Case Report

Acute Pancreatitis in Henoch-Schonlein Purpura Complicated by Large Pseudocyst Formation Requiring Cystogastrotomy

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ABSTRACT

Henoch-Schönlein purpura (HSP) is a small-vessel vasculitis that typically occurs in children and causes characteristic purpuric skin lesions occasionally associated with multisystemic involvement. Gastrointestinal symptoms appear in 70% of the patients varying from mild to severe abdominal pain resembling an acute abdomen but HSP rarely presents with acute pancreatitis. Here, we report the case of an 11-year-old girl admitted with HSP-associated recurrent episodes of acute pancreatitis complicated with large pseudocyst formation. She was treated with pulse methylprednisolone followed by oral prednisolone and a single cyclophosphamide pulse but finally required a cystogastrotomy.

Keywords: Acute pancreatitis, Cyclophosphamide, Henoch-Schönlein Purpura

INTRODUCTION

Henoch-Schönlein purpura (HSP), a systemic small-vessel vasculitis syndrome, typically occurs in children and causes characteristic purpuric skin lesions occasionally associated with multisystemic involvement. Gastrointestinal symptoms appear in 70% of the patients varying from mild to severe abdominal pain resembling an acute abdomen [1]. Acute pancreatitis (AP) is rarely reported; the majority being mild but occasional complications do occur in the form of hemorrhage, necrosis, and pseudocyst formation, which are amenable to conservative therapy [2]. However, in children, a large symptomatic pseudocyst as a complication of HSP requiring cystogastrotomy has not been reported.

CASE REPORT

An 11-year-old girl was admitted with a 3-day history of palpable purpura involving all four limbs associated with pain abdomen and three episodes of non-bilious vomiting. The pain was localized to the epigastric region and non-radiating in nature. There was no history of chronic illness, drug use, allergy, or trauma. On examination, epigastric tenderness was present with stable vital signs. She was normotensive without any gross hematuria.

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On investigation, blood counts, C-reactive protein (CRP), urinalysis, and liver function tests were normal but serum amylase and lipase were mildly elevated (350 IU/L and 410 IU/L respectively). Ultrasound (USG) abdomen showed hepatomegaly without any features of pancreatitis.

She was diagnosed as HSP pancreatitis and because of persistent abdominal pain was initiated on oral prednisolone and discharged. After 6 days of discharge, while on oral prednisolone, she was readmitted with severe abdominal pain and recurrent multiple episodes of vomiting. This time, the pain was severe and generalized as the child presented with an acute abdomen.

On examination, the child looked toxic, had tachycardia (heart rate 150 beats/minute) with decreased urine output. The blood pressure was in the normal range. Blood counts showed leukocytosis 22,700/cmm with 81% neutrophils, raised CRP (51.2 mg/L, normal <5), elevated serum amylase (2866 IU/L, normal 0–100), and lipase (2970 IU/L, normal 0–60). Contrast-enhanced computed tomography (CECT) of the abdomen revealed acute pancreatitis with a peripancreatic collection without necrosis.

Considering the increased clinical severity, she received three pulses of iv methylprednisolone 6 mg/kg/day but as pain abdomen recurred on restarting oral prednisolone, one dose of iv cyclophosphamide 500 mg/m^2 was given. Abdominal pain subsided and she was discharged on oral prednisolone.

After 5 days, she was readmitted with a recurrence of generalized abdominal pain, vomiting, and abdominal distension.

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The child looked pale and tachycardic with cold peripheries. There was leukocytosis 12,300/mm^3 with 91% neutrophils, elevated CRP (73.8 mg/L), falling hemoglobin (7.2 mg/dl), and elevated serum amylase 3156 IU/L as well as lipase 2580 IU/L. Magnetic resonance imaging (MRI) abdomen with magnetic resonance cholangiopancreatography (MRCP) revealed an intrapancreatic fluid collection measuring 8 cm × 4 cm and marked ascites (Fig. 1). Paracentesis was done; peritoneal fluid amylase and lipase were 3053 IU/L and 3160 IU/L, respectively.

She was managed conservatively and prednisolone was continued. In view of persistent abdominal distension and vomiting following feeds, USG-guided drainage of pseudocyst was done twice and 1.5 L of fluid was removed on each occasion but because of rapid reaccumulation causing discomfort, cystogastrotomy was done finally. She remains asymptomatic on follow-up.

DISCUSSION

Henoch-Schönlein purpura is a systemic vasculitis characterized by the deposition of IgA-containing complexes and complement (C3) on arterioles, capillaries, and venules. Clinical manifestations include skin purpura, abdominal pain, arthritis, and occasional glomerulonephritis. AP as a presentation of HSP is very rare [2,3].

The pathophysiologic mechanism of AP is thought to be vasculitic involvement of small vessels within the pancreas, leading to increased vascular permeability and pancreatic edema [3,4]. Endothelial damage-induced embolism causes pancreatic ischemia and hypoxia and the abnormal activation of digestive enzymes leading to inflammation, edema, vascular injury, and cellular death [5].

The biggest case series by Qin Zhang *et al*. showed only 13 among 3212 HSP patients over a period of 13 years in whom AP was the presenting complaint [6]. Xiong *et al*. reported a review of previously published case reports of 13 patients with HSP pancreatitis wherein only one child of 16 years age had pseudocyst formation which was treated with steroids [7]. Formation of pseudocyst in HSP pancreatitis is extremely rare as the majority of them are of the mild variety.

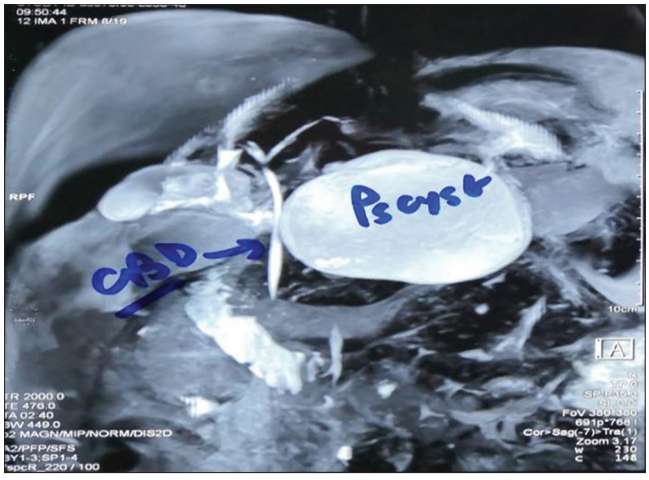

Figure 1: Magnetic resonance cholangiopancreatography of abdomen showing intrapancreatic fluid collection

Early diagnosis can be done by estimation of amylase and lipase in all patients of HSP admitted with pain abdomen. USG or CECT is needed in severe pancreatitis to look for necrosis, peripancreatic collection, and pseudocyst formation. In our case, the child was admitted 3 times, and serial CECT and MRI + MRCP helped to promptly diagnose the complications.

It was previously believed that steroid administration in HSP-related pancreatitis can worsen the outcome with patients developing gastrointestinal bleeding and perforation [2,8]. However, currently, it is established that steroid therapy can actually alleviate the symptoms by decreasing cytokine release and pancreatic secretions [2,9]. In HSP with severe gastrointestinal vasculitis where symptoms persist even after steroid therapy, the use of a single dose of cyclophosphamide has resulted in complete resolution of symptoms [10,11]. Our patient responded to methylprednisolone but had a recurrence of pain on converting to oral prednisolone and needed a single dose of IV cyclophosphamide for complete alleviation of the symptoms.

In our patient, the third admission was due to the complications caused by the expanding pseudocyst causing pressure on the surrounding viscera. A case report published by Pimpa UA yielded similar results involving an adult patient [12]. Considering the rapid reaccumulation of intracystic fluid even after recurrent aspiration, cystogastrotomy was performed as a curative procedure.

CONCLUSION

Amylase and lipase should be done early in HSP patients presenting with abdominal pain. Although the majority of these cases are mild, severe forms do occur rarely. The mainstay of treatment consists of supportive management with early enteral feeding along with iv methylprednisolone in severe cases. When steroid therapy yields a suboptimal response, a single dose of iv cyclophosphamide can help in the complete resolution of symptoms. However, in spite of all conservative therapy, complications like large pseudocysts can rarely develop.

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