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Clinical profile and pattern of Henoch-Schönlein purpura in children

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ABSTRACT

Introduction: This study was done to evaluate clinical profile of Henoch-Schönlein purpura (HSP) in children admitted in Patan Hospital.

Methods: The medical records of all children admitted in children ward with the diagnosis of HSP from January 2008 to December 2015 were analysed for clinical presentation, management and outcome.

Results: Of 59 patients, 37(63%) were boys. The patients' ages ranged from 15 months to 14 years with a mean of 8.3 years. Approximately two third cases presented during winter and autumn. Upper respiratory tract infection preceded HSP in 37 and anti streptolysin O titer was positive in 6 of the 13 (46%) children tested at presentation. Skin purpura was seen in 59 (100%), arthritis or arthralgia 48 (81%), gastrointestinal manifestation 47 (80%) and renal involvement 16 (27%). Forty one (69%) patients received corticosteroid therapy. All children made a full recovery, two with nephritis continued to have hypertension, hematuria and proteinuria. Symptoms occurred in 6 (10%) over a period one month to two years follow up.

Conclusion: The HSP in children had seasonal occurrence, skin rash, pain abdomen with mild disease and recovered well.

Key Words: clinical profile, Henoch-Schönlein purpura, vasculitis, palpable purpura

INTRODUCTIONS

Henoch-Schönlein purpura (HSP) is the most common vasculitis of childhood. Approximately 90% of HSP cases occur in children between the ages of 3 and 10 year.¹ It is characterized by non-thrombocytopenic purpura, arthritis or arthralgia, abdominal pain, gastrointestinal hemorrhage and renal manifestations.² The classical triad seen is palpable purpura, abdominal colicky by pain or hemorrhage and joint pain. However, diagnostic confusion can arise in cases who present with the absence of cutaneous features.³ This study evaluates the clinical profile of HSP in children admitted in Patan Hospital.

METHODS

The medical records of all children admitted in children ward with the diagnosis of HSP from January 2008 to December 2015 were retrieved and analyzed. Diagnosis of HSP was based on presense of at least two of the following four criteria: 1) Palpable purpura not related to thrombocytopenia, 2) Age 20 years or younger at onset, 3) Bowel angina, 4) Histology changes showing granulocytes in the walls of arterioles or venules.⁴ The demography (age and gender) and clinical characteristics, laboratory data at onset, therapy, and recurrence were studied. Renal biopsies were graded according to the classification of the International Study of Kidney Disease in children (ISKDC).⁵

Recurrences was defined as a new flare-up of skin rash or other systemic complications following resolution of the disease for at least one month.⁶ Continuous variables (such as age) were expressed as mean, median, and range. Absolute numbers as well as percentages were presented for the study variables. Descriptive analysis was done.

RESULTS

There were total of 59 (m 37, f 22) children with HSP. All patients had nonthrombocytopenic palpable purpura, distributed mainly over the legs, buttocks, and upper extremities, (Table 1, 2).

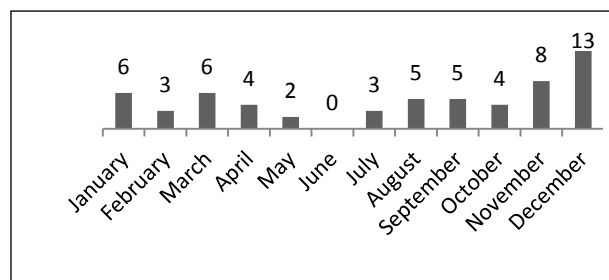


Figure 1. Seasonal occurrence of Henoch-Schonlein purpura (HSP) in children (n=59)

Table 1. Demography and etiology of HSP in children (n=59)

Demographic Data	
Age of onset (years)	
Range	1.25 – 14
Mean	8.3
Median	8
Gender	
Male	37 (62.7)
Female	22 (37.2)
Male to Female Ratio	1.6 : 1
Age Distribution (years)	
< 5	13 (22)
> 5 to < 10	31 (52.5)
> 10	15 (25.4)
Season Pattern	
Winter	22 (37.2)
Autumn	17 (28.8)
Spring	12 (20.3)
Summer	8 (13.5)
Etiological Factor	
URTI	22 (37.2)
Fever	11 (18.6)

Table 2. Clinical features of 59 patients with HSP

Symptom	n (%)
Skin	59 (100)
Joint Involvement	48 (81.3)
Arthritis/Arthralgia	48 (81.3)
Ankle/Feet	45 (76.2)
Knee	41 (69.4)
Wrist/Hand	16 (27.1)
Elbow	13 (22.0)
Gastrointestinal Involvement	46 (80.0)
Abdominal Pain	44 (74.5)
Malena	20 (33.8)
Vomiting	25 (42.3)
Hematechezia	6 (10.0)
Hematemesis	2(3.3)
Intussusceptions	0 (0)
Renal Involvement	16 (27.0)
Microscopic Hematuria	12 (20.3)
Macroscopic Hematuria	2 (3.3)
Proteinuria	9 (15.2)
Nephrotic Syndrome	5 (8.0)
Edema of face, dorsum of feet, hand	3(5.0)
Seizure	2 (3.3)
Recurrence	6 (10.0)

Prednisolone 1-2 mg/Kg/day was prescribed to 41/59 (69%) patients for renal involvement, or joint and GI manifestations. All patients made a full recovery. Two and persistent proteinuria were on regular followup.

The c-reactive protein, elevation of ESR and leucocytosis were seen children with HSP, (Table 3).

Table 3. Laboratory findings among HSP patients

Laboratory Test	Positive/Tested n (%)
Leukocytosis (WBC > 11000/mm ³)	28/59 (47.4)
Thrombocytosis (platelets > 500000 cell/mm ³)	14/59 (23.7)
Anemia (Hemoglobin < 11 gm/dl)	9/46 (19.5)
Elevated ESR	20/39 (51.2)
Positive ASO titer (> 200 IU/ml)	6/13 (46)
Increased C-reactive protein	12/21 (57.1)
Positive throat culture	0/19 (0)
Low C3	3/13 (23)
Renal Biopsy	2/2 (100)

WBC: While Blood cells. ESR – erythrocyte sedimentation rate. C3- complement 3, ASO – anti-streptolysin O titer

DISCUSSIONS

In the present study, nearly 75% of children were less than 10 years of age. Male to female ratio was 1.68:1. Similar results were obtained by Cakir Murat et al⁶ who in their study of 116 children, 73 (63%) were male and 43 (37%) females with a ratio of 1.69:1. Naveed Shahzad et al⁷ in their 27 patients, 17 (63%) males and 10 (37%) females with a ratio of 1.7:1 and a slight female preponderance was found in some studies.^{8,9}

Henoch-Schönlein purpura occurs throughout the year, but a number of studies have noted seasonal variation, with most patients presenting from fall through spring and a paucity of cases in the summer months.^{10,11} In the current study, there were more patients in winter and fall (66%). Mahar khadar et al¹² found distribution of admission was highest in winter months. Nearly 56% of patients

had a potential trigger event before HSP onset, which is consistent with study done by Amer A Lardhi in 2012 found potential trigger event before HSP onset in 55% of cases.¹³

The main clinical features of HSP included purpuric rash, GI symptoms, joint and kidney manifestation. Classic skin purpura was present in all patients in current study. Similar results were observed by I. Kumar et al.¹⁴ Rashes were often seen in the lower extremities and buttocks, but some also had involvement of the arms and face. Joint involvement including arthritis and arthralgia is common occurring in up to 75% of children with HSP, with a predilection for the lower extremities.¹ It was observed in 81% of the patients in our study and mainly affecting ankles and knees. The joints of the upper extremities were involved in few patients.

Gastrointestinal manifestations of HSP occur in up to 80% of children with HSP and include abdominal pain, vomiting, diarrhea and melena; Intussusception, mesenteric ischaemia and intestinal perforation are uncommon.¹ Gastrointestinal manifestation has been described in up to 75% in some series.^{15,16} Abdominal pain was present in 66.6% of cases reported by Naveed Shahzad et al. in May 2015.⁷ Whereas Saulsbury FT¹⁷ found that abdominal pain was present in 63% of patients. Abdominal colic, vomiting and gross or occult bleeding are the dominant features of GI involvement 46 (80%) patients in our study. None of the patients developed intussusception or perforation in the current study.

The long term morbidity and mortality of HSP are related to the severity of renal involvement. The reported incidence of renal involvement varies from 10-50%; with end stage renal disease in 5%.^{18,19} In the present study, renal involvement occurred in 16 (27.0%) patients. Amer A. Lardhi found renal involvement in 19 out of 78(24%) patients.¹³

The low incidence of renal involvement in the current series may be related to local variations in causative and environmental factors. The main clinical features of renal involvement in the present study were microscopic hematuria with or without proteinuria, and nephritic syndrome.

Nephritic syndrome developed in 5(31%) patients. The overall outcome of HSP nephritis is good. All patients with nephritis in the present study made a complete recovery, but two patients who had hypertension, hematuria and persistent proteinuria required treatment for hypertension and were on regular followup.

Other less common potential manifestations of HSP are orchitis, carditis, inflammatory eye disease testicular torsion, and pulmonary hemorrhage.¹ which were not present in our study. Seizure are rare manifestations of CNS involvement in patients with HSP.^{10,20} Two patients (3.3%) developed seizure because of hypertensive encephalopathy in this study which is also consistent with study done by Amer A. Lardhi who found seizure in 2.5% of cases.¹³

In agreement with literature,^{18,20,21} the present data show increased ESR in 20/39 (51%), CRP in 12/21 (57%), leukocytosis in 28/59 (47%), Table 3. Complement component were decreased in 3/13 (23%). Decreased complement levels are reported in 10-18% and increased serum IgA in 0-62% but are not routinely measured.^{18,22,23}

The frequency of relapses varies from 5-66%.^{23,24} In the present study, 6 (10%) patients relapsed within the first 2 years. These figures are similar to Amer A. Lardhi who found 8% relapse in his patients within the first 2 years.¹³

Some of the patients with HSP require only supportive measures. Corticosteroid therapy reduces the duration and severity of abdominal and joint pain but corticosteroids do not prevent the development of nephritis, or alter the natural course of HSP.^{20,25} However, 41 (69%) patients in the present study received steroid for GI and renal involvement. Patients with severe HSP nephritis may require other regimen of treatment. This may include high dose steroid, azathioprine, cyclophosphamide, mycophenolate mofetil, intravenous immunoglobulin and plasma pheresis.^{26,27}

In this study, we did not include all cases attended the outpatients departments in pediatrics, and dermatology. Therefore, these cases might

represents the severe form of clinical presentations only.

CONCLUSIONS

The HSP in children had seasonal occurrence, skin rash, pain abdomen with mild disease and recovered well.

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