American Journal of Medical Case Reports, 2017, Vol. 5, No. 6, 144-147
Available online at http://pubs.sciepub.com/ajmcr/5/6/3

DOI:10.12691/ajmcr-5-6-3

Groove Pancreatitis as a Rare Presenting Feature of Pediatric Henoch-Schonlein Purpura – Case Report

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Abstract Acute pancreatitis (AP) has rarely been reported in association with pediatric Henoch-Schonlein purpura (HSP) and even very occasionally reported as a primary clinical feature presenting before the development of characteristic palpable purpura. We describe here a five years and six months old girl presented with acute pancreatitis in the form of focal groove pancreatitis. The characteristic rash of HSP developed later in the disease course. This rare entity is discussed to alert pediatricians to include acute pancreatitis in the differential diagnosis of abdominal pain complicating HSP.

Keywords: *Henoch-Schonlein purpura, pancreatitis, child*

Cite This Article: Mohammed Ali Tohary**,** Hassan Mohammed Gohal, Ali Abdullah Awaji, Ali Abdullah Kamli, and Hossain Ibrahim Ageel, "Groove Pancreatitis as a Rare Presenting Feature of Pediatric Henoch-Schonlein Purpura – Case Report." *American Journal of Medical Case Reports*, vol. 5, no. 6 (2017): 144-147. doi: 10.12691/ajmcr-5-6-3.

1. Introduction

Henoch-Schonlein Purpura (HSP) is the most frequent form of childhood vasculitis. It is a multisystem disease predominantly affecting the small vessels of skin, joints, gastrointestinal tract (GIT) and kidneys, and rarely, other organs such as central nervous system, lungs, eyes, and scrotum. Acute pancreatitis may rarely develop as a complication of HSP in children [1]. It is even more rare to develop before the appearance of the typical rash of HSP. There were only five cases of pediatric HSP reported worldwide in the English literature with acute pancreatitis as an initial presentation [2,3,4,5,6]. We report here a five years and six months old girl presented with acute pancreatitis as a complication of pediatric HSP before the development of characteristic purpuric rash. To the best of our knowledge, this is the first pediatric case reported in Saudi Arabia.

2. Case Report

A five years and six months old girl referred from a local hospital to the pediatric emergency department at King Fahd Central Hospital, Jazan with a history of an epigastric abdominal pain of five days duration associated with non-bilious vomiting. She had hematemesis and hematochezia one day before admission. No history of recurrent abdominal pain, chronic illness, medication, trauma, allergy or blood transfusion. The child received all vaccines including mumps. Family history was unremarkable.

The patient was irritable because of abdominal pain. She was not pale, or jaundiced. Her body temperature 36.6 °C, pulse 90 beats/min, blood pressure 125/81 mmHg, and SPO_2 99% in room air. Her weight and height at the 50th percentile for her age. No abnormal abdominal findings aside from epigastric tenderness.

On admission, the initial laboratory investigations showed normal results of complete blood count, erythrocyte sedimentation rate, coagulation profile, serum electrolytes, renal and liver function tests, bone profile, blood glucose, lipid profile, and urinalysis. C-reactive protein was 0.67 mg/dL (normal range: 0-0.5 mg/dL). Septic workup was negative. Serum amylase 125 IU/L (normal range: 25 – 115 IU/L), and serum lipase 430 IU/L (normal range: 114 – 286 IU/L) both figuring out slightly elevated levels at admission time. The repeated values of amylase and lipase were higher in two days later, 221 IU/L, and 1323 IU/L respectively. Her serum IgA level was normal (2.4 g/L). She was negative for viral hepatitis markers (A, B, and C), perinuclear antineutrophil cytoplasmic antibodies (pANCA), antinuclear antibody (ANA), and anti-double-stranded deoxyribonuclease (anti-dsDNA). Her serum complements (C3 and C4) were normal. Stool examination was positive for occult blood.

Computed Tomography (CT) of the abdomen showed circumferential mucosal duodenal wall enhancement and thickening with periluminal edema and abnormal fluid density at the groove triangle between the pancreas and second part of duodenum consistent with focal groove Pancreatitis with sentinel loop of duodenum [Figure 1(A, B)].

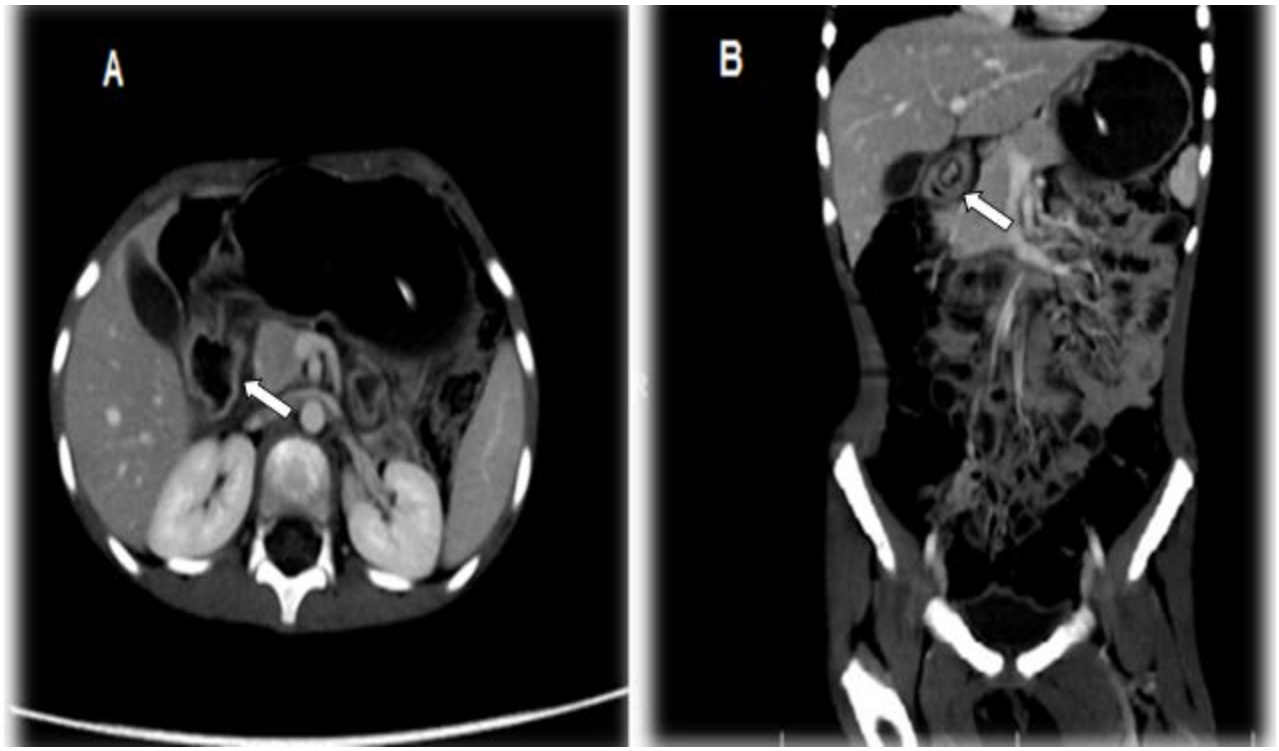

Figure 1. CT scan of the abdomen. (A) Cross section; (B) Longitudinal section, both showed Focal Groove Pancreatitis with sentinel loop of the duodenum

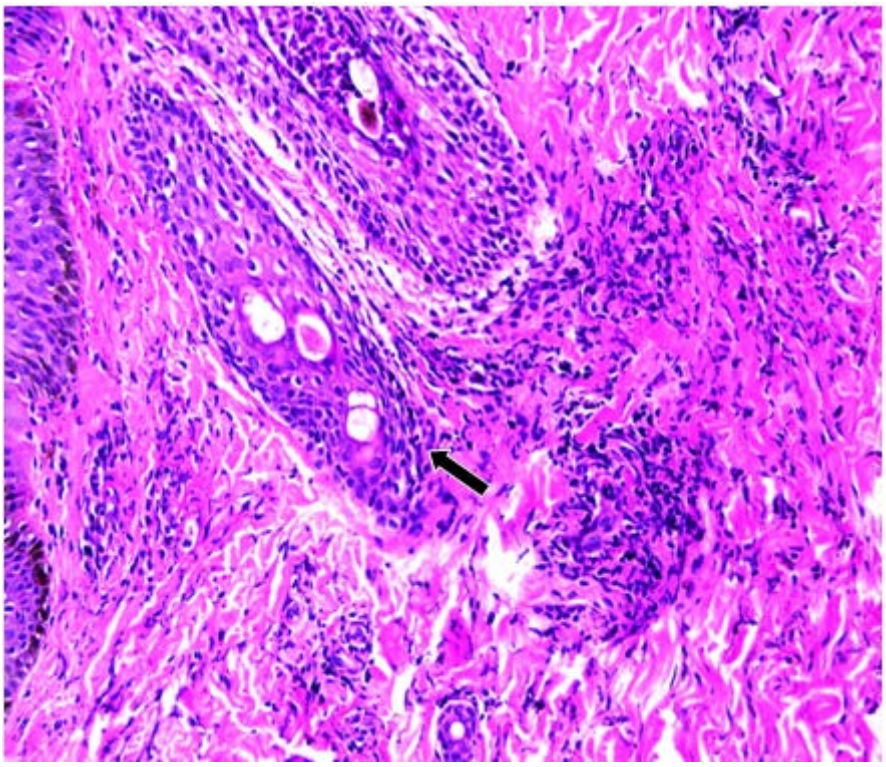

Figure 2. Skin Punch Biopsy of the purpuric rash revealed Leukocytoclastic vasculitis (LCV)

We managed the patient with complete bowel rest and started on intravenous (IV) fluid therapy, IV Ranitidine and analgesia. Her abdominal pain was worsening during hospitalization. On the fourth day of admission, she had developed a non-blanching rash in the buttock and then gradually progressed to involve the trunk, as well as both upper and lower limbs. Dermatologist performed skin biopsy from these lesions. Histopathological studies showed the classical leukocytoclastic vasculitis consistent with the diagnosis of HSP (Figure 2). Later in her disease course, she developed joint pain and swelling of both knees. We started the patient on oral prednisolone 1 mg/kg/day for seven days and then tapered slowly. The abdominal pain resolved, and the patient tolerated oral diet with no exacerbation of her symptoms. Skin rash started to fade, and arthritis improved significantly. She was discharged home in good condition. During follow-up, she remained free of symptoms and her S. lipase level was normal (146 IU/L).

3. Discussion

Acute pancreatitis (AP) is rare in children, but its incidence is increasing in the last two decades [7]. The underlying etiologies in children are often drugs, trauma, infections, congenital anomalies of the pancreaticobiliary system, metabolic disorders and systemic diseases including vasculitis.

HSP is immune-mediated vasculitis, and the deposition of immunoglobulin A (IgA)-containing immune complexes in the walls of small vessels characterizes its pathogenesis [8]. It is predominantly affecting the skin, gastrointestinal tract (GIT), Joints and kidneys. Palpable purpura, arthritis/arthralgia, abdominal pain, and nephritis are the classic tetrad of HSP [8]. More than one-half of children with HSP present with gastrointestinal symptoms and the most common presentation is colicky abdominal pain followed by vomiting [3]. Severe GI complications including massive GI bleeding, intussusception, bowel perforation, protein-losing enteropathy and acute pancreatitis were also reported [3,9].

The first reported case of acute pancreatitis complicating pediatric HSP was in 1977 of a seven-year-old girl presented with purpuric rash followed by signs and symptoms of acute pancreatitis [1]. Since the first report, there are only a few cases of acute pancreatitis described. The classic presentation of pancreatitis in association with HSP usually develops within one week after the characteristic palpable rash. Acute pancreatitis as an initial manifestation of pediatric HSP before the appearance of the rash is very rare with only five patients reported worldwide [2,3,4,5,6]. The first pediatric patient described was in a seven-year-old Chinese boy by Cheung et al. in 2001 [2]. Pediatric HSP reports from Saudi Arabia described gastrointestinal complication in 50-75% of the cases, but no acute pancreatitis identified [10,11,12,13,14]. Recently an 11-year-old Saudi girl was reported to have HSP with acute pancreatitis developed after the rash appearance [15]. Our case is considered the first patient among Saudi pediatric HSP patients to present with acute pancreatitis as an initial manifestation before the rash. The characteristic purpuric rash developed in this patient four days after the initial presentation of pancreatitis.

We established the diagnosis of acute pancreatitis in the presence of abdominal pain, high serum lipase, and radiographic evidence of acute pancreatitis. CT abdomen showed focal groove pancreatitis with sentinel loop sign. HSP was diagnosed based on the clinical features and the histopathological finding of the classical leukocytoclastic vasculitis. Groove pancreatitis is a rare type and indicates a focal chronic pancreatitis [20]. There are two forms of groove pancreatitis, the pure form which involves the groove only, while the segmental form extends up to the pancreatic head [21]. There is no a single report of groove pancreatitis in pediatric HSP. Acute pancreatitis with the presence of inflammatory changes in the groove may mimic the pure form of groove pancreatitis [21]. We think this may be the explanation for the finding of groove pancreatitis in this patient. Investigations showed negative results of hepatitis serological markers for A, B, and C, and auto-antibodies including ANA, anti-dsDNA, and ANCAs which make the possibility of acute pancreatitis due to other causes unlikely.

High serum IgA levels occur in 50% of patients with HSP [16]. 15% of HSP patients may have hypocomplementemia [17]. Interestingly, we noticed that most of the case reports of HSP complicated with acute pancreatitis reveal normal levels of serum IgA and complements [4,6]. We share this finding as well in our case.

The frequent presence of fecal occult blood and rarely massive gastrointestinal hemorrhage in pediatric HSP suggest mucosal lesions. Hemorrhagic erosive duodenitis and gastritis were the most common endoscopic findings [18]. The patient had hematemesis and hematochezia one day before admission. The stool was positive for occult blood. The patient was managed with an acid-suppressing agent and did not require a blood transfusion. We did not perform an upper gastrointestinal endoscopy due to the transient and self-limited nature of the bleeding. Later in her disease course, the child developed arthritis in both knees which is a frequent finding of HSP.

Studies showed that steroid reduces both the severity and duration of abdominal pain during the first two weeks of treatment [19]. HSP-related pancreatitis was treated in several case reports with steroid and showed a dramatic improvement of symptoms and prompt resolution of pancreatitis [4,6]. We share the same experience in this case. The patient did not respond to the initial standard treatment of acute pancreatitis, but she responded well to the steroid.

In conclusion, acute pancreatitis is a rare presentation of pediatric HSP, and we should consider in the differential diagnosis of abdominal pain. The steroid may be of value in the treatment of HSP-related pancreatitis.

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