



CASE REPORT

Henoch-Schönlein purpura

Natalia Zdanowska, Agnieszka Owczarczyk-Saczonek*, Waldemar Placek

Department of Dermatology, Sexually Transmitted Diseases and Clinical Immunology, Faculty of Medical Sciences, University of Warmia and Mazury in Olsztyn, Poland

Abstract: Henoch-Schönlein purpura (HSP) is an acute, systemic vasculitis which usually occurs in young adults and children. The skin involvement may lead to the manifestation of symptoms associated with vasculitis in intestines, kidneys, and the central nervous system. The incidence of bowel perforation in course of HSP is very seldom and it occurs about 10 days after the appearance of the first symptoms. We present a 23-year-old male patient with jejunal intussusception in the course of HSP. The patient was operated urgently with resection of part of the small intestine. Adults rarely suffer from the occurrence of abdominal pain and fever, but sometimes they require careful monitoring and surgical intervention because misdiagnosis can be life threatening.

Keywords: Henoch-Schönlein purpura, jejunal intussusception, haemorrhagic bullous lesions

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*Correspondence to: Agnieszka Owczarczyk-Saczonek, Department of Dermatology, Sexually Transmitted Diseases and Clinical Immunology, Faculty of Medical Sciences, University of Warmia and Mazury in Olsztyn, Poland; aganek@wp.pl

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Introduction

Henoch-Schönlein purpura (HSP) is an acute, systemic autoimmune vasculitis which usually occurs in young adults and children. Skin involvement may lead to the manifestation of symptoms associated with vasculitis in intestines, kidneys, and the central nervous system. Rarely does the disease lead to a patient's death. Typical symptoms of HSP are purpura, arthralgia, abdominal pain, and bloody stools. Histopathological examinations show leukocytoclasia of small vessels with the deposition of immune complexes consisting mainly of immunoglobulin A. Although the etiology is still unclear, symptoms can develop after an upper respiratory tract infection (especially β -hemolytic streptococci) or can be provoked by drugs (e.g., ampicillin, erythromycin, penicillin, quinidine, and losartan)^[1-6]. The incidence of bowel perforation in the course of HSP is 0.38%. Typically, the disease is localized in the ileum and jejunum, where perforation occurs about 10 days after the appearance of

its first symptoms^[4-7].

Case presentation

A 23-year-old male patient, without chronic diseases, was admitted to the Department of Dermatology, Sexually Transmitted Diseases and Clinical Immunology in Olsztyn, Poland, for symmetrical maculopapular and hemorrhagic lesions with blister formation, which may correspond to Henoch-Schönlein purpura. Lesions were localized on the lower limbs, around the hock joints. The first skin lesions, initially papular, which occurred about 15 days before admission, were accompanied by mild upper respiratory tract infection treated symptomatically and were located on the upper and lower limbs. Ten days later, the patient was admitted to the Department of Surgery of the Hospital in Olsztyn due to symptoms of acute abdomen and was diagnosed with jejunal intussusception. The patient was urgently operated on. A 20-cm resection of the small intestine was performed. Histopathologi-

cal changes were described as necrosis of the walls of the small intestine. During hospitalization in the Department of Surgery, a gradual increase of hemorrhagic lesions was observed; for this reason, the patient received two consecutive days of dexamethasone (sodium phosphate) at a dosage of 8 mg/day, with improvement. In laboratory studies collected on admission to the Department of Dermatology, we observed elevated transaminases (alanine aminotransferase (ALT) 42 U/L, aspartate aminotransferase (AST) 57 U/L), elevated markers of inflammation (C-reactive protein (CRP) 38.20 mg/L), leukocytosis, thrombocytopenia (white blood cells (WBC) $10.2 \times 10^3/L$, platelets (PLT) $398 \times 10^3/L$), and elevated antistreptolysin (ASO) (400 U). In successive hospitalization days, we also observed changes in the level of transaminases (4 hospitalization days: AST 42 U/L, ALT 90 U/L; 5 hospitalization days: AST 48 U/L, ALT 104 U/L; 9 hospitalization days: AST 35 U/L, ALT 128 U/L), while serological tests for viral hepatitis B and C were negative. There was absence of antibodies cANCA and pANCA. Pharyngeal swab showed *Moraxella catarrhalis*, and antibacterial therapy in accordance with antibiogram was performed (cefuroxime 3 g/day intravenous therapy (IV) in two divided doses). The patient was treated with intravenous glucocorticoids (dexamethasone 12 mg/day) and with topical application (betamethasone and gentamycin). After treatment, we observed improvements and patient was discharged in good general condition. The patient stayed under the care of the Dermatological Outpatient Clinic and the lesions did not recur. Currently, he has a few post-inflammatory hyperpigmented macules on the lower limbs. Laboratory tests did not show any abnormalities.

Discussion

We decided to describe the case because of the relatively rare occurrence of Henoch-Schönlein purpura in adults. It should be noted that the course of HSP in adults is usually more severe and thus requires careful monitoring. Adults rarely suffer from the occurrence of abdominal pain and fever, but are more likely to have renal involvement^[2]. Symptoms of the gastrointestinal tract occur in 45% to 75% of patients with HSP, and in 14% to 36% of patients, it became purpuric^[8-10]. Approximately 52% of patients with HSP are found to have varying degrees of severe gastrointestinal bleeding, also provoked by the use of corticosteroids to treat the disease. Sometimes patients require surgical intervention, and misdiagnosis can be life threatening^[9,11].

Intussusception in HSP cases is rarely found in adults.

Two literature reviews that evaluated 250 and 115 adults with HSP, respectively, did not report any cases of intussusception^[10,12]. In Japan, colo-colic intussusception associated with HSP in adults has been reported in two cases^[13]. One of the cases underwent colonoscopy, and intussusception was reduced by colonoscopic insufflation^[13].

In contrast, intestinal ischemia, perforation, hemorrhage, fistulas, pancreatitis, and appendicitis are rare^[9,14]. However, Zhang *et al.*^[10] in their assessment of adult patients with gastrointestinal manifestations of HSP showed that the first symptoms may appear as “acute abdomen” without lesions, and that gastroscopy and colonoscopy may be helpful in early diagnosis. Typical results include endoscopic mucosal edema diffuse rash, ecchymoses, or small ulcers, especially in the duodenum or terminal ileum^[10]. Carmichael *et al.* reported a case of a massive, fatal intestine ischemia^[15], and Lawes and Wood described a case of a 41-year-old man with ischemia of the small intestines and symptoms of acute abdomen with the appearance of purpura^[16].

Among the etiological factors rarely mentioned is *Moraxella catarrhalis*. Most cases are caused by bacteria *Streptococcus pyogenes*, *Haemophilus influenzae*, or *Staphylococcus aureus*^[17] and viruses (*e.g.*, adenoviruses, Epstein-Barr virus, or parvovirus)^[18]. The drugs of choice are still glucocorticoids which reduce the risks of developing complications. Weiss *et al.* in 1895 analyzed the hospitalizations of children with HSP and found that early administration of corticosteroids significantly reduced the risk factors for abdominal surgery^[19]. Furthermore, randomized, double-blind, placebo-controlled studies confirmed that prednisolone at a dosage of 1 mg/kg per day for two weeks reduced the onset of gastrointestinal symptoms^[20]. In severe cases, in addition to corticosteroids, drugs such as cyclophosphamide, cyclosporine, and azathioprine can be used^[21].

Conclusion

Intussusception, along with severe complications of HSP, very seldom occurs in adults. Therefore, in each patient who suffers from HSP with abdominal pain, this symptom should be considered.

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