Henoch-Schönlein Purpura with Gastrointestinal Involvement

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Abstract

Henoch-Schönlein purpura (HSP) is a common vasculitis in childhood. It mainly affects small vessel of skin and other systems such as gastrointestinal tract, kidneys, joints etc. Gastrointestinal involvement occurs in 50%-75% of patients whereas gastrointestinal bleeding is rare. Symptoms of GI involvement begin usually about a week after the typically purpuric rash, however, this manifestation may appear before the rash. Our patient is a seven year old boy who presented with vomiting, nausea, abdominal pain and bloody stool. The 3rd day of his admission, purple-violet purpuras were seen on the tip of his left big toe and spread over ankles, legs and buttocks. The patient who responded to steroid therapy was discharged with diagnosis of HSP-gastrointestinal involvement on the 17th day of hospitalization. Conclusion: Purpuric skin lesions are the most common finding in HSP and generally are seen before gastrointestinal involvement. Initially HSP with GIS bleeding before rash may be treated as acute gastroenteritis/dysentery and that may misdiagnosis. Therefore, it is important to consider HSP with GIS involvement when a child presents with bloody stool/abdominal pain.

Key words: Henoch-Schönlein purpura, gastrointestinal bleeding, child

Introduction

Henoch-Schönlein Purpura (HSP), which was originally recognised in 1801 by Heberden and described as a vasculitis having an association with arthritis by Schonlein in 1837, is called HSP. It is acute small vessel leukocytoclastic vasculitis. It is the most common vasculitis seen in children, with an incidence of about 10 cases per 100,000 yearly [1,2]. HSP is a type III hypersensitivity reaction mediated small vessel vasculitis, therefore, it mainly affects the small vessels of skin, joints, gastrointestinal tract and kidneys. The disease is characterized by the triad of palpable purpura, abdominal pain and arthritis. Purpuric skin lesions are the most common findings in HSP and are generally observed before the presentation of the other involvements.
Gastrointestinal involvement (GI) is seen in 50%-80% of patients with HSP [3], who are characterized by mild symptoms such as vomiting, nausea, abdominal pain, and transient paralytic ileus. Also, gastrointestinal hemorrhage, bowel ischemia/necrosis, intussusception, bowel perforation can be seen but they are extremely rare. It is important to determine the GI, especially severe gastrointestinal bleeding, because it is a life-threatening complication. In 15 to 35 percent of patients, gastrointestinal features take precedence of the typical rash, therefore, the diagnosis of HSP is significantly more difficult [4-5]. Initially they may be treated as acute gastroenteritis and bloody stool with abdominal pain before skin manifestation [6].

In this report, we describe a case of a previously healthy child who had complaints of bloody diarrhea and vomiting, two days before being admitted to hospital, and who was later diagnosed with HSP when manifestation of skin occurred.

**Case Presentation**

A 7-year-old boy presented with a six-day history of fever (the highest was 39 degrees) and a two-day history of vomiting, nausea, abdominal pain and bloody stool which occurred four times a day. He was admitted to our clinic with these symptoms. Before admission, he did not have any complaints. There was no family history or past history of chronic disorders. On admission, he was very weak and suffered from moderate to severe abdominal pain. On clinical examination his body temperature was 36.5°C, heart rate was 110/min., respiratory rate was 24/min., and blood pressure was 121/73 mm/Hg. Clinical examination revealed no skin rashes and subcutaneous edema, but signs of abdominal tenderness and increased bowel sounds were noted without defense and rebounds. Routine blood counts and biochemical investigations were normal, and so was his hemoglobin level (14.1 g/dl) in spite of bloody stool. His urine analysis was normal and showed no protein. He was thought to have dysentery, therefore, the patient was started on fluids, and antibiotics (metronidazole and 3rd generation cephalosporin). His bloody stool/diarrhea and colicky abdominal pain had continued 4-5 times per day but tachycardia, hypotension, shock had not occurred. On the second hospital day, due to his colicky abdominal pain and to eliminate the acute abdomen, abdominal ultrasound was performed and it was non-specific. The stool examinations showed no leukocyte cell counts but too many erythrocyte cells, and the stool cultures were bacteriologically sterile. There were no parasites in the stool samples. On daily physical examination, we did not find any pathology but on the 3rd day of his admission we saw purple-violet purpuras on the tip of his left big toe. Routine laboratory tests were repeated but again there was no abnormality in his levels including hemoglobin (13 g/dl). All these combined with typical rash led us to think of HSP with GI involvement. The patient was administered intravenous (IV) corticosteroid (1 mg/kg methylprednisolone) for a short time and received supportive care. On the fourth hospital day, the purpuras had spread over ankles, legs and buttocks. The bloody stool finished on the sixth day and melena occurred. Two days after administering the steroid, abdominal symptoms resolved and melena was not seen again. Stool examination for occult blood was negative. Biopsy from the skin lesions showed features of leukocytoclastic vasculitis. IV steroid treatment was stopped on the fifth day and oral steroid treatment was started. On the 10th day of admission his rash had decreased and abdominal pain and GIS hemorrhage subsided completely but we observed swelling, pain and tenderness on his scrotum. With color Doppler ultrasonography, it was seen that the left testicle had become enlarged and there was epididymitis and no testical torsion. As a result he was diagnosed with acute scrotum/epididymitis. Scrotal involvement was treated with supportive care (analgesic) and scrotum was elevated. On the second day of epididymitis, his symptoms had disappeared and his scrotal examination was normal. The patient was discharged on the 17th day of hospitalization with complete clinical recovery and now he is followed up as an outpatient. All in all intravenous steroid followed by oral steroid therapy was given for one month.

**Discussion**

HSP, which is characterized by a triad of non-thrombocytopenic, non-traumatic palpable purpura, abdominal pain and arthritis, is the most common vasculitis in childhood between 4 and 11 years of age. It is known that the pathogenesis of HSP is related to the deposition of “immunoglobulin A (IgA)-containing immune complexes” and “complement components” within small vessel walls, however, the etiology is still unknown [1, 7]. Some studies have reported that HSP is to be
associated with infections, vaccines, foods, and some medications [8]. Clinical features in which gastrointestinal, renal and the other organ systems such as transient arthritis are usually self-limited, but renal involvement could result in chronic consequences for HSP patients [2]. Skin lesions are generally the first manifestation and typically occur in lower extremities and buttocks [9]. GI is seen in 50% to 70% of patients. Abdominal pain is the most common symptom of GI. Vomiting, anorexia, diarrhea can be seen as well. Due to the involvement of the gastrointestinal tract vessels, digestive complications such as hemorrhagic shock or GI obstruction-perforation, necrosis and bleeding can also occur [1]. HSP with gastrointestinal bleeding occurs in 18% to 52% of patients, the frequency of massive gastrointestinal bleeding is about 2% [10, 11]. Although massive gastrointestinal bleeding is rare, it is important to detect it rapidly because it may result in mortality and morbidity. Symptoms of GI involvement begin usually about a week after the typically purpuric rash, however, this manifestation may appear before the rash so diagnosis of HSP is significantly more difficult in 10% to 15% of patients [4,12]. Initially the patients who presented with GIS bleeding before rash may be treated as acute gastroenteritis/dysentery. Several studies have reported that massive GIS bleeding with HSP is rare, but a recent surveillance study conducted in Japan suggested that the hemorrhagic shock with GIS bleeding-HSP occurred in 1 of 261 cases (0.3%) [3]. (Chen et al). reported that 77.8% of patients showed GI symptoms (162 of 208 cases) and the most common symptom was abdominal pain. They also determined that gastrointestinal symptoms occurred before the manifestation of skin lesions in 41 patients (25.3%) and 35 patients had GI bleeding [1]. (David et al). evaluated the clinical findings of 183 patients with HSP. Ninety-five of 183 (52%) patients presented with GIS bleeding, and about 40% of the patients had bleeding before the typical lesions [13]. Our patient presented with GIS symptoms and bleeding before skin manifestation. He had had abdominal pain and bleeding before the purpuric rash and we detected the rash during his detailed daily physical examinations.

In laboratory measurements for the diagnosis of GIS involvement, blood in stool is seen in affected patients during vasculitis. Ultrasonography (USG) of the abdomen is a sensitive method to find out the intramural bleeding and edema in HSP [14]. Additionally, the USG can detect intussusception and other urgent conditions such as acute appendicitis, pancreatitis etc. at an early stage [15]. Our patient’s ultrasonographic findings were nonspecific and he had no complications requiring surgical intervention.

HSP generally has a self-limiting course; GIS involvement disappears spontaneously in several days in spite of GIS bleeding. Although treatment with corticosteroid is controversial for GIS involvement, it may be helpful for the resolution of pain, may play a role in the subsiding of other symptoms, and may prevent complications of GI tractus in the case of its early administration. [16,17,18]. We administered intravenous (IV) corticosteroid to our patient for a short time, and abdominal symptoms/bleeding resolved in a few days.

**Conclusion**

HSP has good prognosis without serious complications. GIS involvement can occur before the manifestation of skin, and that may cause misdiagnosis. Therefore, it is important to consider HSP with GIS involvement when a child presents with bloody stool/abdominal pain, so the skins of such children must be thoroughly examined during each patient round.

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