Acute pancreatitis as a presenting feature of Henoch-Schönlein purpura

Gönül Dinler¹, Kenan Bek², Yonca Açıkgöz², Ayhan Gazi Kalaycı¹
Departments of ¹Pediatric Gastroenterology, Hepatology and Nutrition and ²Pediatric Nephrology, Ondokuz Mayıs University Faculty of Medicine, Samsun, Turkey


Henoch-Schönlein purpura (HSP) is the most common small vessel vasculitis of childhood. It is characterized by nonthrombocytopenic palpable purpura, abdominal pain, arthritis, and glomerulonephritis. Although HSP is typically known to be self-limited, serious complications can occur. Acute pancreatitis rarely presents as a complication of HSP. It is even rarer as an initial presenting feature of HSP, before the occurrence of characteristic palpable purpura. Herein, we report a 12-year-old girl with HSP who atypically presented with acute pancreatitis.

Key words: child, complication, Henoch-Schönlein purpura, pancreatitis.

Henoch-Schönlein purpura (HSP) is a systemic vasculitic disorder commonly affecting young children. Gastrointestinal manifestations of HSP are common and vary from mild abdominal pain to intussusception¹,². Acute pancreatitis is a potentially life-threatening inflammatory disorder and it may rarely present as a complication during the course of HSP. It tends to develop in the first week of the illness following characteristic palpable purpura but may develop later³,⁴. However, acute pancreatitis, as an initial presenting feature before the typical rash during the course of HSP, is even more exceptional. To our knowledge, there are only two previously reported pediatric cases of HSP initially presenting with pancreatitis⁵,⁶. We herein report a 12-year-old girl with HSP atypically presented with acute pancreatitis.

Case Report
A 12-year-old girl presented with vomiting and epigastric pain of one-week duration. There was no history of trauma, medication, chronic illness, or recurrent abdominal pain. The patient’s family history was unremarkable, and there was no consanguinity between parents. She had a 10-year-old healthy brother. On admission, she appeared in great discomfort because of colicky, intermittent abdominal pain and nausea. Her blood pressure was 110/70 mmHg, body temperature 36.5°C, and pulse regular at 98 beats/min. Her height and weight were at the 50th percentile for age. She had no jaundice, and abdominal examination revealed tenderness in all quadrants but no rigidity or rebound tenderness. There was no hepatosplenomegaly, and bowel sounds were normal.

On admission, initial laboratory investigations revealed normal results of complete blood count, renal and liver function tests and urinalysis. Stool examination for occult blood was negative several times. Acute phase reactants, erythrocyte sedimentation rate, C-reactive protein, and fibrinogen were significantly high, at 82 mm/h, 161 mg/L and 929 mg/dl, respectively. Molecular analysis for familial Mediterranean fever (FMF) revealed heterozygous mutation of M694V. Serum amylase (119 IU/L, normal range: 28-100), lipase (200 IU/L, normal range: 13–60) and pancreatic amylase (113 IU/L, normal range: 13–53) were slightly elevated on admission and increased in a few days (amylase: 349 IU/L, lipase: 404 IU/L and pancreatic amylase: 236 IU/L). Alanine aminotransferase (ALT) and aspartate aminotransferase (AST) levels were also slightly elevated (163 IU/L and 82 IU/L, respectively). Ultrasound imaging
detected an anechoic cyst in the left ovary and minimal fluid accumulation in the bony pelvis, which were not specific for pancreatitis. Acute pancreatitis was defined with laboratory assays of amylase and lipase. Oral feeding was stopped and parenteral feeding started. The etiologic investigations of acute pancreatitis in terms of mumps and hepatitis viral markers were negative. Serum IgA and C3 and C4 levels were all normal.

On the 12th day of hospitalization, she developed palpable purpuric lesions on her lower extremities. HSP was diagnosed and it was proved definitively by skin biopsy, which was compatible with leukocytoclastic vasculitis. When the purpuric lesions developed, oral steroid therapy was started (2 mg/kg per day, tapered gradually after 5 days). Abdominal pain resolved within two days, amylase and lipase levels decreased to almost normal, and the patient was discharged on the 20th day. She remains in good general condition without any abdominal complaints nine months after the discharge.

Discussion

Acute pancreatitis is rarely seen in children but appears to be increasing. It is defined clinically by severe abdominal pain and vomiting with a rise in pancreatic enzymes at least three times the upper limits in blood and urine. The underlying etiologies are variable but the vast majority of cases are associated with trauma, medication, biliary tract disease, viral infections, and systemic diseases including vasculitis7,8. Henoch-Schönlein purpura is a systemic vasculitis characterized by deposition of IgA-containing immune complexes in the skin, joints, gastrointestinal mucosa, and glomeruli. It is the most common small vessel vasculitis of childhood. It usually presents with a classical tetrad of rash, polyarthritis, abdominal pain, and renal disease without any diagnostic difficulty in its typical presentation1,2. The abdominal pain, occurring in more than half of the patients, is characteristically colicky and localized to the periumbilical and epigastric regions, sometimes associated with nausea, vomiting or bleeding. The abdominal pain is thought to be due to localized edema and hemorrhage into the bowel wall secondary to the vasculitis of the small vessels9,10. Although the symptoms are similar, many unusual complications of HSP have been described, such as intussusception, bowel perforation and pancreatitis2,10. The pathophysiologic mechanism of acute pancreatitis is thought to be the vasculitic involvement of the small vessels within the pancreas3. Generally, in the reported cases, acute pancreatitis develops after the occurrence of the characteristic rash3,4,11. This rare complication was first described by Toskin12 in a 20-year-old man, and since then it has been documented in a few reports, including five children. The first pediatric case was reported in 1977 by Garner4. That seven-year-old girl was presented with painful and swollen feet, bruising and purpuric rash around the ankles, and she then developed signs and symptoms of acute pancreatitis within several days. The first pediatric case who presented with acute pancreatitis before rash was described by Cheung et al.5 in 2001. They reported a seven-year-old Chinese boy who presented with severe abdominal pain and was diagnosed as acute pancreatitis with markedly elevated serum amylase level and swollen pancreas on computerized tomography (CT). He developed palpable purpuric rash over his lower extremities on the 11th day. A second report of a pediatric case developing pancreatitis as the initial manifestation of HSP was recently reported in a three-year-old girl by Soyer et al.6 She was admitted with colicky abdominal pain and her laboratory tests were compatible with acute pancreatitis. She was diagnosed as HSP on the 5th day with the occurrence of purpuric rash on the lower extremities.

We report the third case of pancreatitis as an initial manifestation of HSP in childhood. Similar with the other two cases, our patient presented with severe abdominal pain as the initial complaint. Although pancreatitis was not confirmed radiologically by ultrasonography or CT, serum amylase and lipase levels gradually elevated until the occurrence of the rash. Two days after the initiation of steroid therapy, the abdominal pain resolved and serum amylase level began decreasing. Although steroids are not used in acute pancreatitis, they are used in HSP in certain conditions such as severe gastrointestinal, renal, central nervous system, and testicular involvement1,2,13. In HSP-associated pancreatitis, because of the
vasculitic involvement of the small vessels within the pancreas, steroids were used with prompt resolution\textsuperscript{5,6}.

In the reported cases, most of the HSP pancreatitis was mild in nature. There were two cases reported with hemorrhagic pancreatitis of HSP, both in adult ages\textsuperscript{12,14}. Only Cheung et al.\textsuperscript{5} reported a seven-year-old boy who developed a small pseudocyst that resolved spontaneously without any complication. In the present case, although amylase level increased up to threefold the upper limit of normal, there was no radiographic sign of pancreatitis and it resolved completely without any complication. It was reported that, in patients with mild acute pancreatitis, ultrasonography and also CT findings could be normal\textsuperscript{15}. Thus, in patients like the presented case, serum levels of amylase and lipase might be more important and should be measured in the recognition of pancreatitis. Furthermore, in atypically presented vasculitis patients like ours, especially in our geographic region, FMF should always be considered due to the relatively frequent association of these two conditions\textsuperscript{16,17}. However, our patient’s unremarkable family history, absence of another attack and heterozygosity of the M694V mutation were not found to be compatible with FMF.

In conclusion, even if rare, atypical presentation of HSP initially with acute pancreatitis should be remembered in order to plan the specific treatment and avoid unnecessary surgical interventions.

REFERENCES