Clinico epidemiological profile of Henoch-Schonlein purpura in children: A retrospective study

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ABSTRACT

Background: The incidence of Henoch-Schonlein purpura (HSP), the most common systemic vasculitis in children, is steadily increasing. It is an immunoglobulin A (IgA) mediated systemic small-vessel vasculitis. HSP is now referred to as IgA vasculitis as per the Chapel Hill Consensus Conference on the Nomenclature of Systemic Vasculitides in 2012. **Objective:** The objective of the study was to study the clinical profile, pattern of organ involvement, treatment modalities, and outcomes of HSP. **Materials and Methods:** A retrospective study was done from January 2016 to June 2018 using the medical records of our institute. During the study period, 27 children were diagnosed with HSP. The age, gender, clinical presentation, and laboratory data including complete blood count, C-Reactive protein, erythrocyte sedimentation rate, and complete urinalysis, USG abdomen, and recurrence were studied. **Results:** The ratio of male:female was 3:1. Palpable purpura was present in all the patients and was the initial presentation reported in most of the cases. Arthritis was seen in 44% and abdominal pain in 66% of the cases. Renal manifestations in the form of hematuria were reported in 22% and proteinuria in 11% of the patients. Recovery with conservative management was seen in two patients, whereas others required steroids. **Conclusion:** HSP is a common vasculitis seen in childhood and it might have the risks for various complications.

Key words: *Henoch-Schonlein purpura, Immunoglobulin A vasculitis, Scrotal edema in Henoch-Schonlein purpura*

Vasculitis is rare in children [1]. Henoch-Schonlein purpura (HSP) followed by Kawasaki disease is the most common systemic vasculitis in the pediatric population [2]. The American College of Rheumatology classification of vasculitis (ACR criteria 1994) did not include children [3]. However, still, the ACR criteria were used for many years for diagnosis of vasculitis in children. In 2005, the vasculitis working group of the pediatric rheumatology European society (PRES) proposed preliminary classification criteria [4]. Chapel Hill Consensus Conference on the Nomenclature of systemic vasculitides in 2012 renamed HSP to immunoglobulin A vasculitis, due to the deposition of IgA in the vessel wall which is pathognomonic [5]. It affects the small blood vessels of the skin and gastrointestinal (GI) tract, kidney, and joints.

HSP is a benign and self-limiting disease with an excellent prognosis. It takes around 4 weeks for recovery in two-thirds of patients. The disease may recur within the first 4 weeks to as late as 2 years after onset in a third to half of the cases. Although the clinicopathological profile of HSP has been studied before, data from the eastern part of our country are lacking. Hence, this study was planned to evaluate the clinico-epidemiological profile, response to the available therapeutic agents and complications of the disease.

MATERIALS AND METHODS

A retrograde review of the medical records of 35 children diagnosed with HSP, from January 2016 to June 2018, was performed at Kalinga Institute of Medical Sciences. A total number of 35 patients of purpura were identified from the records. The inclusion criteria were children with palpable purpura in the absence of coagulopathy and thrombocytopenia with the presence of at least one of the following features such as diffuse abdominal pain, any biopsy showing predominant IgA deposition, arthritis or arthralgia, and renal involvement in the form of hematuria and/or proteinuria. Purpura associated with other collagen vascular disorders, incomplete medical records, incorrect coding and cases of more than 1 month after initial diagnosis at another hospital were excluded from the study.

Out of the 35 patients, eight were excluded from the study; two had presented more than 1 month after initial diagnosis at another hospital, the medical record was incomplete in three patients, purpura was due to other collagen vascular disorder in two cases, and there was incorrect coding in one patient. Diagnosis of HSP was based on criteria laid by the European League Against Rheumatism endorsed consensus criteria for HSP proposed by Ozen *et al*. [4]. The criteria include palpable purpura which is a mandatory criterion with the presence of at least one of

the following features such as diffuse abdominal pain, any biopsy showing predominant IgA deposition, arthritis or arthralgia, and renal involvement in the form of hematuria and/or proteinuria.

Abdominal pain was grouped into two types among which one category included severe pain in abdomen requiring a radiological examination to exclude other surgical causes and the other one being mild pain in abdomen. Renal involvement included hematuria (>5 red blood cells/hpf) and proteinuria (urine protein creatinine ratio, Upr:Ucr >0.2). In the included patients, the data collected included age, gender, clinical presentations, and findings. Laboratory data including complete blood count, C-reactive protein, erythrocyte sedimentation rate (ESR), complete urinalysis, ultrasound abdomen, and duration of steroid treatment. The treatment was given according to the severity of the disease with oral or injectable steroids or immunosuppressive drugs. Response to steroid treatment was analyzed in 25 patients who were followed up for 6 months period, after exclusion of two patients who did not merit steroid treatment.

All statistical analysis was performed with SPSS version 18.0. Continuous variables were expressed as mean±standard deviation. Categorical variables were expressed as a percentage. Fisher's exact test was used to evaluate the difference between responses to oral and injectable steroids. $p<0.05$ was considered statistically significant.

RESULTS

In the 2 ½ years study duration, 27 patients were included in the study. The average age of onset of disease was 7.3 (7.27±2.65) years, the youngest patient being 2 years. Male:female ratio was 3:1. Rashes were present in all the 27 cases (100%), and buttocks and lower limbs were the first to be affected. The time of onset of rashes was different. Rashes were the initial presenting clinical picture in 13 cases. In seven cases, fever (25%) and in seven others, and pain abdomen (25%) were noted before the appearance of rashes. In two cases, rashes were the only mode of presentation. The details are given in Table 1.

GI symptoms were seen in 8 patients (29%) which included vomiting (14%), organomegaly (14%), and scrotal swelling (7%). When the laboratory parameters were assessed (Table 2), the mean platelet count was found to be 285,000 (240,000–540,000) L/mm^3 and mean hemoglobin level was 7.5 (8.8–13 gm/dl). Antinuclear antibody and rheumatoid factor were done only in 20 patients out of which each one had single positive report. Antistreptolysin O titer of >200 was found in 8 (29%) patients. Abnormal urine analysis was noticed in 9 (33%) patients. Ultrasound abnormalities in the form of hepatosplenomegaly, jejunal thickening, mesenteric adenitis, collection of fluid in the pouch of Douglas were seen in 6 (22%). In one patient (3%), intussusception was also diagnosed.

In 12 patients, oral steroid was given at the dose of 2 mg/kg for 2 weeks in tapering doses while two patients did not require any medication and recovered only with symptomatic management. Injectable steroids were required in 13 cases (Dexamethasone in 9 and methyl prednisolone in 4 patients) because of severe pain abdomen, persistent vomiting, unable to take oral steroid, and extensive rashes. Methyl prednisolone was given in the dose of 30 mg/kg/day for 3 days followed by oral steroids. Similarly, dexamethasone was given in the dose of 0.14 mg/kg/dose in divided doses for 5 days following which it was gradually tapered over 5 days. The details are given in Table 3.

Symptomatic improvement was seen in 83.3% cases with oral steroid within 72 h in comparison to injectable steroids in 61.5%. However, this difference is not statistically significant (p=0.378).

Table 1: Clinical profile

Clinical features	Frequency (n=27)	Percentage
Rashes	27	100
Abdominal pain	18	66
Vomiting	4	25
Hepatomegaly	2	7
Splenomegaly	2	7
Arthritis	12	44
Edema	13	48
Hypertension	1	3

DISCUSSION

HSP is a small-vessel non granulomatous vasculitis. Deposition of IgA in the vessel wall, which is typical to this disease, leads to involvement of skin, joints, intestines, and kidneys. Children between 3 and 15 years are usually affected with male preponderance (male:female=1.5:1) [6,7]. The incidence is 12–53 cases per 100,000 under 17 years.

The main clinical presentation of HSP is rash, pain abdomen with GI symptoms, joint pain, and sometimes renal involvement. In the present study, classical palpable purpura which is predominately seen in the lower extremities and buttocks was

Table 2: Laboratory parameters

Laboratory parameter	n	Frequency	Percentage
Leukocytosis	27	21	77
High erythrocyte sedimentation rate	27	13	48
Anemia	27	8	29
Thrombocytosis	27	8	29
Antistreptolysin O (positive)	20	8	29
Antinuclear antibody	20	1	3
Rheumatoid factor	20	1	3
Hematuria	27	6	22
Proteinuria	27	3	11
Abnormal ultrasound abdomen	27	6	22

Table 3: Comparison between the oral and injectable steroids

Mode of treatment	Response <72 h (%)	Response >72 h	p value
Oral steroids	10 (83.3)	2 (16.7)	0.378
Injectable steroids	8 (61.5)	5 (38.5)	

observed in all the cases. Similar results were observed by Kumar *et al.* [8,9]. Joint involvement, including arthritis and arthralgia is the second common presentation, occurring in up to 75% of the children with HSP. It was observed in 44% of the patients in our study and mainly affected ankle and knee joints.

The classical GI manifestations of HSP consist of nausea, vomiting, diarrhea, constipation, abdominal distension, periumbilical colicky, and abdominal pain due to edema and bleeding. Rare GI manifestations might include intestinal perforation, intussusceptions, esophageal ulcers, and pancreatitis which might require surgical intervention. According to a study by Choong and Beasley, the incidence of pain abdomen was 14–36% and in 11% cases, abdominal symptoms preceded the typical purpuric rashes. The incidence of intussusceptions was 4.6% [9]. In our study, only one (3%) patient developed intussusceptions for which surgical consultation was taken.

Various studies have reported scrotal involvement in HSP cases. The reported incidence of scrotal involvement ranges from 2 to 38%. Chao *et al*. reported 10% of acute scrotum at presentation [10]. In two of our cases, scrotal swelling was seen. Rashes and swelling in the joints developed 4–5 days after the scrotal swelling. Acute scrotal involvement might include scrotal rash and edema of scrotal soft tissue. It might be either unilateral or bilateral, and this pain mimics testicular torsion, though true torsion is rare. Hara *et al*. performed surgical exploration in 11 of their 25 HSP cases and did not identify testicular torsion in any of these patients [11].

Renal involvement was defined as the presence of gross or microscopic hematuria with or without proteinuria. The incidence of renal involvement varies from 10 to 50% with end-stage renal disease in 5%. Severe renal involvement might land up in long-term morbidity and mortality of HSP. Lardhi found renal involvement in 19 out of 78 (24%) patients [12]. In a study by Shah, the incidence of renal involvement was found to be 27% [13]. The outcome depends entirely on the extent of renal involvement. In our study, renal involvement was less in comparison to other studies.

Leukocytosis and elevated ESR are the usual hematological findings in HSP. The distinguishing feature from other forms of purpura is thrombocytosis [14]. The current study shows 29% thrombocytosis which correlates well with the other studies. Leukocytosis and elevated ESR were seen in 77% and 48%, respectively. In our study, none of the patients had central nervous system (CNS) involvement. Although this involvement is rare in HSP, in a study by Lava *et al.*, CNS involvement presenting as posterior reversible encephalopathy syndrome (PRES) was mentioned, as a result of CNS vasculitis or arterial hypertension [15].

Supportive measures might be helpful in some of the patients with HSP. Corticosteroids help in reducing the duration and severity of abdominal and joint pain but, it does not alter the natural course of the disease. In general, prednisolone is a commonly used steroid for the treatment of HSP. Although 9 patients required injection dexamethasone for the treatment in our study, there is no evidence in literature to prove the superiority of one over the other. High dose steroid, azathioprine, cyclophosphamide, mycophenolate mofetil, intravenous Ig, and plasmapheresis might be needed in case of severe HSP nephritis [16,17].

The study had certain limitations. This was a retrospective study with small sample size. The involvement of multiple clinicians in the treatment prevented a standardized steroid therapy with respect to the type of the steroid and their dosage used. Second, most parents did not consent for the biopsy as it was an invasive procedure.

CONCLUSION

HSP appears to be common in the eastern part of our country. Spontaneous recovery with symptomatic management is also seen. The severity as well as involvement of renal system was less marked in our study. Corticosteroids seem to have a role in the management of HSP, but it does not alter the natural course of the disease. There is a requirement for large scale prospective study to establish the standardized treatment protocol for HSP.

REFERENCES

1. Dedeoglu F, Sundel RP. Vasculitis in children. Rheum Dis Clin North Am 2007;33:555-83.
2. Behrman RE, Kliegman RM, Arvin AM. Nelson Textbook of Pediatrics. 20th ed. Philadelphia, PA: Saunders; 2008. p. 1043-5.
3. Ozen S, Pistorio A, Iusan SM, Bakkaloglu A, Herlin T, Brik R, *et al.* EULAR/PRINTO/PRES criteria for henoch-schönlein purpura, childhood polyarteritis nodosa, childhood wegener granulomatosis and childhood takayasu arteritis: Ankara 2008. Part II: Final classification criteria. Ann Rheum Dis 2010;69:798-806.
4. Ozen S, Ruperto N, Dillon MJ, Bagga A, Barron K, Davin JC, *et al.* EULAR/PReS endorsed consensus criteria for the classification of childhood vasculitides. Ann Rheum Dis 2006;65:936-41.
5. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, *et al.* 2012 revised international chapel hill consensus conference nomenclature of vasculitides. Arthritis Rheum 2013;65:1-11.
6. Yang YH, Hung CF, Hsu CR, Wang LC, Chuang YH, Lin YT, *et al.* A nationwide survey on epidemiological characteristics of childhood Henoch-Schönlein purpura in Taiwan. Rheumatology (Oxford) 2005;44:618-22.
7. Peru H, Soylemezoglu O, Bakkaloglu SA, Elmas S, Bozkaya D, Elmaci AM, *et al.* Henoch schonlein purpura in childhood: Clinical analysis of 254 cases over a 3-year period. Clin Rheumatol 2008;27:1087-92.
8. Kumar L, Singh S, Goraya JS, Uppal B, Kakkar S, Walker R, *et al.* Henoch-schonlein purpura: The Chandigarh experience. Indian Pediatr 1998;35:19-25.
9. Choong CK, Beasley SW. Intra-abdominal manifestations of Henoch-Schönlein purpura. J Paediatr Child Health 1998;34:405-9.
10. Chao NS, Wong BP, Leung MW, Chung KW, Kwok WK, Liu KK. Acute gastrointestinal and genito-urinary manifestations in children with Henoch-Schönlein purpura. HK J Paediatr (New Series) 2009;14:168-71.
11. Hara Y, Tajiri T, Matsuura K, Hasegawa A. Acute scrotum caused by henoch-schönlein purpura. Int J Urol 2004;11:578-80.
12. Lardhi AA. Henoch-schonlein purpura in children from the Eastern province of Saudi Arabia. Saudi Med J 2012;33:973-8.
13. Shah G. Clinical profile and pattern of Henoch-Schönlein purpura in children. J Patan Acad Health Sci 2015;2:17-21.
14. Evans-Jones LG, Glough JV. Thrombocytosis in Henoch-Schonlein purpura. Clin Lab Hematol 1990;12:137-9.
15. Lava SA, Peeters GG, Bianchetti MG, Goeggel Simonetti B, Simonetti GD,

Milani GP, *et al.* Posterior reversible encephalopathy syndrome in Henoch-Schönlein purpura. Rheumatol Int 2017;37:461-3.
16. Chartapisak W, Opastiraku S, Willis NS, Craig JC, Hodson EM. Prevention and treatment of renal disease in Henoch-Schönlein purpura: A systematic review. Arch Dis Child 2009;94:132-7.
17. Du Y, Hou L, Zhao C, Han M, Wu Y. Treatment of children with Henoch-Schönlein purpura nephritis with mycophenolate mofetil. Pediatr Nephrol 2012;27:765-71.

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