Unusual Complications of a Common Pediatric Illness : Henoch Schonlein purpura (HSP) with neurological and protracted gastrointestinal complications – A Case Report

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ABSTRACT

Henoch schnolein purpura is the multisystemic vasculitis disorder of childhood. Its common features include palpable purpura concentrated in dependent areas, arthralgia or arthritis, abdominal pain and glomerulonephritis. We report our experience with a boy with seizure and massive gastrointestinal bleed secondary to HSP which followed an unusually protracted clinical course and which was successfully treated with high dose pulse methylprednisolone. Renal involvement and the relapse of the disease was conspicuous by the absence.

Key words: Henoch schnolein purpura (HSP), Gastrointestinal bleed, Seizure, Methyl Prednisolone

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INTRODUCTION

Known to mankind for the last two centuries, Henoch Schonlein Purpura (HSP is the most common form of small vessel vasculitis of childhood, but can occur at any age from infancy to the ninth decade. The disease manifestations are protean ranging from a benign self-limiting disease to fatal multi-system complications. Neurological complications are rare, seen in 1- 8% of patient and though abdominal involvement common, massive GI bleed is rare. We present the case here in view rarity of presentation and scope for treatment.

CASE REPORT

A 7-year-old, previously healthy boy presented with mild fever for two weeks, abdominal pain, hematemesis for 3 days, skin rash of one day duration and one an episode of generalized tonic - clonic seizure. On examination his vital signs were normal. Skin showed scattered palpable purpura over both the extremities, and ears. His sensorium was normal, no focal deficits and systemic examination was normal except for mild epigastric tenderness.

On admission, results of the full blood count, coagulation profile, chemistry tests and urine analysis were normal with no hematuria. Based on this constellation of findings, a diagnosis of HSP was suspected. Histopathology of the tissue obtained from skin biopsy of the lesions was compatible with leukocytoplastic vasculitis with deposition of IgA and C3 in dermal capillaries, confirming the diagnosis of HSP. Neuroimaging showed a small, focal wedge shaped hypodensity in the left parieto - occipital region suggestive of a small infarct. Supportive therapy was instituted he continued to have severe abdominal pain. Parenteral Methylprednisolone at a standard dose, resulted in relief of the abdominal pain and purpura over next 7 days. The treatment was switched over to enteral steroid therapy.

As the steroids were being tapered off, he had massive lower gastrointestinal hemorrhage requiring blood transfusion, with severe crampy abdominal pain. A CT scan and sonography of abdomen ruled out surgical etiology and showed thickened bowel loop. The clinical course was complicated by continued intermittent bouts of lower GI bleed raising a pertinent question: was this a steroid induced bleed or worsening HSP ? A lower GI endoscopy showed multiple large hyperemic areas with central ulcers and necrosis from the rectum to the ascending colon (distal more than proximal) and a large, friable polypoidal ulcerated area at the ileocecal junction suggestive of complicated HSP. A biopsy and histopathological examination revealed nonspecific inflammatory changes. High Dose Pulse Intravenous Methylprednisolone therapy was commenced. He improved with complete resolution of abdominal pain and bleeding. Over the next two weeks, steroids were gradually tapered off and discontinued. He was discharged home after 4 weeks of hospitalization. Follow - up examination 3 months later showed no sequelae.Disese relapse was not observed in the interim period.

DISCUSSION

HSP is a systemic vasculitis with multiorgan involvement. Classical features as symmetrical non-thrombocytopenic palpable purpura, colicky abdominal pain, leucytoclastic vasculitis with IgA and C3 deposits in dermal biopsy were observed in our case confirming the diagnosis of HSP.[1]

HSP is a self-limiting condition, usually resolving within 6 to 8 weeks, but complications may arise. Renal involvement occurs in 30-40% of cases; and 50- 75% of children experience gastrointestinal symptoms. Additional symptoms include fever, scrotal pain, and edema in boys, and rarely pulmonary, cardiac, or neurologic manifestations. [2,3] Although the long-term prognosis of HSP is almost entirely attributable to the kidney disease [4,5], some rare extrarenal features may produce substantial morbidity and mortality. In our case, unusual presentation of HSP was observed with both neurological and severe GI manifestations without renal involvement.

GI manifestations preceding skin purpura is less common (15-30%) which was observed in our case. This is significant as they may simulate a surgical emergency and can even result in an unnecessary laparotomy. We also report massive GI hemorrhage which is an unusual complication. Common GI presentations include nausea, vomiting, acute abdominal pain (65%), GI bleed (35%) and intussusception (5%). These symptoms are caused by submucosal haemorrhage and oedema of the bowel wall, predominantly affecting the proximal small bowel[6]. Other significant, though less common gastrointestinal complications are gangrene of the bowel and bowel perforation. The risk factors for GI involvement were not observed in our case, as the neutrophil to lymphocyte ratio was normal, CRP and procalcitonin was normal, factor XIII assay couldn't be done but a normal coagulation profile rules out any factor deficiency, though that would have been important pointer towards severity. In this case GI involvement was more severe with no involvement of kidneys. In such condition, it can be confused with other conditions like ulcerative colitis, crohn's disease, etc.

Here we also observed neurological involvement in the form of seizure without any renal involvement or hypertension which is an uncommon feature[7] . The CNS involvement has been reported in 1–8% of children. Possible neurological presentations in HSP include headache, altered level of consciousness, seizures, focal neurological deficits, visual abnormalities and verbal disability, peripheral neuropathy, and facial palsy[8,9,10]. CNS dysfunction in HSP results from a vascular obstruction, from an intracerebral haemorrhage or from severe hypertension [5]. Imaging studies might reveal lesions suggestive of small vessel vasculitis as ischemic vascular lesions, intracerebral haemorrhages, diff use brain edema, or thrombosis of the superior sagittal sinus. Posterior reversible encephalopathy syndrome has been described in children with HSP. In patients with suspected CNS or peripheral nervous system Henoch–Schonlein syndrome, it is imperative to rule out other vasculitides syndromes, with detection of IgA in tissue-like skin or kidney [18]

Though mainstay of treatment for HSP is symptomatic and might include mild analgesics such as acetaminophen and nonsteroidal anti-inflammatory drugs for joint pain and fever [2,4] nonsteroidal anti-inflammatory drugs should be avoided in the presence of gastrointestinal or renal manifestations, as they have been shown to aggravate these symptoms.[2,4] Steroid therapy is reserved for HSP with gastrointestinal and renal complications. Corticosteroid dose, 1-2mg/kg/day either oral or parenteral has shown to decrease the mean duration of abdominal pain[5], though exact dosage guidelines are lacking. We found the massive GI hemorrhage and pain abdomen was difficult to control and the standard dose of corticosteroid was effective initially but while tapering, the symptoms reappeared which responded to high dose pulse methylprednisolone. Studies have not shown clear benefit of either high dose steroids over low dose or parenteral over oral route as the initial treatment.[5] Severe gastrointestinal involvement like massive bleeding, mesenteric vasculitis if steroid non-responsive, other therapies have proven to be beneficial. High dose pulse methylprednisolone, IvIg or immunosuppressive medications like cyclophosphamide or azathioprine have been reported to result in complete resolution of abdominal symptoms

along with purpura and arthritis.[11-14] Though IvIg was found to be associated with increased proteinuria [15,16.] It is also important to carefully follow up these patients as severe GI involvement and purpura lasting for more than a month are risk factors for renal involvement [17.]

The initial management of patients with suspected cerebral HSP includes control of arterial hypertension, seizures, and repair of disordered hemostasis. Combined therapy with corticoids and /or cyclophosphamide is appropriate in a patient with relevant ischemic cerebral lesions. Peripheral or cranial neuropathy usually tends to recover spontaneously whereas Guillain Barre Syndrome needs to be treated with intravenous immunoglobulin or plasma exchange.[18]

CONCLUSION

Although HSP is a mild self-limiting disease and most cases resolve with supportive care, severe complications can develop. Intensive care therapy and high doses of steroids result in complete resolution of the disease. Evidence is emerging that steroids in the early phase of the disease will prevent gastrointestinal and neurological complications.

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