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CASE REPORT

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Acute pancreatitis as an atypical presentation of Henoch-Schonlein purpura and cystic fibrosis

Abstract: Acute pancreatitis (AP) is a rare clinical entity in childhood. Two children, aged 10 and 14 years, were hospitalized for acute abdominal pain. Clinical and laboratory findings were compatible with AP. Both patients were given supportive medications. Two days after admission, the first patient had purpuric lesions on both buttocks and lower extremities, and occult bleeding. The clinical picture was consistent with Henoch-Schonlein purpura, with pancreatitis and gastrointestinal involvement. The patient's clinical condition improved rapidly after administration of prednisolone. The second patient had digital clubbing on physical examination, and positive sweat chloride test results led to the diagnosis of cystic fibrosis. AP in association with Henoch-Schonlein purpura and the initial manifestations of cystic fibrosis is very rare, and the presentation of these conditions is discussed.

Key words: acute pancreatitis, cystic fibrosis, Henoch-Schonlein purpura

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Henoch-Schonlein purpurasi ve kistik fibrozisin akut pankreatit kliniğinde atipik prezentasyonu

Özet: Çocukluk çağında akut pankreatit (AP) çok nadirdir. 10 ve 14 yaşında iki olgu ani başlangıçlı karın ağrısı sebebiyle yatırıldı. İki olguya da yapılan klinik ve laboratuvar incelemeleri sonucunda AP tanısı kondu. İlk olgunun başvurudan iki gün sonra kalça ve alt ekstremitelerde purpurik döküntüleri gelişti. Dışkı incelemesinde gizli kanı pozitifti. Hasta bu klinik bulguları ile Henoch-Schonlein purpurasının gastrointestinal ve pankreas tutulumu olarak kabul edildi ve hastanın prednizolon tedavisi başladıktan sonra tüm bulgularının düzeldiği gözlendi. İkinci olgunun fizik muayenesinde çomak parmağı mevcuttu ve yapılan ter testi sonucunda kistik fibrozis tanısı kondu. AP'in Henoch-Schonlein purpurası ile birlikteliği ve kistik fibrozisin ilk bulgusu olarak prezentasyonu çok nadirdir ve bu birlikteliğin mekanizmaları tartışıldı.

Anahtar sözcükler: akut pankreatit, kistik fibrozis, Henoch-Schonlein purpura

Introduction

Acute pancreatitis (AP) is rarely seen in children (1). It is commonly associated with viral infections, trauma, biliary diseases, and drugs (1,2). Herein we report 2 children with AP associated with unusual etiologies: 1 case was associated with Henoch-Schonlein purpura (HSP) and the other with cystic fibrosis (CF).

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Case reports

Case 1

A 10-year-old boy presented to the pediatric emergency department with a 3-day history of abdominal pain and vomiting. He had colicky, intermittent umbilical abdominal pain without diarrhea. There was no history of chronic illness, drug use, allergy, or trauma. On presentation his height and weight were in the 50th and 75th percentiles, respectively. His blood pressure was 100/70 mmHg, his pulse was regular at 88 beats/min, and body temperature was 36.5 °C. Physical examination

was normal, except for umbilical tenderness. There was no hepatosplenomegaly, and bowel sounds and rectal examination were normal.

Laboratory findings, including complete blood count, liver enzymes, renal and liver function tests, blood gas analysis, and urinalysis, were within normal limits. Clotting profile was normal. Serum amylase (1180 IU L^{-1}, normal range: 0-100 IU L^{-1}) and lipase (142 U L^{-1}, normal range: 0-60 U L^{-1}) were markedly elevated. The C-reactive protein concentration and erythrocyte sedimentation rate were 2.8 mg dL^{-1} and 40 mm h^{-1}, respectively. Occult stool blood was negative.

Abdominal ultrasonography revealed parenchymal non-homogeneity at the head, body, and tail of the pancreas, with minimal free fluid within the intestines, suggesting early stages of acute pancreatitis. A diagnosis of AP was made based on clinical, laboratory, and radiological findings. He was hospitalized, restricted from oral feeding, given supportive treatment (attention to volume status and electrolyte balance), and provided pain management using narcotic agents. Within hours of his admission, abdominal pain became more frequent and the amylase level increased (1989 IU L^{-1}), and intravenous octreotide was initiated (2-3 μg kg^{-1} h^{-1}). Serological tests performed to identify the etiological cause of AP, including hepatitis virus, mumps, and other viruses, were negative and lipid levels were within normal limits.

Two days later the patient had purpuric lesions on both buttocks and the lower extremities (Figure 1). His amylase and lipase levels remained high (1245 IU L^{-1} and 98 U L^{-1}, respectively). Occult stool blood was (4+). IgA, C3, and C4 levels were within normal limits. ANA, pANCA, and Anti-ds DNA were negative. Abdominal tomography revealed peripancreatic fluid retention and diffuse pancreatic swelling. According to these clinical features the patient was diagnosed as HSP, with pancreatic and intestinal involvement. Intravenous octreotide was discontinued and prednisolone 2 mg kg^{-1} day^{-1} was

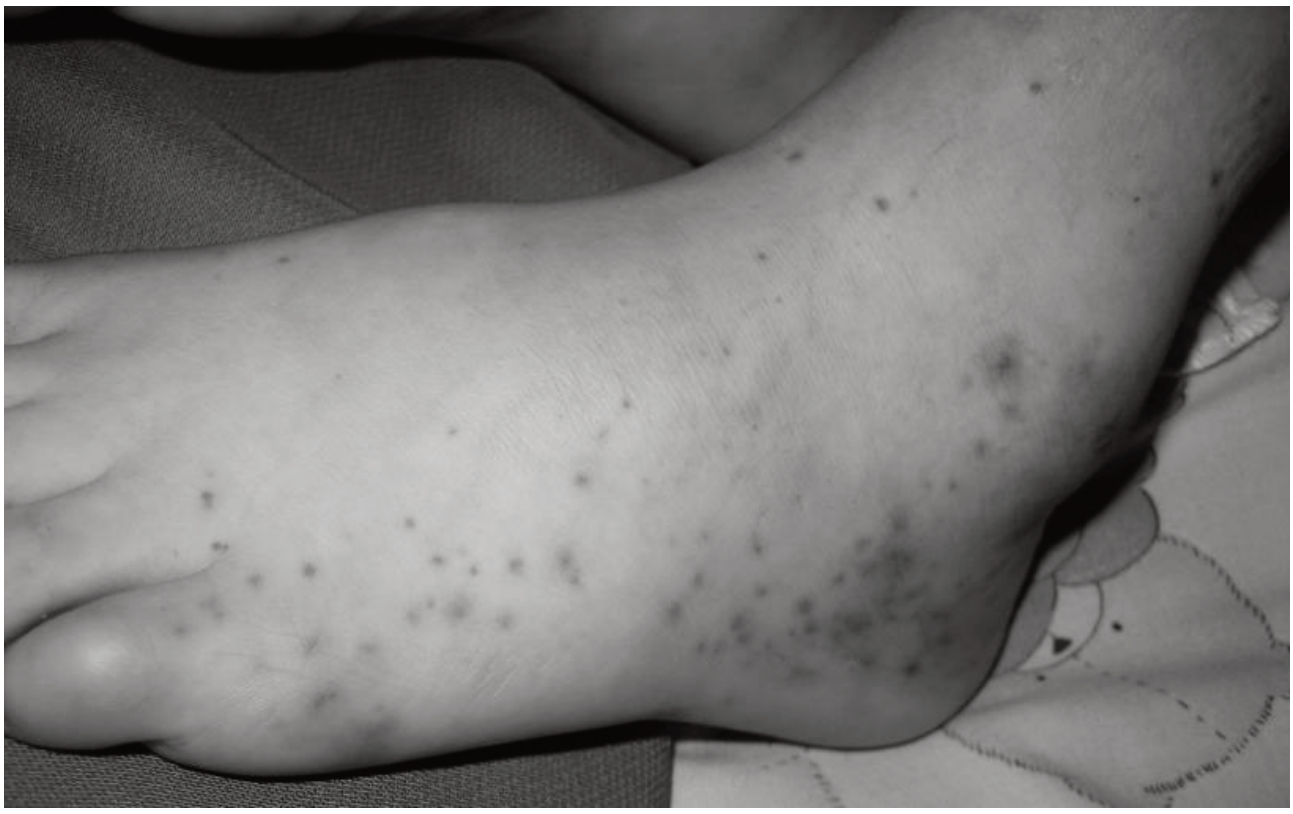

Figure 1. Palpable purpura over the lower extremities in case 1 that developed 2 days after the diagnosis of AP.

administered. Abdominal pain dramatically reduced, amylase and lipase levels decreased, and oral feeding was started 5 days later. He was discharged from the hospital within 1 week, and prednisolone was tapered and discontinued within 1 week at the outpatient clinic.

Case 2

A 14-year-old boy presented to the pediatric emergency service with sudden onset umbilical abdominal pain that spread bilaterally to the lumbar regions. He also had bilious vomiting and watery diarrhea for 2 days. There was no history of trauma, drug use, chronic illness, or recurrent abdominal pain. Family history of chronic illness was unremarkable.

Vital signs were within normal limits at the time of presentation. His height and weight were in the 10^{th}-25^{th} percentile. He had epigastric tenderness, and bowel sounds and rectal examination were normal. Other systemic examinations revealed normal findings, apart from digital clubbing.

Laboratory findings, including complete blood count, liver enzymes, renal and liver functions tests, blood gas analysis, and urinalysis, were within normal limits. Clotting profile was normal. Serum amylase and lipase were 1211 IU L^{-1} and 125 U L^{-1}, respectively. Urine amylase was 2019 U L^{-1} (normal range: 0-460 IU L^{-1}). C-reactive protein and the erythrocyte sedimentation rate were slightly elevated. Abdominal ultrasonography showed pancreatic head enlargement, edema along the body of the pancreas, and retroperitoneal fluid within the anterior pararenal space.

The patient was hospitalized for AP, restricted from oral feeding, given supportive treatment, and provided pain management using narcotic agents. The following day he had fever and attacks of severe abdominal pain; intravenous octreotide (2-3 μg kg^{-1} h^{-1}), in addition to broad spectrum antibiotics, was initiated. In the days that followed his fever persisted, despite antibiotic treatment. Magnetic resonance cholangiopancreatography revealed pancreatic enlargement, heterogenic contrast perfusion, and a small pseudocyst (5 mm in diameter) (Figure 2). Biliary and pancreatic tracts were normal. An antibiotic effective against anaerobic microorganisms was added to the treatment. Five days later

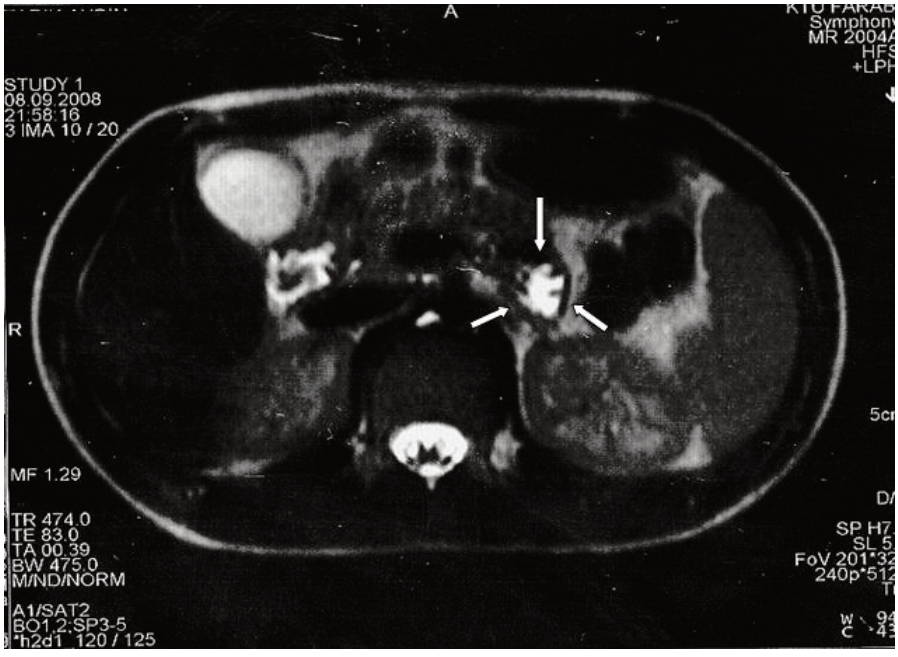

Figure 2. Magnetic resonance cholangiopancreatography of case 2 shows pancreatic enlargement and heterogenic contrast perfusion. Note the pseudocyst formation (with white arrows).

intravenous octreotide was discontinued and enteral feeding was initiated due to reduced abdominal pain and a decrease in the amylase level. Antibiotics were discontinued within 3 weeks and the patient was discharged.

Serological tests to identify the etiological cause of AP, including hepatitis virus, mumps, and other viruses, were negative and lipid levels were within normal limits. The results of 2 sweat chloride tests performed 1 week apart due to the presence of digital clubbing were 85 mmol L^{-1} and 110 mmol L^{-1}, respectively. Mutation analyses, including 12 common mutations for CF, were negative.

During follow-up the patient had a recurrent pancreatic attack 1 month after discharge, which was successfully treated with supportive treatment.

Discussion

We presented 2 children with atypical presentations of AP. The first presented with the clinical features of AP, developed palpable purpura after hospitalization, and was diagnosed with HSP with pancreatic and intestinal involvement. The second had all the clinical features of AP, in addition to digital clubbing. Sweat chloride test results supported the diagnosis of CF. AP is a rare clinical entity in children, and most pediatric cases are related to multisystem diseases, especially hemolytic uremic syndrome, trauma, and biliary diseases (1,2).

Pancreatitis in association with vasculitis has been reported mostly in patients with systemic lupus erythematosus (SLE) and polyarteritis nodosa. Richer et al. studied the abdominal manifestations of 201 children with SLE and reported that 5.9% had clinical pancreatitis (3), which was also observed in 5% of patients with systemic necrotizing vasculitides (4). HSP is the most common systemic vasculitis in childhood. Abdominal manifestations, such as abdominal pain and occult bleeding, may be seen in 50%-70% of patients (5). Intussusception is the most common surgical complication of HSP, occurring in 0.8%-13.6% of patients (6). Pancreatitis in association with HSP is a rare condition and only 5 pediatric cases have been reported in the English language literature (7-10). Chen et al. studied the gastrointestinal manifestations and complications of HSP in 208 children and reported that only 1 child (0.4%) had pancreatitis (11). Vasculitis of the pancreatic vessels or IgA immune complex deposition may cause ischemic necrosis of the pancreas in children with HSP. The classical presentation of pancreatitis in association with HSP develops within 1 week after characteristic palpable purpura, but sometimes as long as 2 months later. Pancreatitis as an initial manifestation of HSP is very rare. Only 2 pediatric cases have been reported and the typical palpable purpura developed within 1 week in both (8,9). Pancreatitis was the initial manifestation of HSP in our case in which typical palpable purpura developed 2 days after the initial diagnosis, and the clinical condition improved after corticosteroid treatment. Pancreatic involvement is generally self-limited and benign, and is rarely complicated by pseudocyst formation.

CF is a rare cause of AP in children. The incidence of pancreatitis in patients with CF is 1.24%, much higher than in pancreatic-sufficient patients, and is usually associated with milder mutations (12). Pancreatitis may be the initial manifestation of the disease, as it was in our case, and patients may be diagnosed after attacks of pancreatitis. Although we did not measure fecal pancreatic elastase or stool fat to assess pancreatic status, the absence of chronic diarrhea and normal growth suggested pancreatic sufficiency in our patient. The presence of digital clubbing prompted us to consider CF in the differential diagnosis. Positive sweat chloride test results confirmed the diagnosis of CF and the recurrence of pancreatitis 1 month later further supported the diagnosis. Mutation analysis for the 12 common mutations of CF was negative in our patient. Most pancreatic-sufficient CF patients with pancreatitis have mild mutations (class IV and V) and a detailed mutation analysis in our patient may have revealed a mild mutation (13). It is important to differentiate the diagnosis of CF in patients with idiopathic AP, not only for the correct diagnosis, but also to prevent recurrences. Fatty meals, stress, alcohol, and medications must be avoided in these patients.

Pancreatic-sufficient CF patients with both high lipase levels and 1 mild/variable mutation indicate an

ongoing inflammatory process in a still-functioning pancreas. mRNA transcript levels in these patients are between 3% and 8%, and they have partially active CFTR protein (13). These patients have semi-viscid pancreatic secretions, leading to recurrent ductular obstruction and attacks of pancreatitis. With time, acinar cells will decrease to below the critical level due to recurrent pancreatic attacks, and patients may become pancreatic insufficient. Recurrent pancreatitis may be the cause of pancreatic insufficiency in CF patients with mild mutations that do not normally cause pancreatic insufficiency.

In conclusion, we presented 2 children with AP as an atypical presentation of HSP and CF. AP may be the initial manifestation of HSP. Pancreatitis must be included in the differential diagnosis of abdominal pain in such children. CF must be considered in patients with idiopathic AP, especially pancreatic-sufficient patients, and digital clubbing may be a clue to the possibility of CF. Long-term follow-up of pancreatic-sufficient CF patients is important, not only for pancreatic functions, but also for identifying complications associated with mild mutations, such as infertility and diabetes mellitus.

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