Gastrointestinal Manifestations and Complications of Henoch-Schönlein Purpura

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Background: Henoch-Schönlein purpura (HSP) is a systemic vasculitic disorder commonly affecting young children. Gastrointestinal (GI) involvement is frequently seen, and it varies from mild symptoms to severe complications.

Methods: In the 5 years from July 1995 to June 2000, 208 children were diagnosed with HSP in our hospital. There were 116 males and 92 females with ages ranging from 9 months to 15 years (mean, 6.4 years; median, 5.5 years). Their medical records were reviewed. We focused on their gastrointestinal manifestations and complications. The hospital course and management of these patients were also analyzed.

Results: GI manifestations were present in 162 cases (77.8%), and among them the most common presentation was colicky abdominal pain in 159 cases (98.1%), followed by vomiting in 64 cases (39.5%). Severe complications included massive GI bleeding in 5 patients, intussusception in 1, protein-losing enteropathy in 1, and a patient with pancreatitis. GI symptoms occurred before the manifestation of skin lesions in 41 patients (25.3%). Five of them underwent a laparotomy. Four cases were operated on due to suspicion of acute appendicitis or peritonitis. Another patient had an emergent operation due to ileo-ileo intussusception.

Conclusions: GI manifestations of HSP are common. Colicky abdominal pain was the most common GI manifestation of Henoch-Schönlein purpura. Some patients may have GI complications, but those requiring surgical intervention in our study were rare.

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Key words: anaphylactoid purpura, Henoch-Schönlein purpura, children, gastrointestinal manifestations, complications.

Henoch-Schönlein purpura (HSP), a multisystemic vasculitic syndrome, manifests as purpuric skin lesions accompanied by such presentations as gastrointestinal (GI) symptoms, arthritis, and nephritis. This disease was recognized as a clinical entity by Lucas Schönlein in 1832 and Eduard Henoch in 1874. However, the etiology of HSP is still uncertain. Some factors such as infection, hypersensitivity, and drugs possibly contributing to this disease have been reported. Immunoglobulin A deposits in the dermis, mesangial area, small vessels, and submucosa of the GI tract result in multiple systemic involvement including dermal, renal, orthopedic, and gut systems. The GI tract is involved in

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50%-80% of patients reported in previous series, who manifested nausea, vomiting, diarrhea, colicky pain, and bleeding from the GI tract.[1] Complications of the GI tract such as massive bleeding, intestinal perforation or necrosis, and intussusception have also been reported.[2] The early detection of these complications is important because they may require surgical intervention, and some complications may result in morbidity and even mortality. Imaging studies such as roentgenography, ultrasonography, and computed tomography have been reported to be helpful in differentiating patients with and without surgical conditions, while laboratory data were not.[2] We reviewed the medical records of patients admitted to our children's hospital with HSP over a 5-year period and focused on symptoms and signs, laboratory studies, and clinical courses. Manifestations and complications of the GI tract were an important aspect we studied.

METHODS

Between July 1995 and June 2000, 208 children were admitted to our hospital with a final diagnosis of HSP. All children presented with a purpuric rash over the buttocks and lower extremities accompanied by 1 or more of the typical manifestations of the syndrome, including hematuria, arthritis, or abdominal symptoms. None of the patients had a bleeding tendency or coagulopathy.

We focused on their GI manifestations. Patients who had experienced abdominal pain, vomiting, diarrhea, upper GI bleeding manifested with coffee-ground vomiting or tarry stool passage, and lower GI bleeding presenting with hematochezia were classified as having GI involvement. Laboratory studies including total white blood cell count, hemoglobin level, and fecal and urinary analyses, and imaging reports were recorded. The results of upper and/or lower GI endoscopies of these patients were also reviewed. GI complications of these patients were defined as unusual presentations of the GI tract such as massive bleeding, bowel obstruction, and peritonitis. Attention was also paid to the operative findings of all of those children who underwent surgery.

The patients' hospital courses and management were analyzed. We compared the age, gender, and days of hospital stay between groups with and without GI manifestations using *t*-test and Chi-square test. The prognosis focused on the pain duration and complication rate of patients with corticosteroid treatment compared to those without such treatment. Finally, we also compared the complication rate of our series with those of past studies.

RESULTS

There were 116 boys and 92 girls with the classical presentation characterized by purpuric skin lesions. Ninety male and 72 female patients had GI symptoms, compared to 26 male and 20 female patients who did not. The average age was 6.4 years with a range of from 9 months to 15 years and a median age of 5.5 years. The mean age of patients with GI involvement was 6.3 years and without GI involvement was 6.5 years. The age distribution included 18 children younger than 3 years, 109 children from 3 to less than 6 years, 57 children from 6 to less than 9 years, 20 children from 9 to less than 12 years, and 4 children from 12 to 15 years. The mean hospital stay of patients with and those without GI involvement was 5.1 and 3.2 days, respectively.

GI manifestations appeared in 162 children (77.8%) (Table 1). Among these 162 patients, 159 cases (98.1%) presented with colicky abdominal pain, and 64 (39.5%) presented with vomiting. Thirty-five patients (21.6%) was found to have gastrointestinal tract bleeding, including 28 episodes (17.3%) of upper gastrointestinal tract bleeding, 9 episodes (5.6%) of lower gastrointestinal tract bleeding, and 2 patients with both upper and lower gastrointestinal tract bleeding. Eleven children (6.8%) had diarrhea.

As to the location of abdominal pain, it most commonly involved the epigastric area in 55 cases (34.6%), and 52 patients manifested periumbilical pain. Forty patients had poorly localized pain.

Table 1. Gastrointestinal Manifestations of HSP in 162 Children

Clinical features	No. of patients	Percent (%)
Colicky abdominal pain	159	98.1
Vomiting	64	39.5
GI bleeding	35	21.6
UGI bleeding	28	17.3
LGI bleeding	9	5.6
Diarrhea	11	6.8

Abbreviations: HSP: Henoch-Schönlein purpura; UGI: upper gastrointestinal; LGI: lower gastrointestinal.[8]

Other uncommon locations included the right lower quadrant, left lower quadrant, and mid-lower abdomen.

GI symptoms occurred before the manifestation of skin lesions in 41 patients (25.3%). The average interval between the 2 events in those patients was 4.8 days. Eighteen patients had abdominal pain which appeared more than 1 week before the skin manifestation.

Some patients had an abnormal hemogram: leukocytosis (> 10,000/mm^3) in 77 patients (47.5%), anemia (Hb < 10 g/dl) in 17 patients (10.5%), and thrombocytosis (> 400,000/mm^3) in 33 patients (20.4%). Twenty-three children were found to have hematuria, and 8 children had proteinuria among the 159 urinary analyses carried out. A stool guaiac test was performed in 160 cases, and obvious occult blood (more than 2+) was found in 52 cases.

Eighty-two patients with acute abdominal pain underwent plain abdominal roentgenography. Their findings were dilated bowel pattern in 24 instances, increased fecal retention in 18 patients, and non-specific findings in 40 patients. Only 8 patients underwent roentgenographic contrast studies, including 5 upper GI with small bowel series and 3 lower GI series. Upper GI with small bowel series contrast studies found thickening of the intestinal fold appearing like a thumbprint in 2 patients (Fig. 1). No specific findings were seen in the lower GI series contrast study. Abdominal sonography was performed in 77 children. It revealed ascites in 19 patients, bowel wall edema in 17, mesenteric lymph node enlargement in 9, hepatomegaly in 8, splenomegaly in 3, and gall bladder sludge in 2 patients. Twenty-seven children underwent upper GI endoscopy, with positive findings in 15 cases. Erythematous hemorrhagic gastritis was found in 9 patients (Fig. 2). Duodenal erosion with erythema was found in 3 cases. A purpuric duodenum lesion was found in 1 case. Colonoscopy was performed in 5 children with lower gastrointestinal bleeding, but no specific findings were seen in those cases.

One hundred twenty-four patients presenting with abdominal pain were treated with prednisolone (1 mg/kg/day) for a duration of from 1 week to 1 month. Most of them (89 patients, 71.8%) improved within 48 hours. For those patients managed with corticosteroids, intractable colicky abdominal pain was the most common indication. The average duration of abdominal pain of patients treated with corticosteroids was 7.5±2.8 days, and that of patients without corticosteroid treatment was 10.2±3.6 days.

Some GI complications were observed in our patients. Five children needed a blood transfusion due to massive blood loss from the GI tract. A 6-year-old boy underwent an emergent operation due to bowel obstruction, and the operative finding was ileo-ileo intussusception and multiple lymphadenopathy. His purpuric skin rashes appeared 13 days later than the presentation of abdominal pain.

We also encountered some uncommon complications of HSP. A 7-year-old boy experienced severe

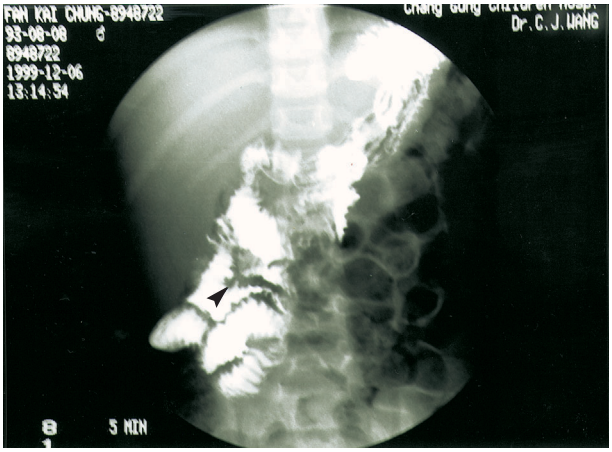

Fig. 1 Upper GI and small bowel series contrast study showing thickening of the intestinal fold and bowel separation with a thumbprint appearance (arrowhead).

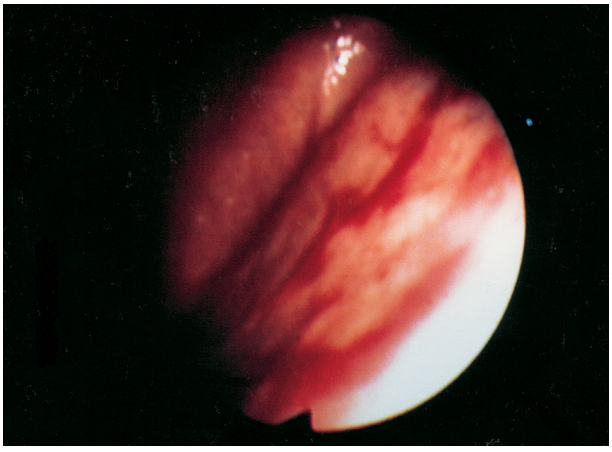

Fig. 2 Upper GI endoscopy showing erythematous hemorrhagic change in the body of the stomach.

epigastralgia with highly elevated serum amylase (384 U/L) and lipase (330 U/L) levels; he presented with dermatologic manifestation 5 days later. Another 6-year-old male suffered from hypoalbuminemia (2.9 g/dl) probably due to protein-losing enteropathy. These 2 patients were successfully managed with conservative treatment. Overall, we had a complication rate of 4.8% (8/162). All patients with complications, but 1, were treated with corticosteroids.

Four children were operated on due to suspicion of acute appendicitis and peritonitis. All of these patients presented with severe and atypical manifestations of HSP, and their skin lesions appeared many days after the GI symptoms. No specific diagnosis was made at the time of the laparotomy. HSP was diagnosed after the appearance of a skin rash.

In the follow-up period of 3 months, recurrence of abdominal pain was found in 21 children, with 1 episode in 17 cases, 2 episodes in 2 cases, and 3 episodes in 2 cases. The interval between the first episode and recurrence ranged from 12 to 44 days, with a mean time of 19.3 days. No patient had a complication which occurred in the course of recurrence. All of these patients with recurrence except 1 were treated with corticosteroids.

DISCUSSION

HSP is a systemic disorder caused by cutaneous and small-vessel vasculitis of unclear etiology characterized by a symmetrical, non-traumatic, non-thrombocytopenic, painless, purpuric rash mostly involving the buttocks or legs, as well as nephritis, arthritis, and GI manifestations. HSP manifests as a systemic illness in about 80% of patients. In our series, there were no differences in the mean age or gender preponderance in patients with and without GI involvement, but the group with GI manifestations required a longer hospitalization (5.1±2.5 vs. 3.2±2.1 days) (Table 2). As in most series,[2-4] our patients were predominantly male.

According to past studies, the GI tract is involved in 50% to 80% of patients presenting with symptoms such as nausea, vomiting, diarrhea, colicky pain, melena, and hematochezia.[1-4] Similarly in our study, GI manifestations appeared in 77.8% of children with HSP. Abdominal pain was the most frequent symptom in our patients (159 of 208, 76.4%). The incidence was higher than in previous series which showed 35%-60% of patients with abdominal pain, which could be diffuse and paroxysmal.[5] In contrast to a previous report[2] which showed that abdominal pain was usually poorly localized, our patients seemed to have pain involving the periumbilical and epigastric areas of the abdomen.

Table 2. Clinical Outcome of our 208 HSP Patients with or without GI Manifestations

	GI (+) (N=162)	GI (-) (N=46)	*p**
Mean age (yr)	6.3±2.1	6.5±2.4	0.5754[a]
Male: Female	90 : 72	26 : 20	0.5474[b]
Mean hospital stay (day)	5.1±2.5	3.2±1.8	0.0127[a]

Abbreviations: HSP: Henoch-Schönlein purpura.
Data are presented as the mean±SD.
GI (+), HSP patients with gastrointestinal manifestations.
GI (-), HSP patients without gastrointestinal manifestations.
[a] Analyzed by *t*-test.
[b] Analyzed by the Chi-square method.
* Significant at $p<0.05$.

The occurrence of severe abdominal pain prior to development of the typical purpuric skin lesions has often posed a diagnostic challenge to physicians.[1,3,6] Extracutaneous signs may precede the rash, sometimes by as long as 28 days.[7] David et al. also reported that 10%-40% of patients showed GI symptoms before manifestation of skin lesions.[2] In our study, GI symptoms occurred before the manifestation of skin lesions in 25.3% of patients.

In our study, plain roentgenography of the abdomen played little role in the diagnosis. This is similar to a previous report in which radiologic findings in HSP of the bowel were nonspecific.[6] Barium studies in patients with HSP might show filling defects as pseudotumors.[6] Two of our 8 patients who underwent GI contrast studies had such a finding. Focal submucosal hemorrhage resulting in mucosal swelling may have served as a lead point for an intussusception in a previous study.[8]

Some have reported that sonography of the intestine represents the most sensitive method for the detection of intramural bleeding in HSP and may reduce diagnostic radiation in children. The findings consisted of circumscribed hypoechoic to non-echoic "half-moon" thickened intestinal walls and cockade

phenomenon outlining intramural bleeding and edema.[9] Another advantage of ultrasound was early detection of intussusception in patients with HSP.[10] We had a patient complicated with intussusception and another complicated with pancreatitis, but their sonographic findings were non-specific. However, ultrasound can help in excluding acute abdomen and preventing unnecessary surgical intervention especially when the purpuric skin lesions appear after the GI symptoms. One study reported that sonography was helpful in follow-up examinations of HSP patients.[9] Serial ultrasonography showed a progressive decrease in mucosal wall thickening, the reappearance of peristalsis, and visualization of valvulae conniventes.[5]

GI endoscopy is indicated in the case of hematemesis, melena, and epigastric pain, irrespective of the presence of skin lesions.[1] In a past report by Tomomosa, mucosal lesions were noted in 6 of 9 patients by upper GI endoscopy. The most prominent finding was severe hemorrhagic erosive duodenitis. It may be regarded as a typical but non-pathognomonic endoscopic finding of HSP.[11] Also in Kato's series, the predominant changes in the second part of the duodenum were characteristic of HSP.[12,13] In a previous analysis of endoscopic findings of HSP children in Taiwan, hemorrhagic erosive duodenitis and gastritis were the most common findings.[14] In our study of panendoscopy in HSP patients, multiple patchy erythema with hemorrhagic-erosive gastritis was found in 9 (60%) of 15 patients. Erythematous erosive duodenitis was visualized in 3 (20%) of those patients.

GI symptoms will disappear spontaneously in a short time. Treatment with corticosteroids may have a role in hastening the resolution of pain.[15] We had similar observations.

The complication rate of the GI tract in HSP reportedly varies from 3.9% to 22.4%.[4] It occurred in 8 (4.8%) of our patients. This is relatively low compared to past studies (Table 3). Early awareness of GI symptoms and their evaluation might have been the reason for the lower rate.

Intussusception is a common surgical complication in patients with HSP. In a previous review, 2/3 involved only the small bowel and 1/2 of the patients had a palpable mass.[16] More than 1/2 of those intussusceptions were of the ileo-ileo type.[10] Lower GI barium reduction is usually not successful for intussusception in patients with HSP. One of our patients was complicated with ileo-ileo intussusception. He was successfully treated using surgical reduction. Patients with massive GI hemorrhage associated with HSP will occasionally require surgical management,[17,18] and up to 14% of them have significant bleeding that requires a blood transfusion.[19] Among children with complications in our series, 3.1% needed a blood transfusion, and none of them underwent surgical management. Other rare complications such as ileal perforation[20] or ileal stricture presenting with chronic intestinal obstruction,[21] and protein-losing enteropathy[22] due to protein loss from the gastrointestinal tract have also been documented to be associated with HSP. There was 1 patient complicated with protein-losing enteropathy in our study. It is interesting to note that we had a patient complicated with acute pancreatitis.

Table 3. Gastrointestinal Complication Rates of HSP in Children Reported in the Literature and Our Study

	No. of cases	No. of complications	Percent
Cull (1982)	183	8	4.4
Harvey (1984)	105	4	3.9
Martinez (1984)	58	13	22.4
Mir (1988)	110	7	6.4
Schmuel (1991)	110	9	8.2
Our study	162	8	4.8

Abbreviation: HSP: Henoch-Schönlein purpura.

HSP might be misinterpreted as surgical abdomen if the skin rash appears later than GI symptoms. Cream et al. stated that 5 of 9 HSP patients who underwent an exploratory laparotomy presented with abdominal pain prior to the development of other signs and symptoms.[22] Similarly, we had 4 patients suspected of acute appendicitis and peritonitis who underwent an operation before the purpuric rash appeared. As was stated by Allen et al.,[3] early recognition and proper management may reduce this hazard. Ultrasonographic[9] and endoscopic[11] findings may help exclude surgical abdomen and establish a correct diagnosis even in the absence of typical skin lesions.

In conclusion, most GI manifestations and complications of patients with HSP are self-limited or can usually be conservatively managed with good success; an operation is rarely needed. HSP should

be considered in children experiencing cramping abdominal pain even without skin lesions at the initial presentation.

REFERENCES

1. Lippl F, Huber W, Werner M, Nekarda H, Berger H, Weigert N. Life-threatening gastrointestinal bleeding due to a jejunal lesion of Henoch-Schönlein purpura. Endoscopy 2001;33:811-3.
2. Cull DL, Rosario V, Lally KP, Irving R, Cull L, Mahour GH. Surgical implications of Henoch-Schönlein purpura. J Pediatr Surgery 1990;25:741-3.
3. Allen DM, Diamond LK, Howell DA. Anaphylactoid purpura in children (Schönlein-Henoch syndrome): Review with a follow-up of the renal complications. Am J Dis Child 1960;99:147-68.
4. Katz S, Borst M, Seekri I, Grosfeld JL. Surgical evaluation of Henoch -Schönlein purpura- experience with 110 children. Arch Surg 1991:126:849-54.
5. Couture A, Veyrac C, Baud C, Galifer RB, Armelin I. Evaluation of abdominal pain in Henoch-Schönlein syndrome by high frequency ultrasound. Pediatr Radiol 1992; 22:12-7.
6. Goldman LP, Lindenberg RL. Henoch-Schönlein purpura: gastrointestinal manifestations with endoscopic correlation. Am J Gastroenterol 1981;75:357-60.
7. Bhaskar B, Salmon R, Edward S, James M. Endoscopic findings in Henoch-Schönlein purpura. Gastrointestinal Endoscopy 1991;37:569-71.
8. Clark JH, Fitzgerald JF. Hemorrhagic Complications of Henoch-Schönlein syndrome. J Pediatr Gastroenterol Nutr 1985;4:311-5.
9. Bomelburg T, Cleasen U, VonLengerke HJ. Intestinal ultrasonographic findings in Schönlein-Henoch syndrome. Eur J Pediatr 1991;150:158-60.
10. Choong CK, Kimble RM, Pease P, Beasley SW. Colocolic intussusception in Henoch-Schönlein purpura. Pediatr Surg Int 1998;14:173-4.
11. Tomomasa T, Hsu JY, Itoh K, Kuroume T. Endoscopic findings in pediatric patients with Henoch-Schönlein purpura and gastrointestinal symptoms. J Pediatr Gastroenterol Nutr 1987;6:725-9.
12. Kato S, Ebina K, Naganuma H, Sato SI, Maisawa SI, Nakagawa H. Intestinal IgA deposition in Henoch - Schönlein purpura with severe gastro-intestinal manifestations. Eur J Pediatr 1996;155:91-5.
13. Kato S, Shibuya H, Naganuma H, Nakagawa H. Gastrointestinal endoscopy in Henoch-Schönlein purpura. Eur J Pediatr 1992;151:482-4.
14. Ting CW, Wu TC, Cheng JH, Hwang B. Gastrointestinal manifestations and endoscopic findings in children with Henoch-Schönlein purpura. Acta Pediatr Sin 1988; 29: 303-8.
15. Rosenblum ND, Winter HS. Steroid effects on the course of abdominal pain in children with Henoch-Schönlein purpura. Pediatrics 1987;79:1018-21.
16. Marnitez FL, Haase GM, Ernster JA, Bailey WC. Surgical complications in Henoch-Schönlein purpura. J Pediatr Surg 1984;19:434-6.
17. Goldbloom RB, Drummond KN. Anaphylactoid purpura with massive gastrointestinal hemorrhage and glomerulonephritis. Am J Dis Child 1968;116:97-102.
18. Weber TR, Grosfeld JL, Bergstein J. Massive gastric hemorrhage: an unusual complication of Henoch-Schönlein purpura. J Pediatr Surg 1983;18:576-8.
19. Feldt RH, Stickler GB. The gastrointestinal manifestations of anaphylactoid purpura in children. Mayo Clin Proc 1962;37:465-83.
20. Chao HC, Huang JL. Ileal perforation in Henoch-Schönlein purpura: report of one case. Acta Paediatr Sin 1996;37:455-7.
21. Lombard KA, Shah PC, Thrasher TV, Grill BB. Ileal stricture as a late complication of Henoch-Schönlein purpura. Pediatrics 1986;77:396-8.
22. Cream JJ, Gumpel JM, Peachey RDG. Schönlein-Henoch purpura in adults. Q J Med 1970;39:461-83.

類過敏性紫斑症之腸胃道表徵及其併發症

陳世彥 江文山

背 景： 類過敏性紫斑症是一種較常見於兒童的全身性血管炎疾患。胃腸道的侵犯常可見，而且影響從輕微的症狀到嚴重的併發症皆有可能。

方 法： 過去5年（從1995年7月至2000年6月），在我們醫院208個病童診斷是類過敏性紫斑症，一共有116個男性和92個女性，年紀最小的9個月大，最大的15歲，平均年齡爲6.4歲，我們回顧他們的醫療記錄，並且將重點放在他們的胃腸道表徵以及併發症。對於他們的住院過程及治療也同時作分析。

結 果： 162個案例有胃腸道表徵，這些表現之中又以腹部絞痛最多，有159案例 (佔98.1%)；其次是嘔吐有64案例（佔39.5%），嚴重併發症包括大量胃腸道出血有5例；腸套疊有1例；蛋白流失性腸症（protein losing enteropathy) 有1例；有一例爲胰臟炎。41個案例其胃腸道表現發生時間比皮膚病灶爲早 (佔25.3%)，其中有5例接受腹部手術。包括四例因爲懷疑急性盲腸炎或腹膜炎而開刀。另一個病人因爲迴腸套迴腸的腸套疊 (ileo-ileo intussusception）。

結 論： 胃腸道表現常在類過敏性紫斑症出現。腹部絞痛是最常見的胃腸道表徵，有時會引起併發症，在我們的研究其併發症很少需要手術處理。

(長庚醫誌 2004;27:175-81)

關鍵字： 類過敏性紫斑症，兒童，胃腸道表徵，併發症。

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