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Acute pancreatitis in IgA vasculitis

Ostre zapalenie trzustki w przebiegu zapalenia naczyń związanego z IgA

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Abstract

IgA vasculitis, also known as the Henoch–Schönlein purpura, is the most common systemic vasculitis in children. Inflammation most often involves small blood vessels of the skin, joints, kidneys and the gastrointestinal tract, but other organs may also be involved. Acute pancreatitis is a rare clinical manifestation of IgA vasculitis. We present a case of a 12-year-old girl with cutaneous and abdominal symptoms, who was diagnosed with acute pancreatitis during IgA vasculitis. In patients with IgA vasculitis who report abdominal pain acute pancreatitis should be taken into account and pancreatic enzymes measurement should be considered.

Keywords: acute pancreatitis, Henoch–Schönlein purpura, IgA vasculitis, amylase

Streszczenie

Zapalenie naczyń związane z IgA, nazywane również plamicą Schönleina–Henocha, jest najczęstszym układowym zapaleniem naczyń wieku dziecięcego. Proces zapalny dotyczy najczęściej drobnych naczyń krwionośnych skóry, stawów, nerek oraz przewodu pokarmowego, ale w przebiegu choroby mogą zostać zajęte również inne narządy. Ostre zapalenie trzustki jest rzadką manifestacją kliniczną zapalenia naczyń związanego z IgA. W pracy przedstawiono przypadek 12-letniej dziewczynki z objawami skórnymi i brzusznymi, u której w przebiegu zapalenia naczyń związanego z IgA rozpoznano ostre zapalenie trzustki. U pacjentów z zapaleniem naczyń związanym z IgA zgłaszających dolegliwości bólowe ze strony jamy brzusznej należy wziąć pod uwagę możliwość występowania ostrego zapalenia trzustki oraz rozważyć oznaczenie stężeń enzymów trzustkowych.

Słowa kluczowe: ostre zapalenie trzustki, plamica Schönleina–Henocha, zapalenie naczyń związane z IgA, amylaza

INTRODUCTION

IgA vasculitis (IgAV), formerly known as Henoch–Schönlein purpura (HSP), is an acute inflammation of small blood vessels associated with IgA-dominant immune-complex deposition in their walls[1]. Although the vessels of the skin, joints, kidneys and gastrointestinal tract are most often affected, other organs, such as the lungs or the central nervous system, may be also involved[2]. In this paper, we present a case of a patient with IgAV, who was diagnosed with acute pancreatitis. The aim of this paper is to draw attention to the fact that abdominal pain in patients with IgAV may be a symptom of acute pancreatitis.

CASE REPORT

A 12-year-old girl was admitted to the Department of Paediatrics, Immunology and Nephrology, Polish Mother's Memorial Hospital Research Institute in Łódź (Poland) due to abdominal pain recurring periodically before hospital stay and petechial rash on the lower limbs persisting for about a week prior to admission. The recurrent abdominal pain was moderate and located in the epigastrium. It was accompanied by nausea without vomiting. The girl underwent a dermatological consultation 2 days before hospitalisation due to the presence of skin lesions. Folliculitis was diagnosed and local treatment was initiated. Three weeks before admission, the child was treated with amoxicillin with clavulanic acid due to acute upper respiratory tract infection. She had a medical history of chronic gastritis at the age of 10 years. On admission, the girl was in good overall condition. Physical examination revealed small petechiae on the skin of the lower limbs, most severe in the area of the external parts of the ankles and the dorsal side of feet, but also present on the upper limbs around the wrists and on the buttocks. The patient had excess subcutaneous tissue, with body mass index (BMI) estimated at 98th percentile. The joints of the upper and lower limbs were painless, without oedema or increased temperature. The throat was red, there was a normal alveolar murmur above the lung fields, and a regular heart rate of 80/min was recorded. The abdomen was soft, with epigastric tenderness on compression, audible peristalsis, and non-enlarged parenchymal organs. There were no peritoneal symptoms.

Laboratory work-up on admission showed leukocytosis 16 × 10^3/μL (N: 4–10 × 10^3) with 80% neutrophilia, C-reactive protein (CRP) 1.28 mg/dL (N: <1 mg/dL), insignificant changes in the coagulation profile: prolonged prothrombin time to 14.0 s, increased D-dimers – 2,173 ng/mL, erythrocyturia in the general urine test – 18/μL, positive faecal occult blood test, normal markers of renal and liver function, and parameters of the acid-base balance within the normal range.

IgA vasculitis was diagnosed based on the medical history, physical examination and laboratory findings. Symptomatic treatment was initiated: bed regimen, hydration, pharmacotherapy to reduce vascular permeability (etamsylate at 250 mg 3 times a day), analgesics (paracetamol at 1 g, up to 3 times a day). Due to the symptoms of upper respiratory tract infection, empirical oral antibiotic therapy with cefuroxime axetil at a dose of 250 mg given every 12 hours was included. In the first days of hospital stay, the patient felt well, periodically reported abdominal pain, the petechiae gradually resolved, but single new lesions occurred on the skin of the lower extremities.

On the 4th day of treatment, several episodes of menstrual lower abdominal pain occurred, but resolved after alternating administration of paracetamol and drotaverine.

On the 7th day of treatment, several night vomiting episodes accompanied by severe epigastric pain occurred. They were described by the patient as acute. The spread of petechiae was observed on the upper limbs in the area of the elbows and on the buttocks and the right lower leg. Laboratory work-up was done and showed: serum amylase 145 U/L (N: 30–100 U/L), urine amylase 1,616 U/L (N: 32–641 U/L), serum lipase 825 U/L (N: 10–180 U/L), leukocytosis 19 × 10^3/μL with neutrophilia, CRP 4.1 mg/dL (N: <1 mg/dL), D-dimers 14,378 ng/mL (N: <500 ng/mL), as well as normal levels of aspartate aminotransferase, alanine aminotransferase, gamma-glutamyl transpeptidase, alkaline phosphatase and bilirubin. Abdominal ultrasound showed a non-enlarged pancreas with homogeneous structure and normal parenchymal echogenicity; gallbladder with normal walls and no signs of gallstones; non-enlarged liver with a hyperechoic parenchymal area 30 mm in diameter, likely to correspond to a haemangioma; and an inhomogeneous cyst 26 mm in diameter in the right ovary.

Based on the clinical symptoms and additional tests, the patient was diagnosed with IgAV-associated pancreatitis. Oral nutrition was discontinued, parenteral nutrition via continuous intravenous infusion was initiated. Due to the increase in the inflammatory parameters, antibiotic therapy was switched to a third generation cephalosporin – ceftriaxone at a dose of 2 g given every 24 hours. Omeprazole 20 mg twice a day was included in the treatment and, in the case of pain, paracetamol at 1 g was administered up to 3 times a day, and drotaverine 40 mg was administered up to 2 times a day.

Over the next 3 days of hospital stay, abdominal pain persisted and new petechiae on the upper and lower limbs occurred. Further increase in pancreatic parameters was observed: serum amylase up to 227 U/l (N: 30–100 U/L), urine amylase up to 2,101 U/L (N: 32–641 U/L), serum lipase up to 2,433 U/L (N: 10–180 U/L). Due to the increasing difficulties in obtaining peripheral intravenous access and newly developing rash, a central vascular access was established.

The treatment led to systematic improvement of the patient's general condition (the pain resolved, no new rash appeared), starting from the 12th day of hospitalisation. Follow-up laboratory tests showed reduced pancreatic parameters, D-dimmers and inflammatory markers. After consultation, the patient was transferred to the Department of Gastroenterology, Allergology and Paediatrics of the Polish

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Mother's Memorial Hospital Research Institute, where the internal jugular vein catheter was removed, parenteral nutrition was discontinued and a soft diet was prescribed. The treatment and diet expansion were continued, achieving further improvement in the child's clinical condition and resolution of skin lesions. Inflammatory parameters and pancreatic enzymes normalised. On day 17 after admission, the patient was discharged home in good general condition, with recommendations for further outpatient care, a light diet, limited physical activity for 2 weeks and the following outpatient follow-up tests: serum levels of pancreatic enzymes and urine amylase concentration after 7 and 14 days, and abdominal ultrasound in 3–4 weeks. The girl's parents were also informed on the need for further specialist care in gastroenterological and dietary clinics.

DISCUSSION

IgAV is the most common systemic vasculitis in children. The estimated annual incidence of IgAV is 10–20 cases per 100,000 population[3,4]. Although the disorder may develop at any age, 90% of cases are children aged 2–10 years, with peak incidence between the ages of 4 and 7 years[1–4]. In most cases, IgAV is preceded by an upper respiratory tract infection, vaccination or pharmacotherapy, including antibiotics or anti-inflammatory drugs[3]. Palpable purpura, which develops in 95–100% of patients, is the most characteristic symptom associated with skin vessel involvement. It appears as macular or maculopapular inflammatory haemorrhagic papules, which do not disappear after applying pressure, and are located mainly on the lower limbs, upper limbs and buttocks. Other clinical manifestations are most often associated with the involvement of joints, kidneys and the gastrointestinal tract[3]. Gastrointestinal involvement is reported in 50% of IgAV cases[4]. It manifests with abdominal pain, typically colic or (less often) severe, imitating acute surgical abdominal diseases, as well as nausea and vomiting or gastrointestinal bleeding (usually latent)[2,5]. Intussusception, perforation, and intestinal ischaemia are the most common gastrointestinal complications[2,6].

Joint involvement is often reported in IgAV, accounting for 60–84% of cases[4]. The knee and ankle joints are most likely to be affected by inflammation and pain. Oedema, pain and limited joint movement are typically observed. However, these symptoms are usually transient and do not cause permanent joint damage[2,3,6].

Urinary symptoms develop in 30–50% of patients with IgAV. Isolated microscopic haematuria, usually developing up to 4 weeks after the onset of the disease, is the most common symptom. The range of urinary clinical manifestations is very wide: from the already mentioned haematuria, through proteinuria, nephrotic syndrome, nephritic syndrome, to acute kidney damage or chronic kidney disease and end-stage renal failure. Although kidney involvement can lead to serious complications, it is mild and fully resolves in most patients[2,3,6]. The diagnosis is based on the clinical picture and the 2010 diagnostic criteria of the European League Against Rheumatism (EULAR), Paediatric Rheumatology International Trials Organization (PRIN-TO) and Paediatric Rheumatology European Society (PRES)[7].

Our patient presented with IgAV-specific relapsing skin lesions, abdominal pain, occult blood in the stool, as well as slight haematuria – before menstruation.

IgAV in children is often self-limiting[4,8]. Mild cases are treated symptomatically with bed regimen, hydration, pharmacotherapy to reduce vascular permeability and analgesics. Non-steroidal anti-inflammatory drugs (NSAIDs) can be used in arthritis with preserved kidney function. If the clinical picture indicates an overlapping infection, antibiotic therapy is included. Steroid therapy may be considered in the case of gastrointestinal or renal involvement[1,2]. The use of other immunosuppressive and immunomodulatory agents is reserved for severe, chronic and complicated cases[1,3].

Abdominal pain, which is the most common gastrointestinal symptom in IgAV, can be also caused by acute pancreatitis. Acute pancreatitis is a rare complication of IgAV, most commonly seen in adolescent girls[9]. It can occur simultaneously, several days or weeks after the onset of a characteristic macular rash, or even precede the typical symptoms of IgAV[8,10,11]. Only few such cases have been reported so far[8–14]. Chen and Kong showed that only 0.48% of 208 patients with IgAV and gastrointestinal symptoms had elevated amylase and lipase levels[14]. Zhang et al. identified 13 children with pancreatitis among 3,212 patients with IgAV (0.4%)[8].

Serum amylase, lipase and urine amylase measurements are reliable diagnostic tools for acute pancreatitis[8,13]. It is suggested that these tests should be performed in any IgAV patient with severe or typically localised abdominal pain. It can be assumed that more common measurements of pancreatic enzyme activity in children with IgAV could show more frequent involvement of the pancreatic vessels than suggested by single clinical reports[10]. Abdominal ultrasound plays an auxiliary role in the diagnosis, allowing for a morphological assessment of the pancreas, and is also useful in differentiating the causes of acute pancreatitis (e.g. gallstone disease)[15]. Changes in ultrasound or computed tomography (CT), such as oedema or necrosis, characteristic of pancreatitis, can be observed in severe acute pancreatitis[15].

According to the definition adopted by the INternational Study Group of Pediatric Pancreatitis: In Search for a CuRE (INSPPIRE), at least 2 out of 3 criteria must be met to diagnose acute pancreatitis: abdominal pain compatible with acute pancreatitis, amylase and/or lipase levels of 2–3 times the upper limit of normal (ULN), changes in ultrasound or CT suggestive of acute pancreatitis[5]. In our patient, acute pancreatitis was diagnosed based

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on the clinical symptoms and tests assessing the parameters of the secretory function of the pancreas. Although acute pancreatitis is mild in most cases, complications such as bleeding, necrosis, and pseudocysts may occur[9]. Recommended management involves discontinuation of oral food intake and initiation of parenteral nutrition, adjusted to the patient's energy requirements[15]. Water and electrolyte imbalance should be corrected, proton pump inhibitors and analgesics should be used[13,15]. Due to the significantly elevated inflammatory parameters and symptoms of upper respiratory tract infection, our patient was also treated with broad-spectrum intravenous antibiotic therapy. Steroid therapy is not routinely used in the treatment of acute pancreatitis, but it should be considered in IgAV[8,9,13]. The recommendations also include the use of somatostatin, which inhibits gastric secretion and reduces the activity of pancreatic enzymes[9].

CONCLUSIONS

Acute pancreatitis is a rare complication of IgAV. In patients with IgAV and severe abdominal pain serum pancreatic enzyme levels measurement should be considered to exclude acute pancreatitis.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organisations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

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