International Journal of Contemporary Pediatrics
Abbas S et al. Int J Contemp Pediatr. 2017 May;4(3):822-826
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20171493

Original Research Article

Clinical profile and outcome of Henoch Schonlein purpura in a tertiary care hospital in South India

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Received: 20 March 2017
Accepted: 25 March 2017

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ABSTRACT

Background: Henoch Schonlein purpura (HSP) is the most common systemic vasculitis in children. It is an immunoglobulin A (IgA) mediated systemic small-vessel vasculitis, with IgA deposition in vessel walls leading to symptoms involving the skin, joints, intestines, and kidneys. The objective of present study was to identify and describe the clinical profile, pattern of joint involvement, histopathological features, treatment modalities and complications of Henoch Schonlein purpura.
Methods: 52 children less than 12 years diagnosed to have Henoch Schonlein Purpura according to the European League against Rheumatism criteria were included in the study. All patients were subjected to detailed history taking and thorough examination. The clinical features, investigations, management and complications of the disease were studied. Descriptive statistics was used to analyse the results.
Results: The common clinical features were palpable purpura (100%) followed by arthritis (66%) and abdominal pain (50%). Renal manifestations were in the form of hematuria in 12% of the patients. Skin biopsy was done in 25 patients of which 19 had findings suggestive of Henoch Schonlein purpura. Steroids were used in 42% of patients. The severity of illness was associated with lower mean age, arthritis, leg edema and stool occult blood.
Conclusions: The clinical features of Henoch Schonlein purpura in the population were different from the previously published studies. Renal involvement was less common. The short-term outcome of the patients were satisfactory.

Keywords: Clinical features, HSP, Treatment

INTRODUCTION

Henoch Schonlein purpura (HSP) is the most common systemic vasculitis in children.[1] It is an immunoglobulin A (IgA) mediated systemic small-vessel vasculitis, with IgA deposition in vessel walls leading to symptoms involving the skin, joints, intestines, and kidneys.[2]

This autoimmune vasculitis is characterised by multi organ involvement in the form of non-thrombocytopenic palpable purpura, abdominal pain, arthritis and hematuria. The annual incidence of Henoch Schonlein Purpura in children is around 14-20 per 1,00,000 population.[3] It usually has a self-limited course but has the potential to cause serious life threatening complications including gastrointestinal perforation and end stage renal disease.

The current knowledge on the treatment of Henoch Schonlein purpura is limited. The aim of this study is to describe the age of onset, pattern of joint involvement, histopathological study of the skin lesions, response to the therapeutic agents and the development of complications of the disease.

METHODS

This is a hospital based descriptive study carried out in children under 12 years of age admitted with palpable purpura in a tertiary care hospital in South India during a period of two years.

The diagnosis of HSP was based on European League against Rheumatism (EULAR) endorsed consensus criteria for HSP classification.[4] which includes: palpable purpura (in absence of coagulopathy or thrombocytopenia) and 1 or more of the following criteria: abdominal pain (acute, diffuse, colicky), arthritis or arthralgia, biopsy of affected tissue demonstrating predominant immunoglobulin A deposition, renal involvement (proteinuria >3 grams/24 hours) hematuria or red cell casts

Histopathology typically is a leucocytoclastic vasculitis with predominant IgA deposit. Renal involvement is proteinuria >30mmol/mg of urine albumin/creatinine ratio on a spot morning sample or >0.3g/24 h,presence of red blood cell casts, hematuria or >5 red blood cells/high power field. Arthritis is defined as arthralgia with limited movement or joint swelling. Arthralgia is defined as joint pain without limitation of movement or swelling.

The cases were subjected to detailed history taking, complete physical examination including pattern of joint involvement, investigations including total and differential counts, platelet count, renal function test, ASO titre, urine examination, stool occult blood and ultrasound abdomen. Skin biopsy was done for almost half of the patients. Treatment was given according to the severity of the disease as supportive management with or without steroids.

Statistical analysis

Continuous variables were expressed as mean ± standard deviation (SD). Categorical variables were expressed as percentages. Inter-group differences on univariate analysis were evaluated by χ^2 test for all parameters except age for which t test was done. All statistical analyses were performed using SPSS 18 (Statistical Package for Social Sciences IBM Corp., New York, NY, USA). P value of <0.05 was considered statistically significant.

All procedures were explained to the parents and a written informed consent was obtained. Ethical clearance was obtained from the institutional ethics committee. The study conforms to the guidelines of Declaration of Helsinki.

RESULTS

During the study period, 52 children were diagnosed to have Henoch Schonlein purpura. The mean age of onset of disease was 6.76 years (range 1.5-12 years), with a male: female ratio of 1:1. The common presenting symptoms were purpuric rash (n = 52, 100%), arthritis (n = 40, 77%) and abdominal pain (n = 29, 56%) (Table 1).

Table 1: Clinical profile in two groups.

Symptoms	<6 years N=23	>6 years N=29
Male:Female	11:12	15:14
Rash	23	29
Arthritis	20	20
Edema	17	15
Abdominal pain	14	15
Vomiting	8	10
Melena	5	5
Hematuria	2	4
Hepatomegaly	9	6
Splenomegaly	1	6

Other gastrointestinal symptoms included vomiting (n = 18, 35%) and melena (n = 10, 19%). Epistaxis and hematemesis were present in 2 (3.84%) patients each.

Genitourinary symptoms included hematuria (n = 6, 11.54%) and scrotal pain (n = 2, 3.84%). None of the patients had oliguria, hypertension or acute renal failure.

Rash preceded arthritis and abdominal pain in 40 patients (76.9%). Arthritis preceded rash in 8 patients (15.3%) and abdominal pain preceded rash in 6 patients (11.5%). Most commonly, the rash first appeared in the lower limbs and buttocks; and gradually spread to other areas.

Physical examination showed pallor (n = 17, 33%), hepatomegaly (n = 15, 29%) and splenomegaly (n = 7, 13.5%). Among the 40 patients (77%) with arthritis, the major joints involved were ankle (n = 30, 58%), knee (n = 20, 39%) and elbow (n = 10, 19%). Other joints involved were wrist, shoulder, small joints of hands and feet and thoracolumbar spine. Monoarticular involvement was seen in 18/40 (45%) patients with arthritis. None of the patients had neurological complications.

Table 2: Laboratory parameters.

Lab parameters	Frequency	Percentage
Anemia	25	48.08
Leucocytosis	9	17.31
High ESR	25	48.08
Thombocytosis	10	19.23
Stool occult blood	10	19.23
AntiStreptolysin O positive	8	15.38
Antinuclear antibody	1	1.92
Rheumatoid factor	2	3.85
Abnormal urinalysis	8	15.38
Abnormal renal function	3	5.77
Abnormal ultrasound scan	8	15.38

An elevated ESR and anemia were present in 25 cases (48%) each. Thrombocytosis was present in 10 (19%) patients with a mean platelet count of 2.85 L/mm^3. ASO titre was available in 36 patients of which a titre of >200 was found in 8 (15.38%) patients. Abnormal urine analysis was noticed in 8 (15%) patients, of which major abnomality was hematuria (n = 6, 11.54%). Persistent proteinuria was present in 2 (3.84%) patients (Table 2).

Skin biopsy was done in 25/52 (48%) patients, of which 19/25 (76%) had histopathology suggestive of HSP. Others had nonspecific findings in histopathology. The most common histopathologic findings were perivascular inflammatory infiltrate and neutrophilic debris. Other findings were extravascular RBCs, endothelial proliferation and fibrinoid necrosis (Table 3).

Table 3: Histopathology of skin rash.

Histopathology	Frequency	Percentage
Perivascular Inflammatory Infiltrate	17	68
Neutrophilic Debris	13	52
Extravascular RBC	4	16
Endothelial profileration	8	32
Fibrinoid necrosis	6	24
Non-specific	6	24

Table 4: Risk factors for severity and treatment.

Factor	Risk category	Steroids given n = 22	No steroids n = 30	Probability (P) value	Significance
Age	Mean age (Standard Deviation)	5.59 (2.33)	7.53 (2.64)	0.008	Present
Gender	Males	12 (54.5%)	14 (46.7%)	0.57	Absent
	Female	10 (45.5%)	16 (53.3%)		
Abdomen	Abdominal pain	19 (86.3%)	10	0.01	Present
	No pain	3 (13.7%)	20		
Joint	Arthritis	21	20	0.01	Present
	No arthritis	1	10		
Renal	Hematuria	5	1	0.07	Absent
	No hematuria	17	29		
Leg	Edema	19	13	0.002	Present
	No edema	3	17		
Onset of rash	Rash first	15	23	0.54	Absent
	Rash later	7	7		
Hemoglobin	Anemia	6	19	0.01	Present
	Normal	16	11		
Leucocyte count	Leucocytosis	15	28	0.02	Present
	Normal	7	2		
ESR	High>30	14	11	0.09	Absent
	Normal	8	19		
Platelet count	High>4 lakhs	7	3	0.07	Absent
	Normal	15	27		
Stool occult blood	Positive	9	0	0.000	Present
	Negative	13	30		

Out of the 52 patients, 22 needed steroids in addition to symptomatic treatment. The factors affecting the severity of the disease were categorised based on the treatment with or without steroids (Table 4). The severity of illness was associated with lower mean age, arthritis, leg edema & stool occult blood. Indications for steroids were bowel symptoms (n = 12, 54.5%), severe arthritis (n = 6, 27%) and extensive rash (n = 4, 18%). In all patients with abdominal pain, symptoms decreased within 24 hours of steroids. The mean duration of treatment in hospital was 6.62 (3.3) days. Steroids was tapered and stopped after discharge in all patients. Surgical consultation was needed in 10 patients. Ultrasound scan of abdomen was done in 15% patients. Bowel wall edema or ascites were present in 70% of steroid treated group-complete recovery was seen in 45 cases (86.5%) and 7 patients (13.4%) had complications of which 4 (7.7%) had acute abdomen, all treated conservatively.

DISCUSSION

The mean age of onset of symptoms was 6.76 years which is comparable to other studies.[5-7] There was an equal sex predilection in our study in contrast to other studies which had male predominance.[3,5,6,8]

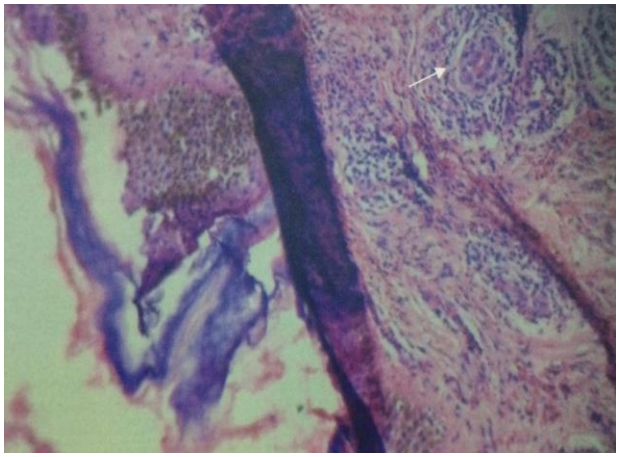

Figure 1: Vasculitis in skin biopsy.

In comparison to our study at the onset, purpura was present in all cases, arthritis/arthralgia in 195 (45.3%), abdominal involvement in 148 (34.4%), and renal involvement in 192 (44.7%) in a study by Anil et al.[6] In a study by Saulsbury the dominant clinical features were cutaneous purpura (100%), arthritis (82%), abdominal pain (63%), gastrointestinal bleeding (33%), and nephritis (40%).[9]

Similar to our study, in previous studies rash has been reported to occur mainly in lower limbs and buttocks.[10,11] Thrombocytosis helps in distinguishing the condition from other thrombocytopenic purpuras.

Occult blood positivity in stools was less when compared to the series by Wang et al which consisted of 49% patients.[11] Four of our patients presented with acute abdomen before the appearance of skin rash. Among these patients two had acute appendicitis, one had pancreatitis and one had bowel infarction. All cases were managed conservatively. Ultrasound was the imaging modality of choice in evaluating the bowel manifestations of Henoch-Schonlein purpura similar to studies by Connoolly et al.[12]

Gastrointestinal involvement occurs in approximately two thirds of children with Henoch-Schonlein purpura and usually is manifested by abdominal pain. Abdominal symptoms preceded the typical purpuric rash of HSP in 11% cases in accordance to study by Choong et al in which the incidence was 14-36%.[13] Major complications of abdominal involvement like intussusception did not occur in our series but developed in 4.6% of cases in their series.[13] The symptoms may mimic an acute surgical abdomen and result in unnecessary laparotomy.

Arthritis preceded rash in 14.8% cases and was present in 77% of the study population. Most children developed joint symptoms simultaneously with rashes or within 11 days of appearance of the rash and 65% developed joint symptoms after having had the rash for 1 to 3 days in a study by Xuehong Wang et al.[11] The most common joint involved was ankle joint followed by knee and elbow joint in contrast to the series by Xuehong Wang et al in which knee joint was most commonly affected.[11]

The prognosis to a large extent, is dependent on the severity of renal involvement.[14-16] Children in >6 years age group had more chance of developing renal involvement similar to study by Nickavar et al and Trnka et al.[17,18] According to Nickaver the mean age at presentation of nephritis was 87.4±30.9 months, which was significantly higher than the age of those without nephritis.[17] Age at presentation was the only predictor of renal involvement in their series. Renal involvement in our study was less compared to other studies.[19]

Corticosteroids are effective in treatment of abdominal pain, arthralgia, and purpura. Clinicians are advised to use their discretion in choosing which patients might benefit most from oral corticosteroids.[20] Early prednisone treatment does not affect the outcome and was not be routinely used in several studies similar to our series.[21,22]

Randomised controlled trials have found no statistically significant relationship between early prednisone treatment and a decrease in abdominal complications, namely hospitalization, intussusception, or surgery.[23-25]

Central nervous system involvement is rare in the disease and was absent in our study. In a study by Lava et al HSP had central nervous system involvement presenting as Posterior Reversible Encephalopathy Syndrome (PRES), as a result of central nervous system vasculitis or arterial hypertension.[26]

CONCLUSION

Spontaneous recovery is seen even in patients with severe initial presentation. Renal involvement was less common. Corticosteroids seem to have a role in the symptomatic management of HSP, specifically in treating abdominal pain, arthralgia, and purpura.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Abbas S, Geetha S, Deepthi RV, Kamar J, Uthup S. Clinical profile and outcome of Henoch Schonlein purpura in a tertiary care hospital in South India. Int J Contemp Pediatr 2017;4:822-6.