Henoch Schonlein Purpura in children: clinical analysis of 120 cases

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Abstract

Background: Henoch Schonlein Purpura (HSP) is a systemic vasculitic disease which is common in children. It is very important to understand the clinical features of this disease for doctors and nurses.

Objectives: To study the clinical characteristics of HSP in children.

Methods: Collect the clinical data of the HSP children, and analyze the clinical characteristics of these HSP patients.

Results: The ratio of M:F was 1.9:1. The mean age was 6.6 ± 1.6 years. The typical onset seasons were spring, winter and autumn. Infection and food allergy were the main etiological factors. The first symptom was skin purpura and these purpura mainly concentrated the lower extremities and buttocks. The dominant digestive clinical features were abdominal pains and vomiting. The knee joint and ankle joint were most frequently affected. The typical kidney symptoms were microscopic hematuria and albuminuria. An increased ESR was reported in 68 patients (56.7%). Serum C3 decreased in 13 cases (10.8%). ASO titer was higher in 57 children (47.5%).

Conclusion: There were gender, season and area differences for the HSP patients. The etiological factors were diverse. HSP patients could have various clinical symptoms and rare complications.

Key words: Henoch Schonlein Purpura, Children

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Introduction

Henoch Schonlein Purpura(HSP), was first recognized in 1801 by Heberden and first described as an association arthritis by Schonlein in 1837.[1] It is a systematic vasculitic disease, and mainly affects the small vessels of skin, joints, gastrointestinal tract, and kidneys. In some series, boys are affected more than girls.[2] The mean age of HSP patients is 6 years; 75% of the patients are under 8 years of age and 90% are less than 10 years of age.[3-5] Although many antigens, such as foods, infective agents, drugs, vaccinations, and insect bites have been reported to be related to HSP, the etiology of this disease remains unclear.[6-8] The pathogenesis also remains unknown. At present, HSP is regarded as an inflammation and immune-mediated disease.[9] IgA and some proinflammatory cytokines have the pivotal role in the pathogenesis of HSP.[10]

The typical clinical characteristics involve the triad of palpable purpura, abdominal pain and arthritis. Progressive renal function impairment, bowel perforation, central nerve system involvement are rare. The occurrence of purpura, which are non-thrombocytopenic, is the essential element for the diagnosis of HSP. The purpura are often located on some parts just as lower extremities and buttocks.[10] HSP, in general, is considered benign and self-limited and the treatment is supportive.

The aim of our study is to analyze the clinical data of the HSP children, and discuss the clinical characteristics of HSP in children. We think knowing about the clinical features could help doctors to make the correct diagnosis and provide the correct remedy and nursing.

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Methods

This is a retrospective study. The patients were HSP children treated from the Qilu Hospital, between 2007 and 2010. We selected the patients who accepted the whole remedy in the pediatric bed-ward of Qilu Hospital. Those patients who left the hospital

before improvement and recovery were excluded form our study. The diagnosis criteria were the American College of Rheumatology 1990 criteria for the classification of HSP[11]. Approval for this research was obtained from the Medical Ethic Committee of the Qilu Hospital.

Definition

The epidemiological data were analyzed: City means the settlements where the non-agricultural industries and non-agricultural population concentrate in. Countryside means the area where the agricultural production laborers inhabited in. Spring includes March, April and May. Summer means June, July and August. Autumn refers to September, October and November. Winter means December, January and February. Previous infection, special food, vaccination, insect bite, and accident were considered as the precipitating factors to be specially retrospectively looked, if they happened in 2 weeks before HSP onset. Preceding infection was self reported.

About the clinical characteristics, we defined as follows: Microscopic hematuria wasdefined when the test result was > +; Albuminuria was defined when the test result was > +; Gross hematuria was defined blood in the urine could be seen with the naked eye; Tubular urine was defined one more urinary cylinder were found per low power field. Rash location means purpura mainly concentrated in the parts of the body.

The laboratory data were defined: Elevated ESR was defined when ESR was >20mm/1hour; increased ASO titer when>300IU/mm; low C3 when <900mg/L.

Statistical analysis

All data were described as means and standard deviations (mean ± SD) or medians and ranges. Categorical variables were expressed by percentages.

Results

Epidemiological features

The main epidemiological data are shown in table 1. There were 120 children fulfilling the diagnosis criteria between January 2007 to December 2010. The 120 HSP children included 78(65%) boys and 42(35%) girls. Males were affected more than the females (Ratio M:F=1.9:1). The age range was from 1 to 12 years (mean age 6.6 ± 1.6 years). The age and gender distribution showed that most HSP children (103 cases, 85.3%) were less than 8 years old, 88.3% were <10 years old, and 98 cases of HSP children (81.7%) were between 5 and 8 years old (figure 1). It was found that 40 cases (33.3%) occurred in spring, 12 cases (10%) in summer, 24 cases (20%) in autumn, and 44 cases (36.7%) in winter. The countryside patients were 73 cases (60.8%), while there were 47 city children (39.2%). For 98 cases, this was their primary presentation. There were 22 relapse patients.

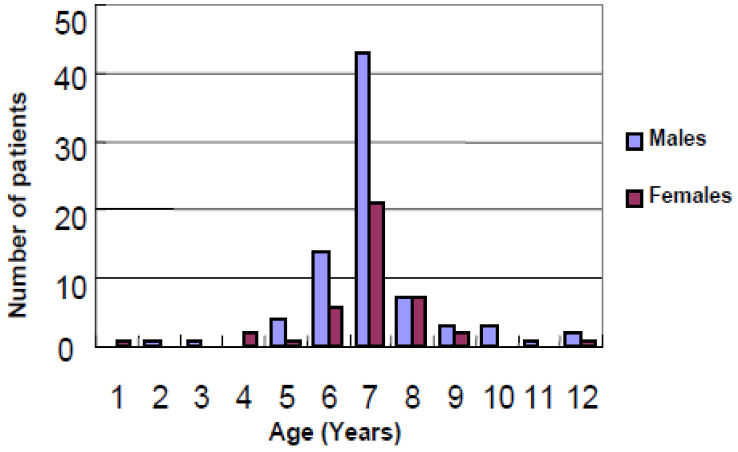

Figure 1: Age and gender distribution of 120 HSP patients

The age range was from 1 to 12 years (mean age 6.6 ± 1.6 years). The age and gender distribution showed that most HSP children (98cases, 81.7%) were less than 8 years old, and 103 cases of the HSP children (85.3%) were part of the age range between 5 and 8 years old (figure 1).

Before HSP onset, there were 73 cases (60.8%) who had an infection, and among these patients, 52 children had an upper respiratory tract infection; 17 cases had bronchial pneumonia; 4 patients had enteritis. The results revealed 32 patients had eaten special food just as fish or shrimp before onset, and there were 2 children who had been given a measles-mumps-rubella vaccination 2 weeks before HSP onset, and 1 child who had been accepted a Hepatitis B vaccine 1 weeks before HSP coming. Parasite infection was reported in 3 patients (2.5%), and among these children, ascarids attacked 1 child, and the other two had been infected by threadworms. Tick bites were announced in 2 cases one week before disease onset. Two children had choked with water seriously during swimming before. There were unknown etiological factors in four children before disease onset.

Table 1: Epidemiological data in 120 children with HSP

	Children (*n* = 120)	Percentage (%)
Age(years)		
Mean ± SD	6.6 ± 1.6	
Range	1-12	
Sex: Male/Female	78/42	
Proportion of boys		65
Ratio M:F	1.9:1	
Possible etiological factors		
Infection	73	60.8
Food allergy	32	26.7
Vaccination	3	2.5
Parasite infection	4	3.3
Tick bite	2	1.6
Emergencies	2	1.6
Unknown	4	3.3
Seasonal pattern		
Spring	40	33.3
Summer	12	10.0
Autumn	24	20.0
Winter	44	36.7
Area distribution		
Countryside	73	60.8
City	47	39.2
Situation of disease onset		
First	98	81.7
Recurrence	22	18.3

City means the settlements where the non-agricultural industries and non-agricultural population concentrate in. Countryside means the area where the agricultural production laborers inhabited in period. Spring are March, April and May. Summer are June, July and August. Autumn are September, October and November.. Winter are December, January and February.

Table 2: Main clinical features of the 120 children with HSP

Symptom	Children (*n* = 120)	Percentage (%)
First symptom		
Skin purpura alone	83	69.1
Abdominal pains	14	11.7
Joint pains	1	0.8
Purpura plus joint pains	11	9.2
Purpura plus abdominal pains	9	7.5
Purpura plus joint pains plus abdominal pains	2	1.7
Rash location		
LE plus buttocks	116	96.7
LE plus buttocks plus UE	36	30.0
LE plus buttocks plus trunk	6	5.0
LE plus buttocks plus hands and feet	6	5.0
LE plus buttocks plus face	5	4.2
LE plus buttocks plus vulva	2	1.7
Digestive tract symptom	**89**	**74.2**
Abdominal pains	68	56.7
Vomit	32	26.7
Blood in the stool	11	9.2
Enterorrhea	3	2.5
Digestive tract complication	**3**	**0.03**
Haematemesis	1	0.8
Intussusception	1	0.8
Intestinal obstruction	1	0.8
Joint symptom	**79**	**65.8**
Knee joint pains	35	29.1
Ankle joint pains	33	27.5
Wrist joint pains	9	7.5
Elbow joint pains	5	4.2
Matecarpophalangeal joint pains	1	0.8
Lumbosacral joint pains	1	0.8
Hip joint pains	1	0.8
Kidney symptom	**65**	**54.2**
Microscopic hematuria	27	22.5
Gross hematuria	5	4.2
Albuminuria	32	26.7
Hematuria plus albuminuria	3	2.5
Tubular urine	1	0.8

Continuation of table 2

Symptom	Children (n = 120)	Percentage (%)
Other symptom		
Calf swelling	5	4.2
Miscellanous		
Orchitis	2	1.7
Pneumonia	3	2.5

Abbreviations: LE, lower extremities; UE, upper extremities.

Microscopic hematuria was defined when the test result was > +; Albuminuria was defined when the test result was > +; Gross hematuria was defined blood in the urine could be seen with the naked eye; Tubular urine was defined one more urinary cylinder were found per low power field period.

Table 3: Laboratory findings in 120 children of HSP

Findings	No. Positive/ No. Tested	Percentage (%)
Elevated ESR	68/120	56.7
Increased ASO titer	57/120	47.5
Low C_3 serum levels	13/120	10.8

Elevated ESR was defined when ESR was >20mm/1hour; increased ASO titer when >300IU/mm; low C_3 when <900mg/L.

Clinical features

The main clinical characteristics of the 120 HSP children were showed in table 2.

First symptom

The first symptoms were diverse. It was found that 83 cases (69.1%) displayed skin purpura alone, while 14 patients (11.7%) told abdominal pains. One child (0.8%) had joint pains, and purpura and joint pains were reported in 11 children (9.2%). We also found nine patients (7.5%) had purpura and abdominal pains meanwhile, and the joint pains, purpura and abdominal pains were complainted in 2 cases.

Purpura

All children appeared skin eruption. The purpura lesions of most patients were concentrated on the lower extremities and buttocks.

Digestive tract manifestation

Among these children, abdominal pains occurred in 68 cases (56.7%). The results showed 32 patients (26.7%) nausea, 11 children (9.2%) had blood in the stool, and 3 cases (2.5%) occured enterorrhea. Serious manifestations were gastrointestinal complications: 1 child (0.8%) haematemesis, 1 case (0.8%) intussusception, and 1 patient (0.8%) intestinal obstruction.

Joint symptom

Joint involvement occurred in many HSP children. These patients complained joint swelling. The results about the affected joints showed knee joints in 35 cases (29.1%), ankle joints in 33 children (27.5%), wrist joints in 9 patients (7.5%), elbow joints in 5 children (4.2%), metacarpophalangeal joints in 1 child (0.8%), lumbosacral joints in 1 child (0.8%), and hip joints in 1 child (0.8%).

Kidney symptom

Microscopic hematuria occurred in 27 children (22.5%), and gross hematuria were found in 5 cases (4.2%). Albuminuria were reported in 32 patients (26.7%), and there were three cases (2.5%) who appeared hematuria except albuminuria. We also found one tubular urine child (0.8%).

Uncommon clinical features

The results showed calf swelling in 5 cases (4.2%), orchitis in 2 children (1.7%). We also found 3 HSP cases had pneumonia during they accepted the remedy.

Laboratory findings

The main laboratory data of the acute phase were showed in Table 3. An increased ESR was reported in 68 patients (56.7%). Serum C3 decreased in 13 cases (10.8%). ASO titer was higher in 57 children (47.5%).

Treatment

The main resolved drug included the glucocorticoid, immunosuppressor, antihistamine drug, calcium channel blocker drug.

Outcome

There were 89 patients (74.2%) had been cured, and the conditions of 31 children of HSP (25.8%) improved.

Discussion

HSP which is characterized by a purpuric rash, arthritis, nephritis and gastrointestinal symptoms is a systemic vasculitis of unknown etiology.[9] It is the most common childhood vaculitis, with an annual incidence of about 10 cases per 100,000.[2] HSP was announced in association with infections, medications, vaccination. The pathogenesis is poorly understood. It was reported an IgA-mediate disease.[10] This multisystem disease mainly affected skin, kidneys, joints, gastrointestinal tract, and other organs still could be affected. It is considered nonfatal and self-limiting. Renal involvement could make the chronic consequences of HSP patients.[2]

The HSP children are mainly between the ages of 5 and 15 years and the mean age is 5 to 6 years.[3,4,6,7,12-15] In our investigation, the patients ranged from 1 to 12 years. According to our study, HSP occurs most frequently between the age of 6 to 7 years old, 88.3% were <10 years old, only 3 cases were <2 years old , and the mean age was 6.6 ± 1.6 years. In our literature, 65% of the patients were boys and 35% were girls. The ratio of M:F was 1.9:1. According to the findings, the boys are affected more often than the girls, and this findings were in accord with Frank T Saulsbury's "Clinical update: Henoch-Schonlein Purpura".[2] Gender distinction about the HSP incidence was also reported in other studies. In these investigations, HSP in children was more common in males. [3,14,16] By contrast, female advantage was found in these studies.[4,17]

According to our results, the incident seasons were spring, winter and autumn. The lowest incidence was in summer. The research outcomes were similar with some reports.[3,4,12-15,17-21] The etiological factors are still unclear. Our reports showed that 73 patients (60.8%) experienced infection before HSP onset and 32 cases (26.7%) occurred food allergy. Infection and food allergy were the main etiological factors in our study. Children often experienced respiratory tract infections in the cold months, so the high incidence in cold months suggested the association between HSP with infection, and this conclusion was alike with some investigations.[3,14,22] Some accounts had reported the trigger events including vaccinations, insect bites, and drugs have been connected with HSP.[23,24] In the present investigation, infection, food allergy, vaccination, parasite infection, insect bite, and emergency all may be potential trigger events for HSP onset. Furthermore we could not find the relevant factors about HSP onset for some patients.

In this study, we found that the incidence of the countryside children had been higher than the patients from the city. One of the reasons leading to this result may be the different hygiene and environment between city and countryside.

In terms of our research, the first symptoms for the HSP children were different. A majority of the HSP patients displayed skin purpura alone at the onset. The purpura lesion concentrated the lower extremities and buttocks. The skin eruption of all patients faded during 2 to 40 days, and the averages were 9.14 ± 5.42 days.

We discovered many HSP children displayed the digestive system symptoms. The dominant digestive clinical features were abdominal pains and vomiting. Digestive complications just as haematemesis, intussusception and intestinal obstruction could also occur. Gastrointestinal involvement buffered during 1 to 30 days, and the averages were 4.15 ± 1.14 days.

Many HSP patients displayed the joint symptom. The knee joint and ankle joint were most frequently affected, and the results were same with Sandra and coworkers.[5] In our study, the other joints just as wrist joint, elbow joint, metacarpophalangeal joint and hip joint were reported to be affected.

In our report, the typical kidney symptoms were microscopic hematuria and albuminuria. We also detected some cases occurred uncommon clinical features, just as calf swelling, orchitis and pneumonia.

Conclusion

We considered that there were gender difference, season difference and area difference for HSP children. The etiological factors were diverse. HSP patients could have various clinical symptoms and rare complications. Knowing well the diversiform clinical features of the HSP could help us to make the correct diagnosis and give the right cure and nursing. Although the epidemiology, clinical characterics, treatments, prognosis of HSP have been researched for many years, several questions still remain. Why do some children have the serious complication? Why do some children progress to end-stage renal disease? So we thought the investigation about the genetic susceptibility to HSP, immune pathogenesis, and more effective treatment still had been needed in this area.

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