

# A review of case and case series reports on Henoch–Schönlein syndrome-related pancreatitis

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To summarize the experience of diagnosing and treating patients with Henoch–Schönlein purpura (HSP)-related pancreatitis, a systematic review of previously published cases was conducted. Among 13 reported cases, there were six males and seven females whose age from 3 to 70 years. The clinical features of these patients indicated that acute pancreatitis could be the initial manifestation of HSP, the radiological change was atypical, and most cases were alleviated with steroidal treatment. Good outcomes can be achieved in patients who are diagnosed early with HSP-related pancreatitis, and it is vital to begin timely treatment of HSP-related pancreatitis with corticosteroid.

**Key words:** Acute pancreatitis, corticosteroids, Henoch–Schönlein purpura

**How to cite this article:** Xiong F, Tao Y, Li H. A review of case and case series reports on Henoch–Schönlein syndrome-related pancreatitis. *J Res Med Sci* 2017;22:43.

## INTRODUCTION

Henoch–Schönlein purpura (HSP) belongs to the group of nongranulomatous small vessel vasculitis. Although HSP can occur at any age, it is overwhelmingly a childhood disease.<sup>[1,2]</sup> Approximately, two-thirds of children with HSP develop abdominal pain. Intussusception, gangrene of the bowel, bowel perforation, and massive hemorrhage are the most common gastrointestinal complications of HSP. Most rarely, life-threatening HSP-related pancreatitis may occur. Since 1965, few cases of HSP-related pancreatitis have been reported.<sup>[3]</sup> A comprehensive search of PubMed, EMBASE, and Web of Science was performed for all relevant papers published before July 1, 2015. Because the full text of six articles was unavailable, only 13 cases described in 12 full-text articles were included in this study.<sup>[4-15]</sup> Then, we investigated the clinical features, treatments, and prognoses of HSP-related pancreatitis cases.

## CASE REPORT

The main clinical characteristics of the 13 patients (six males, seven females) are summarized in Table 1. The patients ranged in age from 3 to 70 years, half of cases were 5–20 years old. Acute pancreatitis was found in the active stage of HSP. Pancreatitis presented as the initial manifestation of HSP in eight cases. In addition, pancreatitis and typical purpura occurred at the same time in the two cases. From the onset of pancreatitis to the diagnosis of HSP, the time elapsed ranged from 1 day to 75 days, and 62.5% (5/8) of the patients began to have typical purpura within 7 days of the onset of pancreatitis. In addition to abdominal pain in all cases, seven patients presented with vomiting, one patient had hematemesis, one patient had hematochezia, and one patient had poor appetite. Six patients (46.2%) had moderate fever. Renal involvement was reported in five cases. Among the 13 cases, one patient was misdiagnosed with acute appendicitis and underwent surgery. Laboratory examinations revealed different

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DOI:

10.4103/jrms.JRMS\_658\_15

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**Received:** 08-08-2015; **Revised:** 30-12-2015; **Accepted:** 17-01-2017

**Table 1: Main clinical characteristics of 12 previously reported cases of Henoch-Schönlein purpura-related pancreatitis**

	Authors												
	Nie et al. <sup>[4]</sup>	Frigui et al. <sup>[5]</sup>	Dinler et al. <sup>[6]</sup>	Nakayama et al. <sup>[7]</sup>	Soyer et al. <sup>[8]</sup>	Sato et al. <sup>[9]</sup>	Sun et al. <sup>[10]</sup>	Cheung et al. <sup>[11]</sup>	Lévy-Weil et al. <sup>[12]</sup>	Lévy-Weil et al. <sup>[12]</sup>	Takamatsu K et al. <sup>[13]</sup>	Garner <sup>[14]</sup>	Diaz <sup>[15]</sup>
Gender	Female	Male	Female	Female	Female	Male	Male	Male	Female	Male	Male	Female	Female
Age (years)	15	53	12	11	3	70	11	7	33	30	51	7	18
Pancreatitis as initial manifestation	No	Yes	Yes	Yes	Yes	Yes	No	Yes	Yes	No	No	Yes	No
Time between pancreatitis and purpura (days)	20	1	19	7	5	75	22	11	7	0	0	2	1
Digestive symptoms	Abdominal pain	Abdominal pain, vomiting	Abdominal pain, vomiting	Abdominal pain, hematemesis	Abdominal pain, vomiting	Abdominal pain, poor appetite	Abdominal pain	Abdominal pain, hematochezia	Abdominal pain, vomiting	Abdominal pain, vomiting	Abdominal pain	Abdominal pain, vomiting	Abdominal pain, vomiting
Fever	Yes	Yes	No	No	No	No	Yes	No	Yes	Yes	No	No	Yes
Serum amylase (IU/L)	149	349	349	230	128	363	198.6	1164	116	87	460	4700	-
Urine amylase (IU/L)	941	476	-	1372	238	-	1193.2	-	1744	380	-	-	-
Kidney involvement	Yes	Yes	-	-	-	-	-	-	-	Yes	Yes	-	Yes
Abdominal CT or ultrasound imaging	Pancreas and peripancreatic swelling, ascites	Pancreas swelling	Pancreatic cyst	Pancreas swelling	Normal	Pancreatic swelling	Normal	Pancreatic swelling, with focal necrosis, ascites, pancreatic pseudocyst	Normal	Normal	Pancreatic hemorrhage, necrosis	-	Normal
The time elapsed until abdominal pain was relieved	7	7	2	-	5	44	-	-	-	-	-	1	-
Outcome of pancreatitis	Improvement	Improvement, no recurrence in a year	Improvement, no recurrence in 9 months	Improvement	Improvement	Improvement, cured in 136 days	Improvement	Improvement	Improvement	Improvement	Death	Improvement	Improvement

CT=Computing tomography

levels of increased serum amylase (87–1164 IU/L) and urine amylase (238–1744 IU/L) in all cases. Abdominal computed tomography (CT) revealed pancreatic edema, ascites, and a wide range of intestinal wall edema in seven cases. A pancreatic cyst was found in one patient, it appeared over the course of 35 days and disappeared in 55 days; however, five patients did not demonstrate any morphological changes in the pancreas. All patients experienced relief through treatment with fasting, gastrointestinal decompression, nutritional support, antacid drug, glucocorticoid, and somatostatin. The abdominal pain relief time (1–44 days) was noted in six cases. Patient prognoses were described in three cases, and these patients were cured without recurrence through follow-up. One patient died from serious acute hemorrhagic pancreatitis.

## DISCUSSION

HSP occurs about twice as often in boys as in girls, half of affected patients are younger than 10 years of age. However, our study suggested that HSP-related pancreatitis usually occurs in adolescent girls. The reason remains unclear; it may be associated with the immune state of adolescent girls.

HSP-related pancreatitis is uncommon. The reason for the low incidence is unclear. Compared with other types of pancreatitis, HSP-related pancreatitis has the following characteristics. In our study, before the onset of HSP-related pancreatitis, there was no prominent cause, such as biliary tract disease, overeating, drinking, hyperlipidemia, viral infection, or drugs. Thus, the pathogenesis of acute pancreatitis remains unclear. We speculate that HSP-related pancreatitis may be associated with small blood vessel thrombosis, vasculitis, and intimal thickening. Although the clinical manifestation was relatively mild,<sup>[16]</sup> acute pancreatitis may be the initial manifestation of HSP. HSP-related pancreatitis is diagnosed clinically but requires CT evaluation or ultrasound imaging to differentiate mild acute pancreatitis from severe necrotic pancreatitis.<sup>[17]</sup> In our study, approximately 41.7% (5/12) of the patients with pancreatic morphology were normal, and only 41.7% of the patients presented with pancreas swelling. The imaging changes in HSP-related pancreatitis were atypical, and pancreatic necrosis or pseudocysts and other local complications were relatively rare. In our study, all patients had elevated serum amylase and urine amylase levels, with an increase of at least three times the upper limits in blood and urine, but the level was not positively correlated with the disease severity. Some researchers believe that elevated amylase creatinine clearance and serum lipase levels are appropriate for the early diagnosis of pancreatitis.<sup>[11]</sup> It has been reported that the measurement of plasma factor XIII could be useful for the early diagnosis of HSP, particularly when the typical

purpura is preceded by abdominal pain.<sup>[16]</sup> In addition to the conventional treatment for pancreatitis, more attention should be paid to HSP. In our study, the symptom of pancreatitis was improved after steroid treatment in all patients. Somatostatin can inhibit gastric and pancreatic secretions, thereby reducing enzymatic activity; it can also reduce capillary permeability, open the sphincter of Oddi, and promote the excretion of pancreatic enzyme. After a diagnosis of HSP-related pancreatitis, somatostatin may be applied as soon as possible.

## CONCLUSION

HSP-related pancreatitis is a special type of pancreatitis that is relatively rare. Pancreatitis may reflect the disease activity or severity of manifestations of HSP. HSP-related pancreatitis should be considered when abdominal pain occurs in HSP patients. In addition to the symptomatic and supportive treatment, the use of corticosteroids to control HSP helps to alleviate HSP-related pancreatitis.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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