

—Report on Experiments and Clinical Cases—

A Case of Henoch-Schönlein Purpura with Rare Complications: Necrosis of the Small Intestine, Neurological Symptoms, and Pericardial Tamponade

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Abstract

A case of Henoch-Schönlein purpura (HSP) with necrosis of the small intestine, neurological symptoms, and pericardial tamponade after frequent recurrence is described. Neurological symptoms were controlled well with steroid pulse therapy, and pericardial tamponade was treated successfully with pericardiocentesis and steroid pulse therapy. To treat necrosis of the small intestine, the necrotic tissue was excised and artificial anuses were constructed. Five months later, the small intestine was anastomosed in a curative operation. Periodic administration of coagulation factor XIII was required from the onset of symptoms until curative surgery, but the activity of this factor returned to normal levels after surgery operation. We report a case of Henoch-Schönlein purpura with extremely rare complications: necrosis of the small intestine, neurological symptoms, and pericardial tamponade.

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Key words: Henoch-Schönlein purpura, factor XIII, complications

Introduction

Henoch-Schönlein purpura (HSP) is a disease occasionally seen in childhood, which is characterized by dermatological symptoms (ecchymosis, papular eruption, and localized edema), joint symptoms (swelling and pain), and abdominal symptoms (abdominal pain and bloody stool)¹. Nephritis is one complication that can greatly influence the long-term prognosis^{1,2}. In HSP, the lesions are caused by vascular abnormalities rather than bleeding tendency due to platelet or coagulation

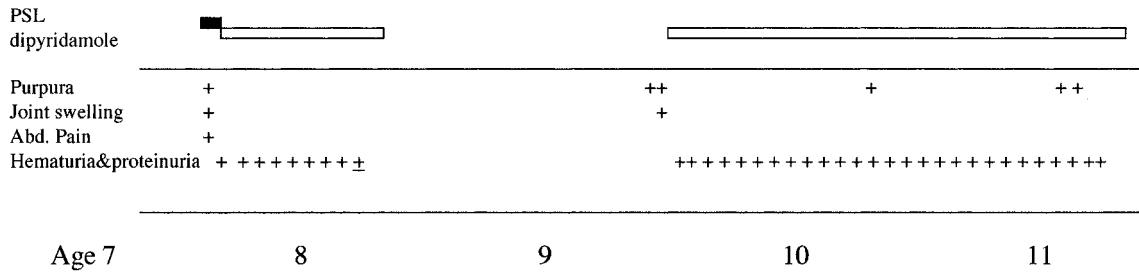
factor abnormalities¹. In the present patient, rare complications, such as disturbance of consciousness, pericardial tamponade, and necrosis of the small intestine, were observed. Although mainly smaller vessels are affected in HSP, intraoperative examination showed that the necrosis of the small intestine in this case was caused by a blood flow disturbance in the jejunal and ileal arteries, which are medium-sized blood vessels.

Case Report

The patient was a 12-year-old boy in whom HSP

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Clinical course(1)



Clinical course(2)

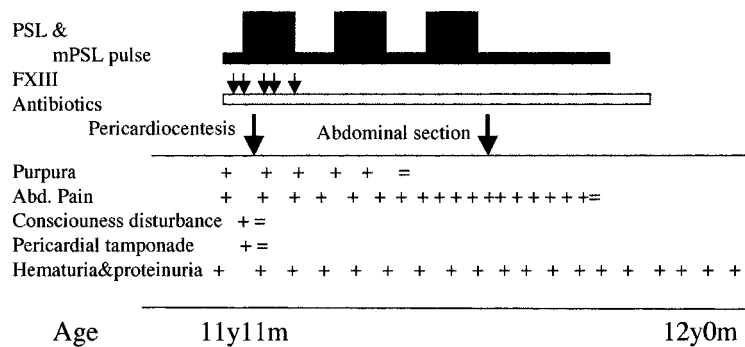


Fig. 1 Clinical course

was diagnosed because of the typical symptoms of HSP: ecchymosis, papular eruption, joint pain of the lower extremities, and abdominal pain. Prednisolone was initially administered for 2 weeks to treat severe abdominal pain. Three weeks later, administration of an antiplatelet agent, dipyridamole, was started because of renal involvement. One year later, as hematuria and proteinuria resolved, dipyridamole was discontinued. HSP accompanied by leg purpura, hematuria, and proteinuria recurred three times during the autumn/winter when the patient was 9, 10, and 11 years old. The abdominal pain was mild, and administration of prednisolone was not required during these recurrences (Fig. 1).

The patient was admitted to our department because of abdominal pain, nausea, and vomiting. He had mild purpura on the legs, and recurrent HSP was diagnosed. At the time of the admission, the patient had intermittent periumbilical pain. Blood tests revealed leukocytosis (mostly neutrophils), but C-reactive protein was negative. That night, abdominal pain increased, and the patient became

more restless and eventually showed disturbed consciousness. Carotid artery engorgement, hypotension, and narrow pulse pressure developed the next day. Echocardiography revealed pericardial tamponade and pericardiocentesis was performed. After aspiration of 90 mL of yellow fluid, blood pressure, and pulse pressure improved. Acute exacerbation of HSP was diagnosed, and pulse therapy was performed with methyl prednisolone. After two cycles of methyl prednisolone pulse therapy, the fever subsided, and the results of a blood test showed improvement (Fig. 1). However, abdominal pain persisted, and paralytic ileus was diagnosed. Abdominal distension gradually worsened, and an abdominal X-ray film showed perforation of the enteric canal. Surgical operation was performed, and necrosis of the small intestine was confirmed (Fig. 2). Ninety centimeters of the jejunum from the ligament of Treitz, and 70 cm of the ileal side of the ileocecum remained unaffected. Therefore, artificial anuses were constructed on each side. Marked vasculitis was seen in the necrotic

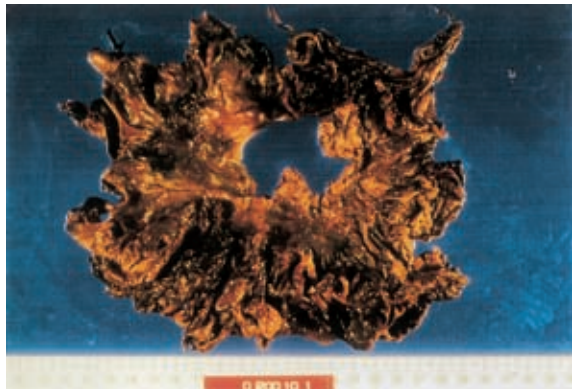


Fig. 2 Section of the small intestine shows necrosis.

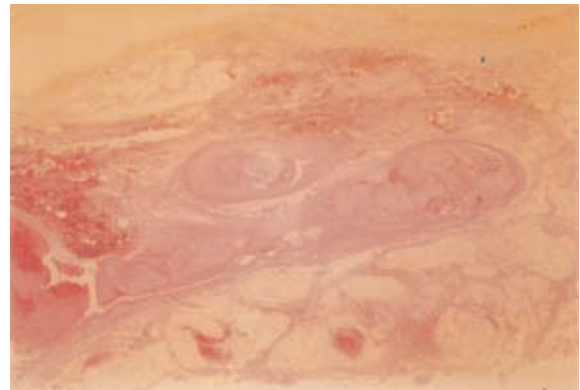


Fig. 3 Microscopic appearance of the necrotic small intestine showing fibrosis and thrombus in vessels.

intestine and in the jejunal and ileal arteries that branched off the superior mesenteric arteries (Fig. 3). The region of the small intestine supplied by these arteries was necrotic. The remaining ileum on the anal side was supplied by the inferior mesenteric arteries. Following surgery, intravenous hyperalimentation was performed to aid nutritional recovery, and intestinal anastomosis as the curative surgery was performed 5 months later. At this stage, biopsy of the feeding vessels of the surrounding intestine was performed, but constrictive lesions were not seen. The activity of coagulation factor XIII decreased to 50% of normal as symptoms appeared. Although the administration of coagulation factor XIII was necessary every 1 or 2 weeks, the activity of this factor returned to normal levels after curative surgery.

Discussion

The prognosis for most patients with HSP is favorable. Although most patients recover within a few weeks, disease recurs in one third of patients. In most of these cases, HSP recurs within 4 months, but recurs after more than 4 months in 6% of cases³. In 50% to 70% of patients with HSP, abdominal symptoms, such as abdominal pain, nausea, and bloody stool, are seen. These symptoms are caused when inflammation of the vessels supplying the enteric canal leads to edema and hemorrhage of the enteric canal wall. Intussusception is seen in 2% of patients with abdominal pain. Sudden ileocolic intussusception does occur, and ileo-ileo

intussusception accounts for two thirds of cases associated with HSP. Although rare, such severe abdominal complications as, perforated enteric canal, massive gastrointestinal bleeding, and protein-losing enteropathy, have been documented⁴. The present patient had neurological symptoms (altered mental state, convulsion, and headache), that was thought to be caused by central nervous vasculitis⁵. There has been a report of cardiomyopathy caused by coronary vasculitis^{6,7}.

The etiology and pathology of HSP remain unclear, but HSP is thought to cause inflammation of small systemic vessels. Vasculitis is classified into three types based on the size of inflamed vessels: large-vessel vasculitis, which affects the aorta and other large vessels in the head and extremities; medium-vessel vasculitis, which affects vessels supplying organs such as the kidney, liver, heart, and enteric canal; and small-vessel vasculitis, which affects vessels within organs⁸. In the present patient, the necrosis of the small intestine may be explained by an obstruction in a branch of the jejunal artery of the superior mesenteric arteries, which are medium-sized arteries. After the patient was admitted to our department, thin-slice computed tomography was performed to evaluate inflammation in other medium-sized vessels in the head and chest, but no clear signs of abnormality were seen. It is still unclear why only the jejunal artery was affected. Because the patient had recurrent HSP, small- and medium-sized vessels around the necrotic enteric canal were examined histopathologically to

determine if all the vessels in the body had become constricted, but no abnormalities were found.

The activity of blood coagulation factor XIII is low in HSP and is correlated with disease severity². Coagulation studies suggest a lack of factor XIII, which is important for both clot formation and wound healing^{9,10}. However, the mechanism of reduced factor XIII activity in HSP is unknown. In the present case, the activity of coagulation factor XIII normalized after curative surgery. This finding suggests that the decrease of factor XIII correlates with severity of HSP, and that factor XIII is involved in the healing process of ruptured vessels in this case.

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