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Clinical Profile and Pattern of Henoch-Schonlein Purpura in Children in Kashmir

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Abstract

Objective: *This study was done to evaluate clinical profile of Henoch-schonlein purpura (HSP) in children in Kashmir.*

Material And Methods: *This study was conducted from August 2010 to July 2011 at G. B Pant Children Hospital which is tertiary care hospital and is associated hospital of Government medical college Srinagar India. The study group included were all children in the age group of 1-18 years who fulfilled diagnostic criteria for Henoch-Schonlein purpura. Diagnosis of HSP was made by using European League against Rheumatism (EULAR) and Pediatric Rheumatology Society classification. The criteria included: Palpable purpura together with at least one of the following finding: Diffuse abdominal pain, Predominant IgA Deposition, Acute arthritis in any joint, Renal involvement. We checked CBC, ESR, Urine examination, KFT, Serum electrolytes, ASO, USG abdomen and skin biopsy.*

Results: *Total number of patients diagnosed was 27. Among these cases 17 were males and 10 were females. 14 cases were from urban areas and 13 cases were from rural areas. Maximum number of cases was seen between November and April. The pattern of clinical features were in the form of purpuric rash 17 (100%), Abdominal pain 18 (66.6%), arthritis/arthralgias 16 (59.26%), HSP nephritis 9 (33.3%), G I bleed 8 (29.6%). Thrombocytosis was present in 66.6% cases, ASO titre was raised in 37%, ESR was raised in 55.5%, CRP in 40.7% and 29.6% cases had proteinuria.*

Conclusion: *We conclude that HSP in Kashmir is as common as in other parts of the world with pattern and clinical features almost similar as revealed by other authors but because smaller sample size further studies need to be done over extended period of time to have statistically more significant results.*

Keywords: *Henoch-scholein purpura, Children, pattern, clinical profile,*

I. Introduction

Henoch-Schonlein purpura is most common vasculitis seen in children. It is also known as Anaphylactoid purpura or purpura rheumatica and Schonlein-Henoch purpura[1]. The exact cause of Henoch-Schonlein purpura (HSP) is unknown, although it may occur after certain viral and bacterial infections, as well as adverse drug reactions to some medications. HSP occurs more often in children than in adults, and usually follows an upper respiratory infection. Half of the affected patients are below the age of six, and 90% occur under 10 years of age. It occurs about twice as often in boys as in girls. Typical symptoms include palpable purpura, joint pains and abdominal pain. Most cases are self-limiting and require no treatment apart from symptom control, but the disease may relapse in a third of cases and cause irreversible kidney damage in about one in 100 cases[2]. The incidence of HSP in children is about 9 per 100,000 children per year; this makes it most common vasculitis in childhood[3]. HSP is a systemic vasculitis, most of its features are due to the deposition of abnormal antibodies in the wall of blood vessels, leading to vasculitis. These antibodies are of the subclass IgA_1 in polymers; it is uncertain whether the main cause is overproduction (in the digestive tract or in the bone marrow) or decreased removel of abnormal IgA from the circulation[4].

Multiple standards exist for defining HSP. These include the 1990 American College of Rhematology (ACR) classification [5]and the 1994 Chapel Hill Consensus Conference (CHCC)[6]. The ACR classification criteria, of which two are necessary to make diagnosis are; age < 20 years at onset, Palpable Purpura, Bowel angina (diffuse abdominal pain or bowel ischemia usually with bloody diarrhea) and Biopsy evidence of granulocytes in the walls of arterioles or venules. A more recent classification is the 2006 European League Against Rhematism (EULAR) and Pediatric Rheumatolgy Society (PRS) Classification[7]. This classification recommends; Palpable purpura as a mandatory criteria, together with at least one of the following findings; diffuse abdominal pain, predominant IgA deposition (confirmed on skin biopsy), acute arthritis and renal involvement (as evidenced by the presence of blood and/or protein in the urine). The diagnosis is based on the combination of the symptoms, as very few other diseases cause the same symptoms together. Blood tests may

show elevated creatinine and urea levels, raised IgA levels (in about 50%)[8], raised C-reactive protein (CRP) or erythrocyte sedimentation rate (ESR) results; none are specific for HSP[9]. If there is doubt about the cause of the skin lesions, a biopsy of the skin may be performed to distinguish the purpura from other diseases that cause purpura, such as vasculitis due to cryoglobulinemia. However, overall serum complement levels are normal[10]. Overall prognosis is good in most of the patients, with one study showing recovery occurring in 94% and 89% of children and adults, respectively[11].

II. Material And Methods

This study was conducted in G. B Pant children Hospital Srinagar which is a tertiary care hospital and is associated hospital of Government Medical College Srinagar India. This study was a prospective study and was conducted from August 2010 to July 2011. The study group included all children in the age group 1 to 18 years who fulfilled diagnostic criteria for HSP. All the patients diagnosed as HSP were included in this study. Informed consent was taken from the parents/guardians of all patients. Detailed history was recorded with emphasis on onset of rash, type of rash, location of rash, its progression or regression, itching or not, joint pain and/or swelling, location of joint involved, abdominal pain, type and localization of pain, history of any blood and/or mucus in stools, history of dark colored urine, hematuria, any change in sensorium or any convulsions. General physical examination was recorded on a predesigned proforma which included anthropometric data like weight, height, head circumference, chest/mid upper arm circumference, vitals like pulse, respiratory rate, temperature, blood pressure and physical signs like palpable purpura, peri-orbital puffiness, edema of scalp, peno-scrotal area, feet or legs, arthritis and other significant findings. Hypertension was defined as an average systolic or diastolic blood pressure $\geq 95^{th}$ percentile for age, sex and height. HSP nephritis (HSN) was defined as the presence of gross or microscopic hematuria with or without proteinuria. Hematuria was defined as small amount (+) of haemoglobin on dipstick testing or >5 red blood cells per high power microscopic field in a centrifuged specimen. Proteinuria was defined as small amount of protein (+) on dipstick testing or proteinuria >4 mg/m^2/hour obtained from 24-hour collected urine. Nephritic and Nephrotic syndrome were defined using standard criteria. Systemic examination of chest, CVS, abdomen, CNS, and Skeletal system was done. All patients underwent investigations like CBC, ESR, Urine examination, KFT, Serum electrolytes and ASO. USG Abdomen was done in those cases that had abdominal complaints. Skin biopsy was done in all cases and biopsy was subjected to light microscopy and direct immunoflourence study.

III. Results

The results of this study showed total of 27 cases of HSP over a period of one year in our setup.

Table-1 Gender Distribution of cases.

SEX	Total number of cases	%
Male	17	62.9
Female	10	37
Total	27	100

As depicted in above table, there were 27 cases, out of which 17 (62.96%) were males and 10 (37.04%) were females. Our study showed that maximum number of cases 14 occurred in the age group of 7-12 years with 9 males and 5 females giving ratio of 1.8:1 compared to other groups.

Table-2 Distribution of cases as per residence

Residence	Number	%
Rural	13	48.14
Urban	14	51.8
Total	27	100

Above table shows that most of cases were from urban area than rural area with a ratio of 1.07:1.
In our study there were seasonal distribution of cases with 22 (81.4%) cases occurring between November and April. Highest number of cases 6 (22.23%) were seen in March and there were no cases seen in June and July.

Table-3 Monthly distribution of cases (n=27).

Month	Number	%
January	4	18.81
February	3	11.11
March	6	22.23
April	4	14.81
May	1	3.71
June	0	0
July	0	0

August	1	3.71
September	2	7.40
October	1	3.71
November	2	7.40
December	3	11.11
Total	27	100

Purpuric rash was most common presentation and was present in all 27 cases, abdominal pain was present in 18 cases, arthritis/arthralgias in 16, HSP nephritis in 9 cases, Gasrtointestinal bleeding in 8 cases.

Table-4 Clinical presentation of HSP.

Clinical Feature	Number	%
Purpuric rash	27	100
Abdominal pain	18	66.66
Arthritis/arthralgias	16	59.26
HSP nephritis	9	33.33
G.I bleed	8	29.62
Others	6	22.22

Among laboratory findings thrombocytosis was present in 66.6% of patients, ASO titre was raised in 37%, ESR was raised in 55.5%, CRP in 40.7% and 29.6% patients had proteinuria.

Table-5 Laboratory findings among HSP patients

Laboratory test	Total number	%
Thrombocytosis	18	66.6
ASO titre>200 IU/ml	10	37
ESR(raised)	15	55.5
CRP(raised)	11	40.7
KFT(abnormal)	5	18.5
Hematuria	4	14.8
24 hour urinary protein(>4mg/m^2/hour)	8	29.6

Table-6 Skin biopsy changes in HSP

Skin biopsy change	Tatal	%
Leukocytoclastic changes	20	74
IgA±C_3 Deposits	14	51.8
Leukocytoclastic changes±IgA Deposits	25	92.59

Table-7 Correlation of raised ASO titre in HSP patients with positive skin biopsy

	ASO Titre>200 IU/ml	ASO Titre<200 IU/ml	Total
Skin biopsy (+ve)	9	16	25
Skin biopsy (-ve)	1	1	2
Total	10	17	27

This correlation has sensitivity of 90% and specificity of 5.8% with p value=0.076

IV. Discussion

Our study revealed that out of 27 patients there were 17 (62.9%) males and 10 (37%) females with a ratio of 1.7:1. Similar results were obtained by Cakir Murat et al[12] who studied 116 children out of which 73 (63%) were males and 43 (37%) were females; Lata Kumar et al[13]studied 46 patients and found M:F ratio of 2:1 which is almost similar to our results. In our study, out of 27 cases 11 (40.7%) were in the age group of 1-6 years, 14 (51.8%) were in age group 7-12 years and 2 (7.4%) were in age group of 13-18 years. The mean age of distribution was 7.33 (3.07) Almost similar results were seen by A Bagga, et al[14] who studied 47 cases of HSP and found that the mean age at onset was 8.5 years. Assar Shideh et al[15] found that out of 63 cases the mean age of the cases was 6.4 (±3.15) years.

Our study demonstrated a seasonal variation with 22 (81.4%) cases seen between November and April (winter months). Highest number of cases 4 (14.8%) seen in April and there was no case seen in June and July. Similar results were observed by Y H Yang [16] who found that disease onset was more common in autumn and winter months, Maher Khader et al[17] found distribution of admission was highest in winter months. In our study purpuric rash was the most common presentation and was present in all 27 cases, similar results were observed by L Kumar et al[18]. Abdominal pain was present in 18 (66.6%) cases. Saulsbury FT[19] found that abdominal pain was present in 63% of patients. In our study arthritis/arthralgias was present was present in 16 (59.26%). Lata Kumar et al[20] found large joint arthritis occurred in 60% of cases. In our study HSP nephritis occurred in 9

(33.3%) of cases. Similar results were seen by Lata Kumar et al[21] who found. In our study we found that peno/scrotal edema in 4 (14.8%). Chana Ophir Mintzer, et al[22] studied scrotal involvement in 86 children over a 20 year period and found similar results. Intussusception was seen in one patient in our study. Trapani S et al[23]studied 150 children and found intussuception in one patient. Our study demonstrated thrombocytosis in 66.66% of patients, ASO titre was raised in 37.03%, ESR was raised in 55.55%, CRP in 40.7%, 29.62% patients had proteinuria, 5 (18.5%) had abnormal KFT and 4 (14.8%) had hematuria. Trapani S et al[24] studied 150 children with HSP and found most frequent laboratory abnormalities were high ESR (57%), hyper-IgA (37%), and proteinuria (42%). Maher Khader MD et al[25] studied 30 patients and found ASO titre >200IU/ml in 4 (36.36%) out of 11 patients tested.

In our study 25 patients had positive skin biopsy findings, 20 patients had leukocytoclastic changes and 14 patients had IgA±C_3 deposits. Lata Kumar, et al[26] studied skin biopsy changes in 36 patients and found 34 had histological (Leukocytoclastic changes, n=27) and immunofluorescent changes (deposition of IgA with/without C_3 deposits in dermal capillaries, n=18).

V. Conclusion

Our study reveals that HSP in Kashmir is as common as in other parts of the world with pattern and clinical features almost similar as revealed by other authors but because of smaller sample size further studies need to be done over extended period to have statistically more significant results.

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