Henoch-Schönlein Purpura in Thailand: Review of 41 Children

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Abstract

The clinical features of 41 Thai children with Henoch-Schönlein purpura were presented, with particular emphasis on the cutaneous features and date of onset. There were 20 boys and 21 girls, ages ranged from 2 to 15 years. All patients had palpable purpura on the lower limbs. Palpable purpura were also present on forearms and pinna in 25 (61%) and 5 (12%) respectively. Scalp edema was found in five patients. Hemorrhagic vesicles and bullae were found in only one patient. Twenty six patients (63%) had skin lesions as the first sign. Thirty four patients (83%) had skin lesions in the first four days of the clinical course. Other dominant features included were abdominal pain, arthralgia and nephritis 73, 66 and 39 per cent respectively. The pertinent literature is reviewed.

Henoch-Schönlein purpura is a syndrome characterized by nonthrombocytopenic purpura accompanied by arthralgia, gastrointestinal symptoms, or renal involvement[1-3]. The diagnosis of Henoch-Schönlein purpura is based on specific clinical features. The characteristic skin lesions are the most helpful aid in diagnosis. The skin lesions consist of small, 2- to 10-mm hemorrhagic macules, papules, or urticarial lesions that appear symmetrically distributed over the lower part of the legs, buttocks and forearms[1-3]. The clinical manifestations of Henoch-Schönlein purpura have been studied extensively elsewhere[1,2,4-8]. We presented the clinical manifestations of Henoch-Schönlein purpura, with particular emphasis on the cutaneous manifestation, date of onset and review the pertinent literature.

PATIENTS AND METHOD

The medical records of all patients with Henoch-Schönlein purpura up to 15 years of age were seen at Chulalongkorn Hospital at the onset of their illness, during a period of eight years (1988-1995), were reviewed. The diagnosis of Henoch-Schönlein purpura was made by a characterized palpable purpura in conjunction with one or more of the following: abdominal pain, joint symptoms or nephritis.

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Nephritis may be demonstrated by gross or microscopic hematuria, with or without proteinuria. All patients had a complete blood count with platelet quantification. The following tests were performed on some patients if palpable purpura was the only manifestation at the first visit: antinuclear antibody, rheumatoid factor, total hemolytic complement titer and serum C_3 concentration.

RESULTS

There were 20 males and 21 females, age ranged from 2 to 15 years of age, all exhibited a characteristic purpuric eruptions concentrated on the lower extremities. Thirty patients (73%) had abdominal pain, 27 patients (66%) had joint involvement and 14 patients (39%) had nephritis.

The first symptoms and signs of Henoch-Schönlein purpura in our patients are shown in Table 1. Skin lesions, palpable purpura, appeared as the first sign in 26 patients (63%). Abdominal pain and joint involvement appeared as the first symptoms and signs in 4 patients (10%) equally. Only one patient presented with gross hematuria as the first manifestation.

The clinical manifestations of Henoch-Schönlein purpura in this series compared with other series are shown in Table 2. All patients had palpable purpura on the lower extremities (Fig. 1). Lesions on the forearms were found in 25 patients. Five patients had typical palpable purpura at both pinnas. Extremities edema were found in 14 patients. Four patients, aged 3, 4, 5 and 7 years, had scalp edema. One patient had high fever, hemorrhagic vesicles and bullae. Twenty six patients (63%) had skin lesions as the first manifestation. Thirty four patients (83%) had skin lesions in the first four days of the clinical course.

Table 1. Presenting symptoms and signs in Henoch-Schönlein purpura of 41 Thai children.

Skin	63.4%
Palpable purpura	61.0%
Extremities edema	2.4%
Abdominal pain	9.7%
Joint	9.7%
Arthritis	7.3%
Arthralgia	2.4%
Kidney	2.4%

Table 2. Clinical manifestations of 41 Henoch-Schönlein purpura patients compare to previous report.

Reference No.	1	2*	4	5	6	7	8	This series
No. of Patients	131	10	26	75	25	52	71	41
Skin (%)	100	100	100	100	100	100	100	100
Palapble purpura	100	10	100	100	100	100	100	100
Superficial necrosis	-	-	-	4	-	-	-	-
Bullae	-	100	-	-	-	2	-	2.4
Extremities edema	46	-	-	}45	-	10	4.1	34.15
Scalp edema	24	-	-	-	4	4	-	9.75
Scrotal edema and ecchymosis	-	-	-	-	-	-	-	2.4
Joint (%)	68	80	62	74	84	83	55.4	66
Gastrointestinal (%)	69	100	42	61	76	60	62	73.17
Pain	53	100	42	61	76	58	14.9	73.17
Bleeding	56	80	26	29	40	30	24.4	9.75
Volvulus	-	-	-	-	-	-	1.4	-
Perforation	-	-	-	-	-	-	-	2.4
Renal (%)	40	90	35	100	44	46	9.8	39.02
Microscopic hematuria	24	60	}23	100	32	-	8.4	24.39
Gross hematuria	16	40	-	47	12	-	1.4	14.63
Proteinuria	27	70	35	43	32	-	9.8	24.39
Recurrence (%)	40	30	38	13	55	8	-	21.95

- = no record

* Exclusion of two adult patients

Abdominal pain was the second common clinical feature, occurring in 73 per cent of patients. Gastrointestinal bleeding was documented in 4 patients, all of whom had abdominal pain. One patients had ileal perforation despite systemic corticosteroid. Joint involvement, particularly the ankles and knees, was found in 66 per cent of patients. Most of the patients had arthritis, only 8 patients had arthralgia. Sixteen patients had nephritis; 6 patients had gross hematuria, 10 patients had microscopic hematuria and 10 patients had proteinuria.

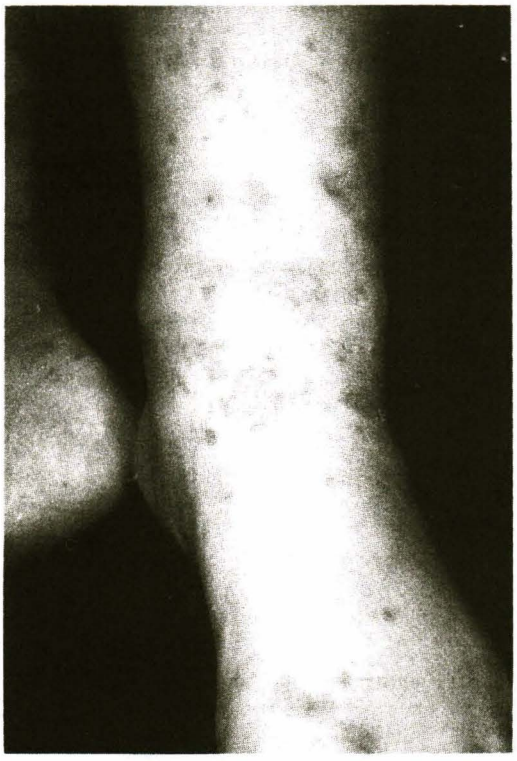

Fig. 1. Palpable purpura on the lower extremities, characteristic skin lesion of Henoch-Schönlein purpura.

The laboratory data are depicted in Table 3. Hematocrit values were normal in all patients. White blood cell counts ranged from 6,000 to 25,000/mm^3. Platelet counts ranged from 220,000/mm^3 to 680,000/mm^3. Serum immunoglobulin concentration were determined in 14 patients: serum IgG was elevated in 2 patients; serum IgM was elevated in 3 patients; but serum IgA was all within normal range. Antinuclear antibody and rheumatoid factor were negative in all patients tested. Serum C_3 concentrations and total hemolytic complement titers were normal in all patients tested. None of the laboratory findings listed above correlated with the clinical features of Henoch-Schönlein purpura of our patients. Skin biopsies were done in 10 patients. All specimens showed leukocytoclastic vasculitis of the dermal vessels. Direct immunofluorescent studies revealed granular deposits of IgA in the wall of dermal vessels in 3 patients.

All patients had been routinely followed-up for 6 months or more. Nine patients (22%) had one or more recurrences of their disease within 4 months except one patient, a 5 year old girl, who had her initial recurrence at 11 months after onset of the disease.

DISCUSSION

The clinical manifestations of Henoch-Schönlein purpura were similar to those in other reported series[1,2,4-8]. The diagnosis of Henoch-

Table 3. Laboratory features in 41 patients with Henoch-Schönlein purpura.

	Low	Normal	High	Total test
Hematocrit	0	41	0	41
White blood count	0	33	8	41
Platelet count	0	33	8	41
Serum IgG	0	12	2	14
Serum IgA	0	14	0	14
Serum IgM	0	11	3	14
Antinuclear antibody	0	18	0	18
Rheumatoid factor	0	18	0	18
LE cell	0	18	0	18
Total hemolytic complement titer	0	18	0	18
Serum C_3	0	18	0	18

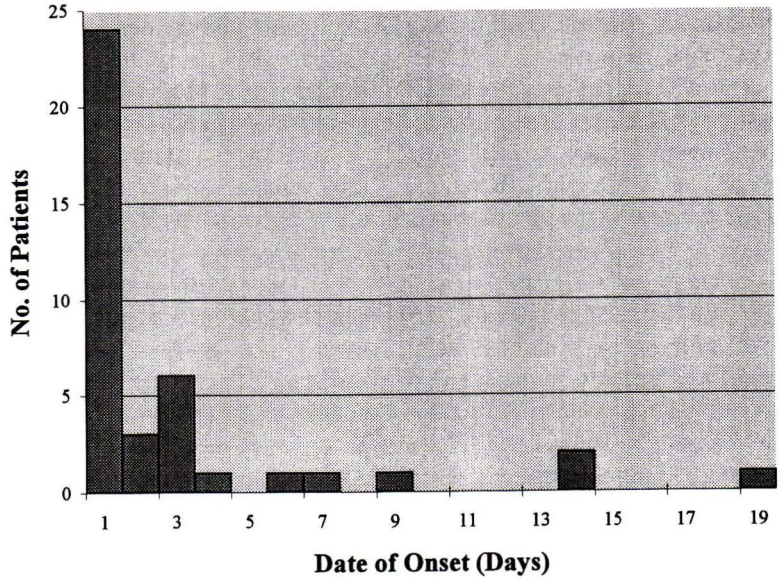

Fig. 2. Date of onset of skin manifestation in 41 Henoch-Schönlein purpura's patients.

Schönlein purpura is based on specific clinical features. The characteristic skin lesions are palpable purpura, 2 to 10 mm in diameter, initially seen as generalized erythematous macules, papules, and urticarial plagues. Within 48 hours these lesions become purpuric papules and plagues interspersed with petechiae. They are symmetrically distributed on the lower part of the legs, buttocks and forearms[1-3]. In this series, the lesions on the lower extremities were found in all patients but the lesion on forearms were found only in 60 per cent of the patients. Palpable purpura at pinnas are not rare, we found them in 12 per cent of our patients. Hemorrhagic vesicles and bullae in pediatric patients are quite rare[2,5,7,9]. We had one patient, a 5.5 year old boy who presented with high fever and hemorrhagic vesicles and bullae varying in size from 2 to 50 mm in diameter, on both pinnas, the hard palate, gums, dorsum of the hands, buttocks and both legs. Histopathology showed dense perivascular nutrophilic infiltrate with nuclear dust. Direct immunofluorescence of normal skin adjacent to the lesion showed deposition of IgA, C_3 and minimal IgM in blood vessel walls, which was characteristic of Henoch-Schönlein purpura. The other cutaneous manifestations of Henoch-Schönlein purpura are soft tissue edema of the scalp which is more common in those under 3 years[1]. In our series, three patients were over 3 years, their ages were 3, 4, 5 and 7 years. Soft tissue edema of the extremities were found in 34 per cent of our patients. Soft tissue edema of the extremities are common[1,2,4-7]. We found only one patient who showed petechiae at mucous membrane. Petechiae may be found rarely on the face, genitalia and mucous membrane[1,2,5]. One patient had scrotal edema and ecchymosis which is rare[10].

Histopathology of the cutaneous lesions showed leukocytoclastic vasculitis of the dermal vessels characterized by the presence of scattered nuclear dust owing to degenerating polymorphonuclear leukocytes[11]. Immunofluoresence studies of uninvolved skin showed stippled deposits of IgA in the walls of dermal vessels[12]. In our patients, only 3 patients had positive results. This might due to biopsy of late lesions because direct immunofluorescence studies of lesions older than 48 hours are frequently negative due to destruction and removal of antibodies in the affected necrotic blood vessels[13].

We found abdominal pain in 73 per cent of our patients. Gastrointestinal symptoms are potentially the most serious manifestation of Henoch-Schönlein purpura which results from edema of bowel wall and hemorrhage owing to the vasculitis. The dominant features are abdominal

pain, vomiting, hematemesis, melena and bleeding which can be found in 60-70 per cent[1,2,4-7].

Ten per cent of our patients presented with abdominal pain preceding skin lesions. Diagnosis of Henoch-Schönlein purpura is usually not difficult if the classic rash is present. Abdominal pain and gastrointestinal bleeding may precede palpable purpura for days or weeks and thus simulate a number of inflammatory or surgically treated bowel diseases[1,14,15]. One of our patients, a 5 year old boy, came with severe abdomen pain, vomiting and melena for nineteen days before skin lesions. He was treated as segmental enteritis for eighteen days without any improvement and finally exploratory laparatory was done and only mild inflammation of terminal ileum was found. Palpable purpura on lower extremities was then first noted on the second post operative day which was the nineteenth day of the disease and skin biopsy revealed leukocytoclastic vasculitis. Systemic steroid was started with gradual clinical improvement. Another patient, a four year old girl, presented with edema of the extremities, palpable purpura and severe abdominal pain on the first, third and fourth day consecutively. Steroid was started on the fifth day but the clinical state of the patient worsened despite meticulous conservative treatment for 7 days and finally showed signs of bowel perforation. The operative findings revealed perforation at terminal ileum, size 0.5 x 0.5 cm, 1 foot from ileocecal valve. Although most of the abdominal pain resolved within three to seven days, with or without steroid therapy[16], steroids may have a role in hastening the resolution of self-limited pain but the efficacy of corticosteroids is debatable and are still used in severe cases, particularly those with significant gastrointestinal complications[17]. Bowel infarction, perforation, intussusception[18], pancreatitis [19,20], cholecystitis[21] and hydrops of the gall bladder[22] may occur. Jejunum and ileum are the most commonly affected sites. Direct immunofluoresence of the bowel showed IgA deposits in the vessel walls similar to those observed in the skin and kidney[23]. Esophageal stricture[24] and ileal stricture[25] can be found as late complications.

Joints were affected in 63.2 per cent of our patients, the same incidence reported 60-80 per cent in other series[1,2,4-7]. The most common joint involvement is the ankles, the knees, and the dorsum of hands and feet. Elbows and fingers are occasionally involved. Joint involvement was relatively transient and left no deformity.

In our series, only 4 patients had joint involvement as the first clinical presentation and all 4 patients involved ankle joints but edema extended above the mid calf region.

Nephritis was diagnosed clinically in 39 per cent of our patients. Ten in fourteen of our patients showed gross hematuria. Proteinuria was found in 10 patients. The reported incidence of nephritis, in unselected series, the incidence is about 35-46 per cent[1,6,7,18]. Nephritis may be demonstrated by gross or microscopic hematuria, with or without proteinuria. Although we found renal involvement in fourteen patients, only one patient had gross hematuria as the first presentation. The onset of nephritis may be delayed for weeks or months, but over 90 per cent of patients with renal involvement will show hematuria within the first three months[27,28].

Nephritis is the only feature of Henoch-Schönlein purpura that may become chronic[1,3, 6,25,29,30]. Patients with isolated microscopic hematuria have good prognosis. Those who present with hematuria and proteinuria have a 10 per cent chance of renal insufficiency or active renal disease by 10 years[26]. Although neurological involvement varies in severity from headache, seizures, focal neurological deficits, mononeuropathies and polyradiculoneuropathies can be seen[31,32]. None of our patients showed neurological involvement. Other manifestations such as pulmonary hemorrhage[33,34], pleural hemorrhage[35], and myocardial infarction[36,37] were reported as rare manifestations.

We describe the clinical manifestations of Henoch-Schönlein purpura which have been recognized for over 100 years, but the etiology still remains poorly understood. A wide variety of infectious agents and drugs have been linked to Henoch-Schönlein purpura such as streptococcal infection[3,28], mycoplasma[39], varicella[40], hepatitis B[41], drugs[42,43], insect bites[44] and exposure to cold[45]. Henoch-Schönlein purpura can involve multiple organs and has a wide range of severity and course of disease, further studies are needed to understand more about the pathogenesis of Henoch-Schönlein purpura.

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เฮน็อค–เชินลาย เพอเพอรา ในเด็กไทย : รวบรวมผู้ป่วย 41 ราย

ศิริวรรณ วนานุกูล, พ.บ.,*
เทวี วัฒนา, พ.บ., ประไพ พงษ์ประสิทธิ์, พ.บ.**

ได้ศึกษาอาการและอาการแสดงของผู้ป่วยเด็กไทยที่เป็น Henoch–Schönlein purpura ที่มารับการรักษาที่ภาควิชากุมารเวชศาสตร์ โรงพยาบาลจุฬาลงกรณ์ จำนวน 41 ราย เป็นผู้ป่วยเด็กชาย 20 ราย และเด็กหญิง 21 ราย อายุระหว่าง 2 ถึง 15 ปี ทุกรายมีผื่นแบบ palpable purpura ที่ส่วนล่างของขา ส่วนผื่นที่แขนและใบหูพบร้อยละ 61 และ 12 ตามลำดับ พบหนังศีรษะบวมในผู้ป่วย 5 ราย ผื่นแบบ hemorrhagic vesicles และ bullae พบในผู้ป่วยเพียง 1 ราย ผู้ป่วยร้อยละ 63 มีอาการแสดงทางผิวหนังก่อนอาการอื่น และร้อยละ 83 มีอาการแสดงทางผิวหนังในระยะ 4 วันแรกของโรค ส่วนอาการปวดท้อง ปวดข้อ และอาการทางไตพบร้อยละ 73, 66 และ 39 ตามลำดับ

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