the CMAP was noted. F waves had poor persistence with prolongation of minimum F wave latencies. H reflex latency on stimulation of the right

Figure 1 Diffuse purpuric rashes over bilateral lower limbs.

Figure 2 Diffuse purpuric rashes over lower limb.

To cite: Aggarwal P, Sahoo R, Gadpayle AK, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-007945

tibial nerve was prolonged. CMAP amplitude on right peroneal nerve stimulation at ankle, below neck of fibula and above neck of fibula was 1, 0.2 and 0.2 mv, respectively. The motor conduction velocity of the peroneal nerve across neck of fibula and over below neck of fibula-ankle segment was 12 and 10 m/s, respectively, with distal latency of right peroneal nerve at ankle being 21 m/s. On stimulation of the right peroneal nerve at ankle F waves were seen occasionally with minimum latency at 98 m/s. The nerve conduction velocity in case of median as well as ulnar nerves were <40 m/s. Abnormalities of motor conduction were more marked in the nerves of the lower limbs as compared with the upper limbs. Sensory nerve conduction studies were within normal limits. The findings were consistent with demyelinating motor neuropathy involving all the four limbs.

Skin biopsy revealed keratinised stratified squamous epithelium with underlying dermal blood vessels showing swelling of endothelial cells and deposits of strongly eosinophilic strands of fibrin and fragmented nuclei within and around the vessel walls; with oedema and neutrophilic infiltrate around them suggestive of leucocytoclastic vasculitis (figures 3 and 4) with deposits of IgA and C3 in dermal capillaries.

Upper gastrointestinal endoscopy revealed multiple gastric ulcers with punctate bleeding, erosive ulcers in the duodenum and multiple erosions in lower one-third of oesophagus.

TREATMENT

The patient was managed with blood transfusion, proton pump inhibitors and intravenous fluids. After a clinical diagnosis of HSP and GBS, patient was treated with intravenous immunoglobulin (IVIg) infusions over 5 days for a total dose of 2 g/kg body weight. Rehabilitation with regular physiotherapy was performed.

OUTCOME AND FOLLOW-UP

A gradual improvement in symptoms and clinical status was observed. Patient had no further episode of haematemesis. After 5 days of IVIg treatment patient started showing improvement in weakness.

At 3 months of follow-up and rehabilitation, patient showed complete recovery from weakness.

DISCUSSION

HSP is an autoimmune disorder that preferentially affects children. It is characterised clinically by punctate haemorrhages,

Figure 3 Skin biopsy of the patient showing leucocytoclastic vasculitis in low power.

Figure 4 Skin biopsy of the patient showing leucocytoclastic vasculitis in high power.

arthralgia and gastrointestinal symptoms; histological examination reveals systemic necrotising vasculitis of small vessels. The diagnosis of HSP in our patient was based on three criteria. *The American College of Rheumatology 1990 criteria*¹ which includes the presence of two or more of the following criteria:

1. Palpable purpura
2. Age at onset <20 years
3. Abdominal pain
4. Wall granulocytes on biopsy

*Ankara 2008 classification definition*² for diagnosis of HSP includes purpura or petechiae (mandatory) with lower limb predominance and at least one of the four following criteria:

1. Abdominal pain
2. Histopathology
3. Arthritis or arthralgia
4. Renal involvement

*International Consensus Conference, 2006*³ which includes palpable purpura in the presence of one or more of the following criteria:

1. Diffuse abdominal pain
2. Any biopsy showing predominant IgA deposition
3. Arthritis (acute, any joint) or arthralgia
4. Renal involvement (any haematuria or proteinuria)

Neurological manifestations are rare in patients with HSP^{4 5} Involvement of peripheral nervous system is even rarer in HSP⁶ In a review by Garzoni *et al.*⁷ GBS is seen in 6% of patients with neurological involvement in HSP.

The diagnosis of GBS is based on characteristic signs and symptoms, with confirmation from laboratory and nerve conduction studies. The characteristic laboratory findings are an elevated CSF protein in the absence of pleocytosis. Nerve conduction studies supportive of GBS include delayed motor nerve conduction velocity of lower limb nerves and decreased M wave altitude. The patient in the present report fulfilled the modified criteria proposed by an ad hoc committee of the U.S. *National Institute of Neurological Disorders and Stroke*⁸ which is as follows:

Features required for diagnosis

- ▶ Progressive weakness in both arms and both legs
- ▶ Areflexia

Features strongly supporting diagnosis

- ▶ Progression of symptoms over days to 4 weeks
- ▶ Relative symmetry of symptoms
- ▶ Mild sensory symptoms or signs
- ▶ Cranial nerve involvement, especially bilateral weakness of facial muscles

- ▶ Recovery beginning 2–4 weeks after progression ceases
 - ▶ Autonomic dysfunction
 - ▶ Absence of fever at onset
 - ▶ High concentration of protein in CSF, with fewer than $10 \times 10^6/l$
 - ▶ Typical electrodiagnostic features
- Features excluding diagnosis
- ▶ Diagnosis of botulism, myasthenia, poliomyelitis or toxic neuropathy
 - ▶ Abnormal porphyrin metabolism
 - ▶ Recent diphtheria
 - ▶ Purely sensory syndrome without weakness

Involvement of the peripheral nervous system in HSP is extremely rare. Belman *et al*⁶ in 1985 described three patients of HSP with neurological manifestations including one with brachial plexopathy and transient leg weakness. Sanghavi and Sharma⁹ and Moreau *et al*¹⁰ also demonstrated similar presentation of HSP in an adult patient.

Pathogenesis of GBS in HSP is still not clearly understood. Initially an allergic or hypersensitivity mechanism was proposed.⁹ Moreau *et al*¹⁰ demonstrated IgA immune complexes, a characteristic of HSP were implicated in the causation of peripheral demyelination and consistent histological lesions were seen on neuromuscular biopsy.

The myelin destruction in GBS may be mediated through the immunoglobulin receptors present in various cells in the peripheral nervous system, particularly the Schwann cells and endoneurial macrophages,¹¹ or through the interaction of these antibodies with various target antigens like gangliosides, myelin-associated glycoproteins, etc in the peripheral nerves.^{12 13}

In HSP complicated by GBS, management similar to the classical form of this polyneuropathy, with IVIg (or plasma exchange) is recommended.¹⁴

Learning points

- ▶ Henoch-Schönlein purpura should be considered as a possible diagnosis in a patient with palpable purpura irrespective of age.
- ▶ Neurological manifestations are rare in patients with Henoch-Schönlein purpura.
- ▶ Guillain-Barre syndrome is one of the neurological manifestations seen in Henoch-Schönlein purpura.

Acknowledgements Special thanks to Dr Ketan Garg, ICMR & department of pathology, VMCC & SJH for providing the histopathological diagnosis of the patient.

Contributors PA contributed to work up of patient, manuscript design, writing, forming and review of the literature. RS contributed to review of the literature. GK and AKG proof read the manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Mills JA, Michel BA, Bloch DA, *et al*. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura. *Arthritis Rheum* 1990;33:1114–21.
- 2 Ozen S, Pistorio A, Iusan SM, *et al*. EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria. *Ann Rheum Dis* 2010;69:798–806.
- 3 Dillon MJ, Ozen S. A new international classification of childhood vasculitis. *Pediatr Nephrol* 2006;21:1219–22.
- 4 Ashwal S, Schneider S. Neurologic complications of vasculitic disorders of childhood. In: Swaiman KF, ed. *Pediatric neurology, principles and practice*. St. Louis, MO: The CV Mosby Company, 1989:625–41.
- 5 Ritter FJ, Seay AR, Lahey ME. Peripheral mononeuropathy complicating anaphylactoid purpura. *J Pediatr* 1983;103:77–8.
- 6 Belman AL, Leicher CR, Moshe SL, *et al*. Neurologic manifestations of Schönlein-Henoch purpura: report of three cases and review of the literature. *Pediatrics* 1985;75:687–92.
- 7 Garzoni L, Vanoni F, Rizzi M, *et al*. Nervous system dysfunction in Henoch-Schönlein syndrome: systematic review of the literature. *Rheumatology (Oxford)* 2009;48:1524–9.
- 8 Asbury AK, Cornblath DR. Assessment of current diagnostic criteria for Guillain-Barré syndrome. *Ann Neurol* 1990;27(Suppl):S21–4.
- 9 Sanghavi LM, Sharma R. Guillain-Barré syndrome and presumed allergic purpura. *Arch Neurol Psych* 1956;76:497–9.
- 10 Moreau BA, Schuller E, Georges B. Association of Guillain-Barre syndrome and Henoch-Schönlein purpura: is immunoglobulin A responsible for the neurological syndrome? *Am J Med Sci* 1988;296:198–201.
- 11 Vedeler C, Ulvestad E, Nyland H, *et al*. Receptors for immunoglobulin in the central and peripheral nervous system. *J Neurol Neurosurg Psychiatry* 1994;57(Suppl):9–10.
- 12 Steck AJ, Kappos L. Gangliosides and autoimmune neuropathies: classification and clinical aspects of autoimmune neuropathies. *J Neurol Neurosurg Psychiatry* 1994;57(Suppl):26–8.
- 13 Vander Meche FGA, van Dröon PA. Guillain-Barre syndrome and chronic inflammatory demyelinating polyneuropathy: immune mechanisms and update on current therapies. *Ann Neurol* 1995;37(Suppl 1):S14–31.
- 14 Kuwabara S. Guillain-Barré syndrome: epidemiology, pathophysiology and management. *Drugs* 2004;64:597–610.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow