Case Report

Intestinal Infarction and Portal Vein Thrombosis in a Patient with Henoch Schonlein Purpura

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Henoch Schonlein purpura is a systemic vasculitis that commonly affects children and teenagers but also affects adults of all ages. In most instances it has a benign course. Organ involvement, particularly in adults, and notably the kidneys and gastrointestinal tract may require therapeutic intervention and may have a less favorable outcome. We report a case of a 58-year-old man who presented with purpura and who rapidly developed catastrophic intestinal vasculitis, leading to his demise.

1. Introduction

Henoch Schonlein purpura (HSP) is a systemic vasculitis characterized by involvement of the skin, joints, kidney, and gastrointestinal tract [1–3]. It is primarily a disease of children but can occur at any age [2, 3]. In most cases, especially in children, it is a benign self-limiting disorder, but adults may require immunosuppressive therapy for complete recovery [2]. Gastrointestinal (GI) manifestations occur in 55–75% of adult HSP patients [1]. The most common GI symptoms include colicky abdominal pain, GI bleeding, and vomiting. Rarely, intussusception, bowel infarction and hemorrhagic ascites can complicate HSP [4–7]. Complete recovery with or without treatment usually occurs, and fatal complications are rare. We report an adult patient with HSP complicated by extensive infarction of the intestine, intraperitoneal hemorrhage, and portal vein thrombosis with a fatal outcome.

2. Case Report

A 58-year-old white man was referred to the outpatient nephrology office by his gastroenterologist because of microscopic hematuria and proteinuria in the setting of an evolving purpuric rash. The rash had progressed proximally from both feet to his thighs, upper extremities and abdomen over the past two weeks. He complained of nausea, and vomiting for one day but denied hematemesis, melanotic stools, or arthralgia. His significant comorbidities included hypertension, untreated hepatitis C, and alcohol abuse. He took no medications.

On physical examination blood pressure was 105/62 mmHg, pulse was 84 beats/minute and regular, and temperature 37.7°C. The skin showed nontender, palpable purpuric lesions involving both upper and lower extremities and the abdominal wall. Lungs were clear to auscultation and the cardiovascular exam was unremarkable. On abdominal examination no organomegaly was appreciated. There was no pedal edema.

Laboratory tests revealed a white blood cell count of $13.1 \times 10^3/\text{mcL}$, hemoglobin 14.7 g/dL, hematocrit 42.9%, and platelet count of $234 \times 10^3/\text{mcL}$. Serum chemistry showed sodium 136 mEq/L, potassium 4.2 mEq/L, chloride 101 mEq/L, bicarbonate 24 mEq/L, blood urea nitrogen (BUN) 34 mg/dL and serum creatinine 2.0 mg/dL. Liver
Intravenous methylprednisolone 1 g daily for three days was administered and then 30 mg every 12 hours. Postope- ratively he continued to have an elevated white blood cell count and persistent fever, but his renal function improved. A repeat CT scan of the abdomen revealed persistent small bowel wall thickening. A second exploratory laparotomy was performed, but no additional ischemic bowel was identified. The patient was continued on broad-spectrum antibiotics and maintained on a ventilator due to respiratory failure. Eight days after the second laparotomy he developed severe lactic acidosis, right upper abdominal quadrant tenderness jaundice and worsening renal function. Ultrasound of the right upper quadrant showed findings compatible with portal vein thrombosis and cholelithiasis. Repeat CT scan of the abdomen showed increased ascites with intraperitoneal hemorrhage. Hemodialysis was initiated due to worsening oliguric acute kidney injury. His condition continued to deteriorate, and he died one day later, a total of 27 days after admission.

3. Discussion

HSP is a systemic small-vessel vasculitis that is mainly a disease of early childhood [1–3, 10]. Overall prognosis is good in both children and adults, with one study showing complete recovery occurring in 94% of children and 89% of adults [2]. In this regard, recovery is usually spontaneous in children, whereas in adults immunosuppressive therapy may be required in up to 63% of cases [2]. We present a case of a middle-aged man who developed extensive gangrenous bowel and intraperitoneal hemorrhage due to HSP vasculitis, with a fatal outcome.

GI symptoms are one of the commonest manifestations of HSP in adults, involving 55–75% of individuals [1]. In a retrospective analysis of 115 adults with the diagnosis of HSP, GI symptoms were reported in 90 patients (78.2%), abdominal pain being the most common (89%) followed by vomiting and GI bleeding [4]. Furthermore, 24% of patients had GI symptoms prior to the development of a cutaneous rash [4]. In another retrospective study involving 116 children and 46 adults, adults had a lower frequency of GI involvement (5%) at disease onset, but during the clinical course GI involvement was the same in both age groups (56.5% of adults versus 63.8% of children) [2]. In a retrospective analysis of 250 adults with HSP, GI involvement was observed in 48%. In 13 of these cases (11%) serious GI complication developed requiring transfusion or surgery or leading to death [3]. The small intestine is the most common site involved, terminal ileum (60%) and the second portion of the duodenum (53%). The rectum (80%) is the most frequently affected areas in the lower GI tract [4, 8]. Endoscopic findings include mucosal congestion, redness, petechiae, multiple ulcers, and hemorrhagic erosions [4, 8, 9]. Rarely, severe GI complication such as bowel infarction, perforation, fistula, intussusceptions (ileoileal), hemorrhagic ascites, and pancreatitis can occur [4–7, 10].

A number of inciting antigens have been implicated in the causation of HSP. These include bacterial and viral infections, vaccinations, drugs, malignancy, and autoimmune
phenomena. The pathogenic mechanism of organ involvement in HSP is thought to be due to the deposition of antigen antibody complexes in the small vessel walls. This leads to activation of the alternate complement pathway resulting in inflammation and vasculitis [9, 10]. IgA is the antibody class most often seen in the immune complex [10]. The presence of hepatitis C virus could have been a triggering factor in our patient as has been suggested by others [10].

To the best of our knowledge, the extensive small and large intestine infarction with gangrene that led to a fatal outcome in our patient is rarely seen in HSP. In this case, in contrast to the majority of those reported, the arteritis was extensive and involved medium sized vessels (0.3–0.5 cm) in many areas as well as the small arteries, arterioles, and venules. The involvement of the transverse colon and appendix as seen in our patient is also rare in HSP. An additional complicating factor in this case was the occurrence of portal vein thrombosis. In reviewing the literature, there is only one reported case of portal vein thrombosis complicating HSP [11]. However, in the patient presented here, we are unable to conclude whether the portal vein thrombosis was associated with underlying hepatitis C or was a complication of HSP. Hepatitis C, without cirrhosis would be unlikely to cause portal vein thrombosis.

The mainstay of therapy in HSP with severe organ involvement has been high-dose steroids. Other immunosuppressive agents such as cyclophosphamide, azathioprine, and mycophenolate mofetil have also been used [12, 13]. There has been only one prospective randomized trial comparing steroid therapy with and without cyclophosphamide in cases of severe visceral HSP [14]. The results of that study showed that addition of cyclophosphamide to steroids did not improve the outcome of the disease as compared to steroids alone. Our patient was treated with pulse steroids alone, with no significant improvement.

In conclusion, given the potential severity and fatal outcome of HSP involving the intestines in adults, prompt and early recognition of this entity is crucial. The mainstay of therapy remains high-dose steroids, with little evidence to support the use of other immunosuppressive agents. In catastrophic circumstances as described in this instance, even high-dose steroids may not alter the course of events.

References


