CASE REPORT

Posterior reversible encephalopathy syndrome in a child with Henoch-Schönlein purpura

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SUMMARY

Henoch-Schönlein purpura (HSP) is a small vessel vasculitis that affects the gastrointestinal and central nervous systems and the kidneys. The disease primarily affects children, but may occur in elderly children with allergic purpura and also in adults. Central nervous system involvement may be the first sign; however, it is rarely encountered. Posterior reversible encephalopathy syndrome (PRES) is a clinical syndrome of encephalopathy, headache, visual disturbance and seizures. Its radiological signs can be observed in grey and white matter at the posterior region of the cerebral hemispheres. HSP should be considered in children with PRES in the presence of rash, joint and gastrointestinal symptoms. We reported a 5-year-old patient who developed acute renal failure and PRES by reason of HSP.

BACKGROUND

Without clinical data of Henoch-Schönlein purpura (HSP), the radiologist may misinterpret the MR findings as progressive multifocal leukoencephalopathy, gliomatosis cerebri, demyelinating-dysmyelinating disease, cerebral ischaemia or postictal changes. HSP should be considered in children with posterior reversible encephalopathy syndrome (PRES) in the presence of rash, joint and gastrointestinal symptoms.

CASE PRESENTATION

A 5-year-old girl presented with purpuric rash on lower extremities, arthralgia, headache as well as abdominal pain. On examining the medical history, it was found that she had nasal discharge and a sore throat due to an upper respiratory tract infection 1 week ago. She did not have a food or drug allergy or an insect bite. And she was not on any medications.

On clinical examination, she was found to be conscious and afebrile. Pulse rate was 110/min, arterial blood pressure (ABP) 180/110 mm Hg and respiratory rate 18/min. Purpuric lesions looked like the knob of a needle and were tiny, red and slightly raised from the skin. They were detected on the extensor surfaces of the lower extremities and the buttocks. She had knee, elbow, wrist and ankle pain without any swelling. Bilateral cortical blindness was present in the neurological examination. She had periumbilical tenderness in the abdominal region.

INVESTIGATIONS

Laboratory examination revealed microcytic anaemia: haemoglobin: 9 g/dL, white cell count: $8500 \times 10^3/\text{mm}^3$, thrombocytes: $150 \times 10^3/\text{mm}^3$,

antistreptolysin O: 1000 IU/dL. Sedimentation rate was 70 mm/h. The throat culture was negative. Blood urea was 200 mg/dL and creatine was 3 mg/dL. The urine examination revealed 20–25 erythrocytes and no leucocytes. We did not find any other abnormalities.

We planned to perform a dialysis owing to acute renal failure. Cranial MR was carried out before the dialysis because the patient had a seizure attack. According to grey matter mild hypointense on T1-weighted view and high signal on fluid-attenuated inversion recovery and T2-weighted views and gyral swelling were seen in the both bilateral occipital-parietal lobes (figures 1 and 2). Diffusion restriction was not determined in the diffusion weighted imaging (DWI). Before the dialysis, contrast examination was undertaken for the differential diagnosis of the designated lesions. Cortico-subcortical asymmetric patchy contrasts were observed in the described areas and in the right frontal lobe (figure 3). These findings were compatible with PRES. The case was diagnosed as HSP after the skin biopsy. We considered that the cranial findings were due to hypertension and uraemic encephalopathy caused by acute renal failure.

DIFFERENTIAL DIAGNOSIS

- ▶ Progressive multifocal leukoencephalopathy
- ▶ Gliomatosis cerebri
- ▶ Demyelinating-dysmyelinating disease
- ▶ Cerebral ischaemia
- ▶ Postictal changes

Figure 1 T1-weighted image shows marked gyral swelling and isointense areas according to the cortex in the posterior parietal regions (arrows).

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Figure 2 T2-weighted (A) and fluid-attenuated inversion recovery (B) images show cortico-subcortical gyral oedema and hyperintense areas in bilateral parietal lobes and the left frontal lobe (arrows).

TREATMENT

The patient was treated as symptomatic for hypertension and seizure attack. Control of hypertensive emergency was an important part of the symptomatic management. Hypoglycaemia should be looked for routinely and corrected. The dialysis was performed owing to acute renal failure.

OUTCOME AND FOLLOW-UP

Clinical and recurrent MR findings improved completely after 4 weeks. We considered that the cranial findings were due to hypertension and uraemic encephalopathy caused by acute renal failure.

DISCUSSION

HSP is a small vessel vasculitis which affects the skin, joints, gastrointestinal and central nervous systems and the kidneys. Specific products like IgA suggest that an abnormal immune response affects small vessels and causes the disease.¹ The most frequent sign of HSP is the purpuric rash on the buttocks and the extensor surfaces of the lower extremities. Joints are affected in 65% and kidneys in 20–35% of cases. Acute renal failure has been reported in 1–5% of cases. The reason behind the

Figure 3 Contrast-enhanced T1 image shows cortico-subcortical patchy contrasts in bilateral parietooccipital areas (arrow).

neurological abnormality is not exactly known. The first neurological abnormality was defined by Osler in a patient with HSP who later became hemiplegic.² De montis and Turpin reported that the incidence of repetitive seizures, confusion and stroke was 6.9% in 244 patients with HSP. Thirty one per cent of cases had headache and 46% had EEG abnormality in a prospective study containing 26 patients.³ The reported central nervous system symptoms were headache, altered mental status, bilateral cortical blindness, seizures and focal neurological deficits. Mononeuropathy, polyneuropathy and plexiopathies were peripheral nervous system symptoms.⁴ We observed headaches and seizures in our patient.

Appearance of neurological signs in patients with HSP occurs with a variety of mechanisms. Ischaemic infarct areas may exist after small vessel vasculitis in the brain. In this instance, cytotoxic oedema will occur in the cells and cause diffusion restriction on DWI. In our case, we did not observe diffusion restriction. However, there was signs of vasogenic oedema. We considered that vasogenic oedema developed due to hypertension, uraemic encephalopathy and electrolyte disturbances resulting from acute renal failure. Cranial MR impressions on the parieto-occipital and frontal lobes were consistent with PRES. Intracerebral haematoma and vasculitis signs were reported in the literature and these signs resulted in permanent symptoms.⁵ In PRES, we detected that symptoms and MR signs improved completely after 4–6 weeks.

The pathophysiology of PRES is complicated. The blood–brain barrier breaks down due to dilation of the arteriols, then transudation and extravasation of the fluid and the macromolecules from the arteriolar wall occur. Interstitial fluid accumulation takes place in the cortical and subcortical white matter. Eventually vasogenic oedema occurs instead of cytotoxic oedema.³ The parieto-occipital region is particularly affected. One might consider that this is because the posterior circulation has fewer sympathetic innervation than the anterior circulation. PRES also affects border zone areas in the frontal lobe, cerebellum, basal ganglia and rarely the peduncle.^{2–5}

The causes of PRES are acute–subacute hypertension,^{6 7} uraemic encephalopathy⁸ (acute glomerulonephritis, hemolytic uremic syndrome, thrombotic thrombocytopenic purpura), drugs (cyclosporine, FK-506, tacrolimus, cisplatin, interferon- α , erythropoietin) and porphyria. It is considered that immunosuppressive drugs break down the blood–brain barrier and cause PRES. In our case we considered that PRES has happened due to hypertension and uraemic encephalopathy resulting from acute renal failure which may rarely be observed during the course of HSP. Clinical and MR findings were completely improved 4 weeks after therapy.

Learning points

- ▶ Henoch-Schönlein purpura is a small vessel vasculitis which affects the skin, joints, gastrointestinal and central nervous systems and the kidneys.
- ▶ Henoch-Schönlein purpura should be considered in children with posterior reversible encephalopathy syndrome in the presence of rash, joint and gastrointestinal symptoms.
- ▶ Clinical and MR findings start to improve 4–6 weeks after therapy.

Competing interests None.

Patient consent Obtained.

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