Case Report

Henoch-Schonlein Purpura with Intracerebral Haemorrhage

AK Misra*, A Biswas*, SK Das**, PK Gharai+, T Roy***

Abstract

Henoch-Schonlein purpura is a leucocytoclastic vasculitis commonly seen among children and young adults. Neurological complications, though rare, include focal cerebral deficit, coma, convulsion, subarachnoid hemorrhage and chorea. We are reporting a 12 years boy with Henoch-Schonlein purpura who developed a large intracerebral hematoma in right occipital lobe. He made an uneventful recovery with conservative treatment and one year follow up revealed no major neurological sequelae.

INTRODUCTION

Henoch-Schonlein purpura (HSP) is a distinct, common, self-limited, subacute type of leucocytoclastic vasculitis, seen primarily among children and young adults. It is characterized by non-thrombocytopenic symmetrical purpuric skin rash, arthralgia, abdominal pain, gastrointestinal haemorrhage and immune complex nephritis.

Neurological complications include focal cerebral deficit, coma, convulsion, subarachnoid haemorrhage and chorea in 2 to 5% of patients.1 Till now few cases have been reported.

We are reporting a case of 12 years boy with HSP who had a large intracerebral haematoma in the right occipital lobe.

The case is reported here because of the rarity of this sort of complication of HSP and scope for treatment.

CASE REPORT

A 12-year-old boy presented with colicky pain in abdomen, nausea, symmetrical reddish papular skin lesions over the lower limbs and buttocks, severe arthralgia of knee and ankle joints, irritability, headache, generalized tonic-clonic seizure. There was no history of visual disturbances, rectal or gum bleeding, haematuria, food or drug allergy, insect bite or immunization. Family, personal and drug history are non-contributory. He had a history of upper respiratory tract infection with sore throat one week prior to the onset of illness.

Clinical examination revealed that the patient was conscious, febrile and irritable. He had mild pallor, pulse 104/minute, BP - 110/70mm of Hg, palpable purpura on both the buttocks and extensor surface of legs, tender knee and ankle joints without swelling. There was no focal neurodeficit including visual field loss. Examination of other systems was normal.

Investigations showed RBC morphology of microcytic hypochromic type with a hemoglobin label of 10.1 gm.%, TLC - 8500/cu mm, DC - N65L30M01E04, ESR -110 mm in 1st hour, and platelet - 1,50,000/cumm. Throat swab culture showed growth of Streptococcus β hemolyticus. The ASO titre was 1200 IU/ml. The coagulation profile and clot retraction time were normal. The estimation of factor XIII was not possible due to practical difficulties. Blood biochemistry and collagen vascular profile were normal. EEG revealed inter-ictal discharges of generalized seizure disorder without focal abnormality. The cranial CT scan showed moderately large hyperdense lesion at the right occipital area with surrounding edema (Fig. 1). The MRI of brain revealed subacute haematoma and subsequent MR-angiogram of brain showed no arteriovenous malformation or aneurysm. Skin biopsy showed small vessel leucocytoclastic neutrophilic vasculitis (Fig. 2).

Treatment with steroid therapy and anticonvulsant was successful and two year follow up was uneventful.

DISCUSSION

In our patient, classical features of symmetrical palpable purpura, colicky abdominal pain, arthralgia with preceding history of streptococcal sore throat, and histopathological picture of leucocytoclastic vasculitis are suggestive of HSP.

Neurological complication in HSP is often underestimated and intracerebral haemorrhage is quite rare. The exact mechanism of haemorrhage is not obvious, but probably may be due to CNS vasculitis.2,3 Scattarella et al described Henoch-Schonlein purpura in a seven year old boy with right occipital haematoma where symptoms of meningeal irritation, convulsion, hemiparesis and chorea were incriminated due
to CNS vasculitis. Altinos et al reported a boy with sudden onset large intracerebral haematoma causing rapid neurological deterioration. The haematoma resulted from intraparenchymal bleeding due to Henoch-Schonlein purpura and the boy made excellent improvement following urgent surgical evacuation of it. Henoch-Schonlein purpura with intracerebral haemorrhage has been reported by Imai et al where intracerebral haemorrhage was found to have resulted from a marked decrease in factor XIII.

Though we failed to arrange the estimation of factor XIII because of practical difficulties, normal coagulation profile exclude common haematological disorders. Neuroimaging ruled out vascular malformation and neoplasia. Skin biopsy confirmed small vessel leucocytoclastic neutrophillic vasculitis compatible with HSP.

Hence, we feel that this haematoma is primarily related to necrotizing vasculitis leading to intracerebral haemorrhage. Though vasculitis classically gives rise to haemorrhagic infarct in neuroimaging study, several confluent foci of haemorrhagic infarction in neuroimaging may look similar to primary intracerebral haematoma. Despite moderate haematoma, patient did not notice any focal neurodeficit, possibly due to slow tempo of development of haematoma.

Prognosis of this condition in all the documented cases are good and haematoma with other symptoms resolved with passage of time as is in our case. Though Henoch-Schonlein purpura is a self-limited disease, we had to use steroid for intractable headache, as it was unresponsive to oral glycerine and acetazolamide.

REFERENCES