# Henoch-Schönlein purpura

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*Caso clínico comentado*

A 15 - year - old female adolescent presented with paroxysmal tolerable dull pain around the umbilicus two weeks ago. The pain could relieve spontaneously, and there was no obvious acid reflux, belching, nausea, diarrhea, constipation, fever, etc. The local hospital conducted a series of evaluation examinations and tests. The blood routine showed that WBC was $11.7\times10^9$/L, CA199 was 77.18 U/ml. The stool routine showed ++ red blood cells, + occult blood, interleukin-6 was 9.75 pg/ml, and D-dimer was 1.33 mg/L. Abdominal vascular CTA showed: 1. Multiple gallstones; 2. Edema and thickening of the wall of a part of the ileum in the right lower abdomen, considered possible enteritis, autoimmune diseases to be excluded; 3. Multiple lymph node shadows at the root of the mesentery; 4. A cystic lesion beside the left iliac vessel, possibly originating from the left ovary, and dilated intestine could not be excluded; 5. Endometrial thickening; 6. Pelvic effusion; 7. No obvious abnormality was found in the CTA of the upper and lower abdominal vessels. Tests such as coagulation routine, liver and kidney function showed no obvious abnormalities. After symptomatic treatment in the local hospital, the patient's abdominal pain did not improve significantly. At the same time, she developed nausea, vomiting of gastric contents, diarrhea, and passed reddish - brown watery stools. Therefore, the patient came to our hospital for further treatment.

After admission, the abdominal plain film showed local jejunal pneumatosis and slight dilation in the left middle and lower abdomen (Figure 1A and 1B). The blood routine showed that white blood cell (WBC) was $14.41\times10^9$/L, neutrophil percentage (NEUT%) was 85.0%, and 25 - hydroxyvitamin D was 8.00 ng/ml. Electrocardiogram, chest CT, tuberculosis antibody, glycosylated hemoglobin, procalcitonin, TORCH (IgG, IgM), EB virus antibody, immunoglobulin + complement, erythrocyte sedimentation rate, and pre - transfusion tests (in the ward) showed no obvious abnormalities. Considering the previous results, small intestinal diseases could not be excluded as the cause of the patient's symptoms. On the second day after admission, the patient underwent a contrast - enhanced CT of the small intestine (Figure 1C). The results suggested possible intussusception of a local jejunum in the left abdomen, thickening and obvious enhancement of the pelvic ileum wall, possible inflammatory lesions; slightly thickened and enhanced appendix wall, considered possible inflammatory lesions; multiple enlarged lymph node shadows around the mesentery; abdominal and pelvic effusion; gallstones; a cystic shadow in the left adnexal area, possibly a cyst or follicle. After communicating with the general surgery experts, the patient and her family refused surgical laparotomy but agreed to undergo gastroscopy and colonoscopy. On the fourth day after admission, the patient underwent gastroscopy and colonoscopy (Figure 2). Gastroscopy showed chronic non-atrophic antritis, and colonoscopy showed mucositis at the terminal ileum, with a smooth mucosa at the appendiceal orifice and no special abnormalities. On the fifth day after admission, the patient complained of intermittent pain around the umbilicus, but the abdominal pain was improved compared with before. During this period, the patient's D

- dimer was slightly increased compared with before. After a bowel ultrasound, no intussusception was found. On the sixth day after admission, the patient underwent capsule endoscopy, which suggested multiple ulcers in the ileum, ischemic enteritis, and Henoch-Schönlein purpura (HSP) could not be excluded (Figure 3). On the eighth day after admission, scattered red rashes appeared on the back of the hands and feet (Figure 4). Considering the patient's clinical manifestations, abdominal HSP was highly suspected. HSP might induce intussusception, leading to intestinal ischemia and possibly intestinal ulcers. After a multidisciplinary consultation and discussion, the patient received steroid hormone treatment,. The patient's abdominal pain gradually improved, and the skin purpura gradually recovered. The treatment ended on the 13th day after admission. During the follow-up, the patient did not experience abdominal pain or purpura again.

**Figure 1.** *The abdominal plain film and CTE of small intestine*

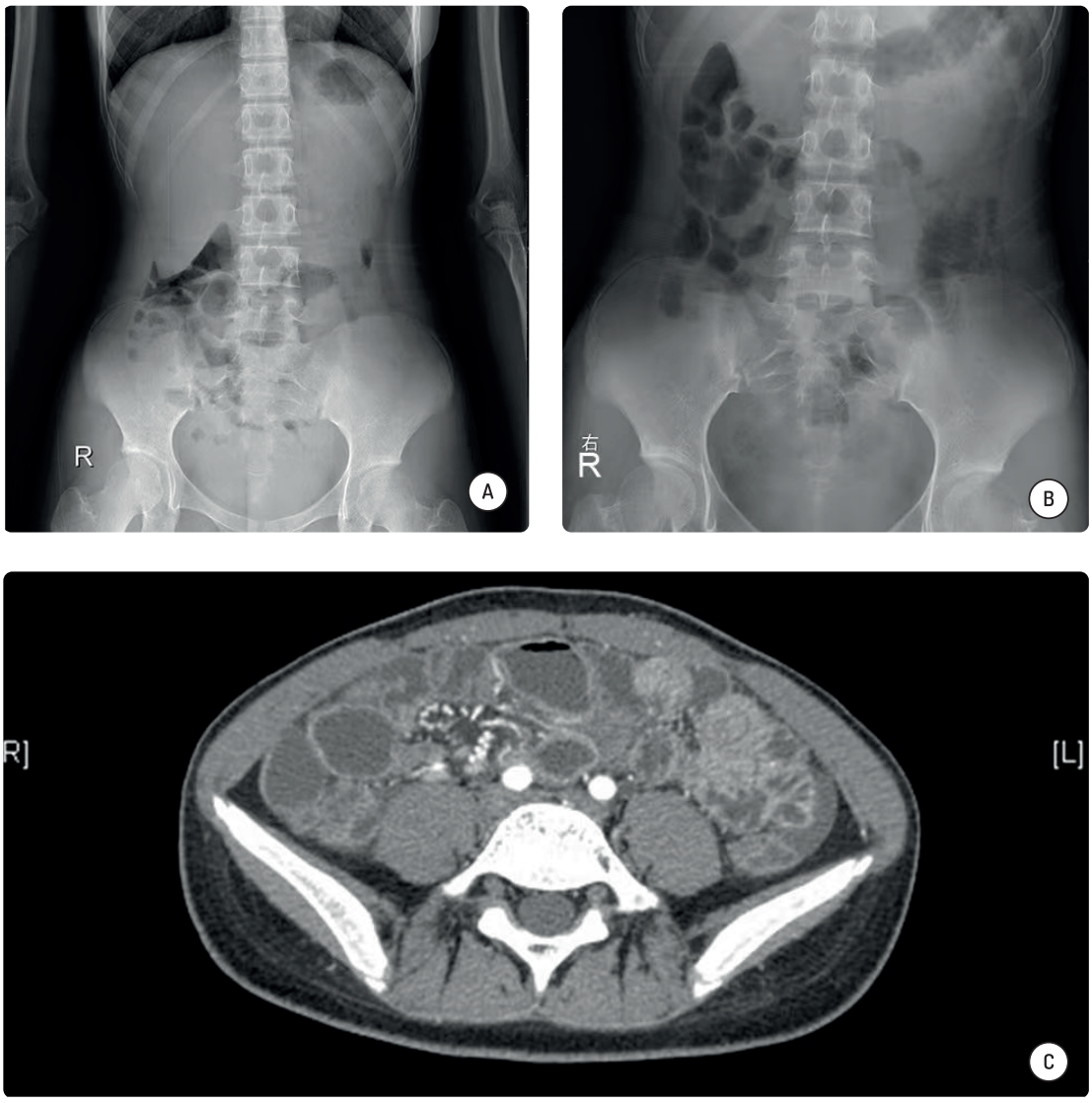

**Figure 2.** *Gastroscopy and Colonoscopy*

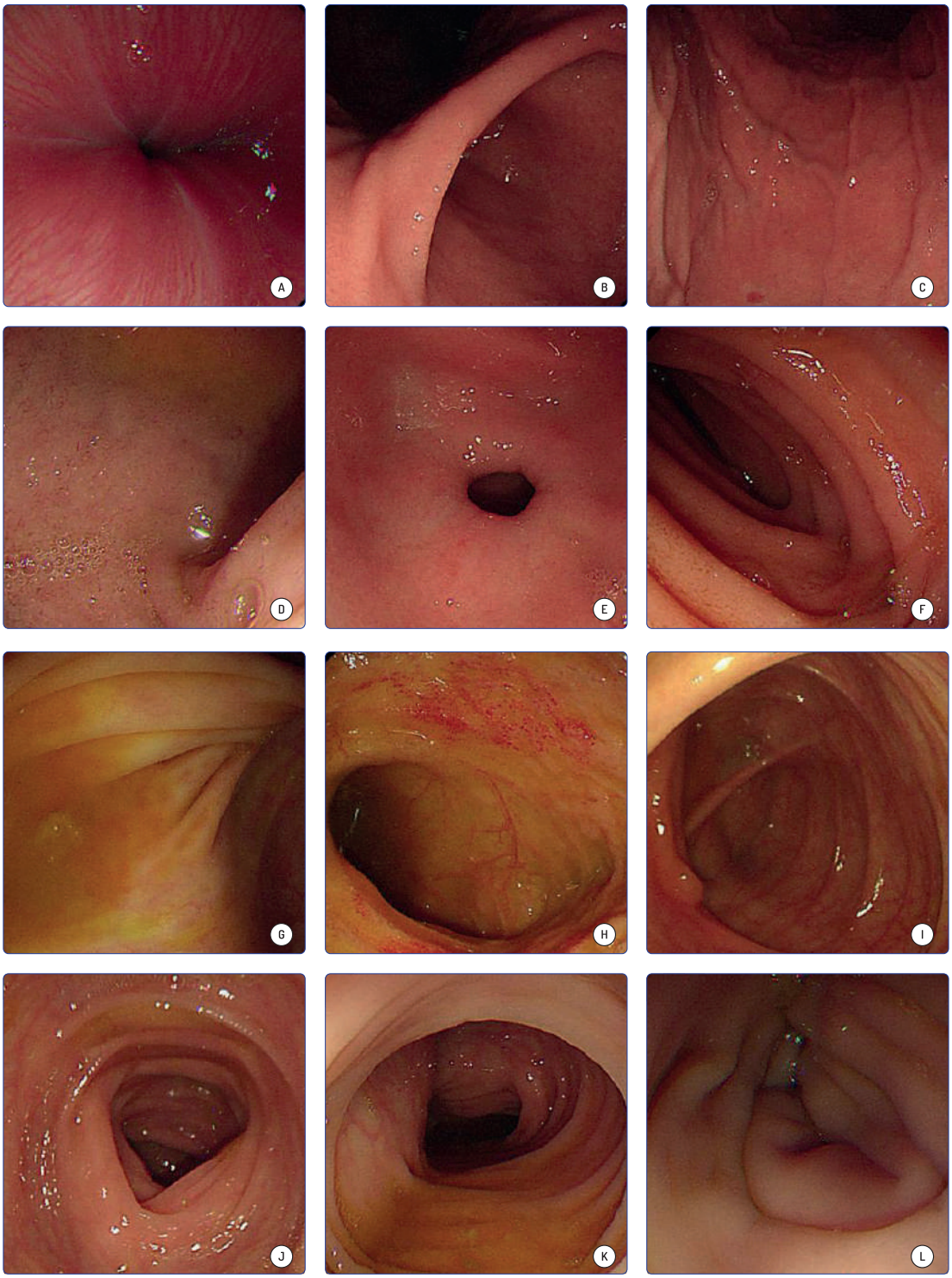

**Figure 3.** *capsule endoscopy*

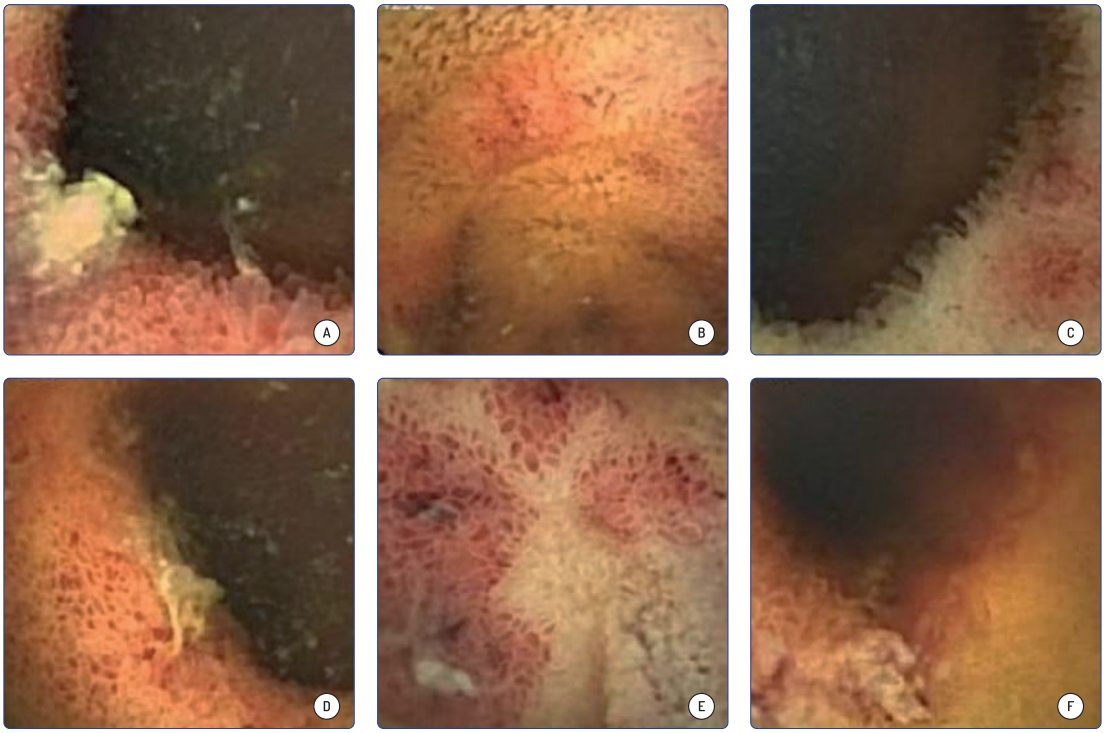


**Figure 4.** *Skin purpura like changes*

## Discussion

HSP also known as IgA vasculitis (IgAV), is an IgA-mediated systemic small-vessel vasculitis(1). HSP with the digestive tract as the initial symptom is also called abdominal HSP(1). It is prone to being misdiagnosed as various internal medicine diseases and surgical acute abdomen, and may even lead to unnecessary surgical treatment(2). Abdominal HSP can cause intestinal mucosal bleeding and necrosis, with clinical manifestations such as abdominal pain, vomiting, diarrhea, and bloody stools(3). It may also be complicated by intussusception, intestinal obstruction, intestinal perforation, and hemorrhagic enteritis. Especially when purpura occurs after or simultaneously with abdominal pain, it is difficult to distinguish from surgical acute abdomen. In this case, the patient's skin purpura appeared three weeks after the onset of abdominal pain, which brought difficulties to the diagnosis of the disease. According to the "Chinese Guidelines for the Diagnosis and Treatment of IgA Vasculitis in Children (2023)", abdominal ultrasound and abdominal X - ray are recommended for the diagnosis of severe digestive tract complications (intussusception, intestinal perforation) of IgAV. Routine gastrointestinal endoscopy (GPS) is not recommended for children with IgAV. Gastrointestinal endoscopy can be considered for children with IgAV or suspected IgAV who have severe abdominal pain or digestive tract bleeding (acute massive bleeding or recurrent digestive tract bleeding) and need further differential diagnosis. In this case, the patient had recurrent unrelieved abdominal pain and reddish - brown watery stools, so during the treatment process, the patient underwent gastroscopy, colonoscopy, and capsule endoscopy. At the same time, the UK Kidney Association's "The management of complications associated with IgA Vasculitis in children and young people" recommends that for children with intussusception in the abdomen, advice from surgical and radiological experts should be sought(4). Abdominal HSP can increase capillary permeability, leading to extensive edema, congestion, hematoma of the intestinal wall, enlargement of mesenteric lymph nodes, and gastrointestinal dysfunction, resulting in irregular intestinal peristalsis. The spasm caused by irregular intestinal peristalsis can lead to intussusception(5). If the invaginated intestinal tract is ischemic and paralyzed, the tension on the mesentery increases, and the blood circulation disorder is severe, which is likely to cause ischemic enteropathy and even intestinal necrosis. For adolescents and children with unexplained abdominal pain, the possibility of abdominal HSP should be considered during diagnosis(6), even if the skin purpura may not appear at the beginning.

## Reference


1. L. Xu, Y. Li, X. Wu, IgA vasculitis update: Epidemiology, pathogenesis, and biomarkers. Front Immunol 13, 921864 (2022).
2. B. V. Reamy, J. T. Servey, P. M. Williams, Henoch-Schonlein Purpura (IgA Vasculitis): Rapid Evidence Review. Am Fam Physician 102, 229-233 (2020).
3. H. Wang et al., Clinical outcome in pediatric refractory gastrointestinal Henoch-Schonlein purpura treated with mycophenolate mofetil. Eur J Pediatr 179, 1361-1366 (2020).
4. C. Day, R. Shute, UK Kidney Association guideline review: 'The initial management of IgA vasculitis (Henoch-Schonlein purpura) in children and young people' in conjunction with 'The management of complications-associated IgA vasculitis (Henoch-Schonlein purpura) in children and young people'. Arch Dis Child Educ Pract Ed 109, 196-201 (2024).
5. Y. Song et al., Pathogenesis of IgA Vasculitis: An Up-To-Date Review. Front Immunol 12, 771619 (2021).
6. A. Felix et al., Common Seasonal Pathogens and Epidemiology of Henoch-Schonlein Purpura Among Children. JAMA Netw Open 7, e245362 (2024).