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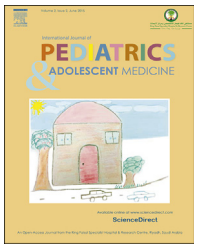

CASE REPORT

Successful treatment of severe gastrointestinal manifestations of Henoch—Schonlein Purpura and factor XIII deficiency using cryoprecipitate transfusion

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Abstract Henoch—Schonlein Purpura (HSP) might present with severe gastrointestinal (GI) involvement. Herein, we report 3 cases of HSP with severe GI manifestations in the form of hematemesis, melena, pancreatitis, and erosive gastritis. Different treatment modalities were not successful. Low factor XIII levels were found in all patients and Cryoprecipitate transfusion resulted in significant immediate clinical improvement.

1. Introduction

Henoch—Schönlein Purpura (HSP), a non-granulomatous, immunoglobulin A-mediated small vessels vasculitis. It is the most common vasculitis in pediatric age group with an incidence of 20 per 100,000 in children less than 17 years of age with a peak incidence of 70 per 100,000 in children between the ages of 4 and 6 years [1—5]. It presents with a tetrad of palpable purpura, arthritis or arthralgia,

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abdominal pain, and renal disease. Gastrointestinal (GI) manifestations may vary from solely colicky abdominal pain in about one-half of patients to GI bleeding in approximately 20—30 percent of patients [2,3]. However, rarely GI complications including acute pancreatitis and bowel perforation may occur [1]. A previous study by Prenzel have reported an association between severe HSP and factor XIII deficiency [5].

In this study, we report 3 cases of HSP with severe GI manifestations in the form of melena, hematemesis, acute pancreatitis, and erosive gastritis. All were unresponsive to corticosteroids. Low levels of factor XIII were found in all patients. Fortunately, they showed dramatic response to Cryoprecipitate transfusion.

2. Case 1

An 11-year-old girl, previously healthy, admitted with history of skin rash, abdominal pain, and vomiting for 12 days. The skin rash started at the lower extremities then progressed gradually to the buttocks area, upper extremities and face. The abdominal pain was periumbilical, colicky in nature, severe, and associated with vomiting. The vomiting had been associated with streaks of blood. She was admitted initially to a local hospital where she was treated conservatively, but she had worsening symptoms with significant abdominal distension and severe pain. She was referred to our hospital for further workup and management.

Physical examination upon arrival revealed ill-looking girl, in pain, conscious, oriented, and well hydrated. Her vitals were stable. She had purpuric rash over the face in a malar distribution (Fig. 1), upper and lower extremities with ulcerated lesions over both elbows and malleoli (Figs. 2—4). She had a distended abdomen with generalized tenderness, more in the epigastric and umbilical regions. Other examinations were unremarkable.

Laboratory data revealed: high white blood cell count at 22×10^9/L (normal 5—15) with predominant neutrophils (79%), low hemoglobin 10.4 mg/dl (normal 11—14), high platelet count 358×10^9/L (normal 140—350), normal erythrocyte sedimentation rate (ESR) at 2 mm/hr, high C-reactive protein (CRP) at 39.8 mg/L (normal $>$ 3), normal renal functions and electrolytes, high amylase at 275 U/L (normal range 3—110), high lipase at 584 IU/L (normal range 0—60), and normal complements levels. Antinuclear antibodies, anti-SSB, anti-SSA, prothrombin time (PT), partial thromboplastin time (PTT), international normalized ratio (INR), occult blood in stool and urinalysis were negative.

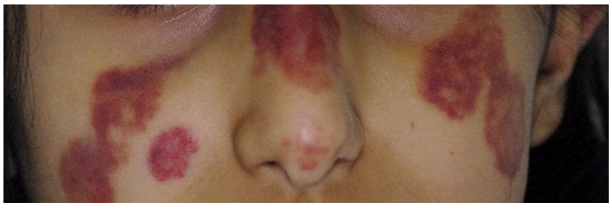

Figure 1 Purpuric rash in a malar distribution.

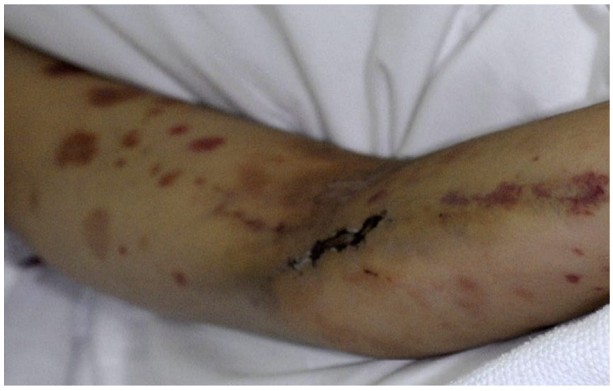

Figure 2 Purpuric rash over upper and lower extremities.

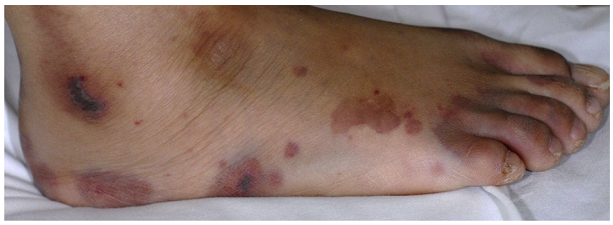

Figure 3 Purpuric rash over upper and lower extremities.

Abdominal radiograph showed evidence of paralytic ileus with multiple air-fluid levels suggestive of intestinal obstruction. Abdominal ultrasound reported moderate, free intraperitoneal fluid with inability to visualize the pancreas due to marked bowel gas shadowing. Abdominal computerized tomography (CT) showed diffuse swollen edematous pancreas, which confirmed the diagnosis of pancreatitis.

She was started on daily intravenous (IV) methylprednisolone pulse (30 mg/kg/dose) for 3 days without significant improvement. She was kept NPO (null per oral), on nasogastric tube (NGT) suctioning and analgesia. Skin biopsy showed leukocytoclastic vasculitis with IgA deposition confirming the diagnosis of HSP (Fig. 5). A week later, she developed melena; hence, upper endoscopy was performed and showed evidence of severe erosive gastritis with normal esophagus and duodenum. Therefore, NGT suctioning was continued and IV omeprazole and total parental nutrition (TPN) were started. IV methylprednisolone and IV immunoglobulin (IVIG) were administered, but it was without any significant improvement. Assay of Factor XIII showed low level at 0.3 IU/ml (normal range 0.7—1.2 IU/ml). Subsequently, Cryoprecipitate transfusion (60 ml equivalent to 3 units) was given 2 weeks after admission (on

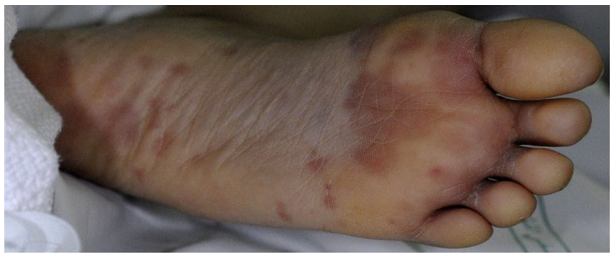

Figure 4 Purpuric rash over upper and lower extremities.