

HENOCH- SCHONLEIN PURPURA WITH MULTIPLE RECURRENCES**Dr. Saiprasad Onkareshwar Kavthekar***

Associate Professor, Department of Pediatrics, DD.Y. Patil Medical College and Hospital, D. Y. Patil Education Society (Institution Deemed to be University), Kolhapur (416006), Maharashtra, India.

***Corresponding Author: Dr. Saiprasad Onkareshwar Kavthekar**

Associate Professor, Department of Pediatrics, DD.Y. Patil Medical College and Hospital, D. Y. Patil Education Society (Institution Deemed to be University), Kolhapur (416006), Maharashtra, India.

Article Received on 07/08/2019

Article Revised on 28/08/2019

Article Accepted on 18/09/2019

ABSTRACT

Henoch-Schonlein Purpura (HSP) is characterised by presence of palpable, non-thrombocytopenic purpura, arthritis and arthralgia, abdominal pain, gastrointestinal haemorrhage and /or nephritis. HSP is typically a childhood disorder, self-limited, benign disease and distinctly less common in adults, in whom severe and chronic complications are often encountered. Approximately one third of patients have at least one recurrence, generally involving cutaneous and abdominal manifestations, especially during first two years period after the initial outbreak. Studies regarding recurrences are very few and incidence of recurrence reported from 2.7% to 51.7% in various studies. Here I report a case of 16 years boy who presented with typical clinical presentation of HSP with initial episode and subsequent six recurrences over the next 3 years with milder clinical presentation than initial episode except one severe recurrence episode which presented with gastrointestinal tract manifestations in the form acute abdominal pain and hematemesis. Since last one year, this young boy is totally asymptomatic.

KEYWORDS: Henoch-Schonlein purpura, recurrence, vasculitis.**INTRODUCTION**

Henoch- schonlein purpura (HSP) is an IgA immune complex mediated leukocytoclastic small vessel vasculitis and the most commonly affected organs are small vessels in the skin, joints, gastrointestinal tract and kidneys.^[1] HSP occurs worldwide and affects all ethnic groups but is common in white and Asian population. The incidence of HSP is estimated at 14-20/100000 children per year and affects males more than females.^[1] HSP is typically childhood disorder, self-limited, benign disease and distinctly less common in adults, in whom severe and chronic complications are often encountered.^[1] HSP is characterised by presence of palpable, non-thrombocytopenic purpura, arthritis and arthralgia, abdominal pain, gastrointestinal haemorrhage and or nephritis.^[1,2]

Recurrences are common in HSP and approximately one third of patients have at least one recurrence, generally involving cutaneous and abdominal manifestations, especially during a two year period after the first outbreak.^[3] However studies regarding recurrences are very few and incidence of recurrence reported from 2.7% to 51.7% from various studies.^[4,5,6] Recurrence of HSP is defined as the presence of a fresh episode after a period of at least three months without signs and symptoms. Chronic HSP is defined as when cutaneous, gastrointestinal and or renal manifestations persisted for a continuous period of 12 months or more.^[6]

Here I report a case of HSP who initially presented with petechial-purpuric rash, arthritis, abdominal pain and urinary microscopic abnormalities and responded well to the treatment. Subsequently he presented with multiple recurrences over the next three years with milder clinical presentation with each recurrence than initial episode except one severe episode.

CASE**Initial Episode (10th November 2015)**

16yrs boy presented in the Out Patient Department of Pediatrics, D.Y. Patil Medical College and Hospital, Kolhapur with acute abdominal pain, bilateral ankle joint swelling and petechial to purpuric rash on both legs, buttocks and posterior aspect of forearm since last 2 days. On examination, he was afebrile, pulse 72bpm, B.P:120 /70 mm of Hg and respiratory rate 20/minute Per abdomen examination showed tenderness in umbilical region and no organomegaly and rest of the systemic examination was normal. His complete blood counts showed HB14.6gm/dl, WBC13600 cells/cmm, N 81%, L 19%, Platelet count 2480000/cmm, Bleeding time, Clotting time and Prothrombin time were normal. Blood Urea was 46mg/dl and Serum Creatinine was 0.70mg/dl. Urine examination showed protein 2+, Pus cells 18-20/ hpf and RBC24-25/ hpf. Ultrasonography abdomen showed edematous bowel loops. In view of typical clinical finding and investigations, he was diagnosed as HSP and treated with intravenous fluids for

two days and oral prednisolone 1 mg / kg / daily in three divided doses for four weeks and tapered over next two weeks and then stopped. His abdominal pain, joint swelling resolved over next two days and rash disappeared over next three weeks. But urinary microscopic abnormalities persisted for almost four weeks. At that time, we did renal biopsy to rule out HSP glomerulonephritis but which was normal. We monitored his renal function and microscopic urinary abnormalities for next one month and his urinary microscopic abnormalities became normal over next two weeks.

1st recurrence (31st March 2016)

This boy again presented with petechial lesion on legs and few purpuric lesions on buttocks with abdominal pain and few urinary microscopic abnormalities without any arthritis. He was treated with oral prednisolone 1mg/kg/ day for 5 days and tapered over next 5 days. His symptoms relieved and rash disappeared over 1 week.

2nd Recurrence (7th August 2016)

This time he presented with few petechial lesions on legs and bilateral ankle arthritis. He was treated only with non steroidal anti inflammatory drug (NSAID) and did not receive oral steroids. His rash and arthritis relived over one week duration.

3rd Recurrence (14th December 2016)

He had minor petechial lesions on legs and no other system involvement. This time we observed for progress in petechial lesion and other system involvement and treated only with NSAID, if needed. The petechial lesions regressed over five days.

4th Recurrence (7th July 2017)

He presented with complaints of acute epigastric pain with frank hematemesis since 1 day and without any petechial rash, arthritis or urinary microscopic abnormalities. Complete blood counts showed HB 11.6%, WBC 12770 cells/cmm, platelets 237000/cmm, blood Urea 25mg/dl and bleeding time, clotting time, and prothrombin time was normal. Urinary microscopic examination were normal. Ultrasound abdomen examination showed stomach wall thickening with irregular mucosa, inflamed small intestine and both the kidneys were normal. He was treated with intravenous fluids, injection Ranitidine and Injection Hydrocortisone 100mg 12 hourly for 3 days and switched to oral Prednisolone 30mg BID for next 1 week and tapered over next one week and stopped. His acute epigastric pain and hematemesis subsided in 24 hours. Repeat ultrasonography abdomen after one week was normal

5th Recurrence (26th December 2017)

He developed only few petechial lesions on foot and mild ankle swelling without any gastrointestinal manifestations and urinary abnormalities. He was treated with NSAID only for 2 days and he responded very well and petechial lesions disappeared in three days.

6th Recurrence (5th August 2018)

This time, he had few petechial lesions on foot without any other clinical manifestation. We observed the rash for progression carefully and rash disappeared within next 5 days on its own without any treatment.

We are following this young boy, who is presently 20 years old and absolutely asymptomatic, without any recurrence since almost last one year and normal renal functions without any urinary microscopic abnormalities.

DISCUSSION

HSP was first described by Heberden before 1800; in the 1830s, Schonlein described the typical rash and joint manifestations, and in the 1870s Henoch recognized the gastrointestinal and renal manifestations.^[7] American college of rheumatology classification criteria for children and adult include two of the following must be present 1.palpable purpura 2.Age at onset <20 years 3.Bowel angina(postprandial abdominal pain, bloody diarrhoea) 4.Biopsy demonstrating intramural granulocytes in small arterioles and /or venules.^[8] Many cases of HSP follow a documented upper respiratory infection.^[1]

Treatment for mild self-limited HSP is supportive, with an emphasis on assuring adequate hydration, nutrition, and analgesia. Steroids are most often used to treat significant gastrointestinal involvement or other life threatening manifestations.^[1] Overall, the prognosis for childhood HSP is excellent, and most children experience an acute, self-limited course lasting on average four weeks.^[1] Chronic renal disease is the major long term complication, occurring in 1-2% of children with HSP and approximately 8% of those with HSP nephritis go on to have end stage renal disease.^[1]

Children with more severe initial course of HSP are at higher risk for recurrence typically within 4-6 months of first episode.^[1] Byun et al,^[4] described age more than 30 years, the presence of underlying disease, persistent purpura more than one month, abdominal pain and the presence of hematuria as the predictors of recurring HSP. Shin et al^[9] reported severe bowel angina, age more than 10 years, persistent purpura more than one month and leukocytosis as the best predictors for HSP recurrence. Calvo Rio et al^[10] identified joint and gastrointestinal manifestations and no history of previous infection at the time of the diagnosis of HSP as the risk factors for recurrence of HSP. Relapses in children occurred more commonly in those who presented with joint manifestations whereas in adults in those who experienced gastrointestinal manifestations. In addition the protective role of infection against the risk of relapse was mainly seen in children with HSP. Calvino et al,^[11] observed that children who experienced renal sequelae were at greatest risk for recurrences than those patients without renal impairment. Trapani et al^[12] observed an association between the use of corticosteroids at the time of disease diagnosis and the risk of relapses in children.

The risk factors for recurrence in our patient were age more than 10 years, severe initial presentation, no history of previous respiratory tract infection at the time of diagnosis, purpura persisting for 4 weeks and presence of prolonged microscopic hematuria and proteinuria.

The pathogenesis of recurrences in HSP is still unknown. Nathwani D et al,^[13] revealed role of human leukocyte antigen (HLA B35 haplotype) to recurrent HSP. Abnormal auto immune test like RF and ANA were more frequently observed in patients with HSP who experienced recurrences than in those who never had flares of the disease.^[10]

CONCLUSION

HSP may recur multiple times over the years but the clinical manifestations and severity will be milder as compared to initial episode. The children with a more severe and prolonged initial first course are at higher risk for recurrence.

REFERENCES

1. Stacy P, Ardoin, Fels E. Henoch - Schonlein Purpura. In Kliegman R.M, eds. Nelson Textbook of Pediatrics. First south Asia edition, 2016; 2(167.1): 1216-18.
2. Saulsbury FT. Clinical update: Henoch-Schonlein Purpura. Lancet, 2007; 369: 976-78.
3. Saulsbury FT. Henoch-Schonlein Purpura. Curr Opin Rheumatol, 2001; 13: 35-40.
4. Byun JW, Song HJ, Kim L et al. Predictive factors of relapse in adult with Henoch-Schonlein Purpura. Am J Dermatopathol, 2012; 34: 139-44.
5. Prais D, Amir J, Nussinowitch M. Recurrent Henoch-Schonlein Purpura in children. J Clin Rheumatol., 2007; 13: 25-28.
6. Alfredo CS, Nunes NA, Len CA, Barbosa CMP, Terrerri MT, Hilario MO. Henoch-Schonlein Purpura: recurrence and chronicity. J d Pediatr (Rio J), 2007; 83: 177-80.
7. Jane Green Schaller. Henoch - Schonlein Purpura or vasculitis. In Behrman RE, Kliegman R.M, Arvin AM eds. Nelson Textbook of Pediatrics. 15th edition, 1996; 152(1): 676-78.
8. Mills JA, Michel BA, BlochDA, et al. The American college of Rheumatology criteria for classification of Henoch-Schonlein purpura. Arthritis Rheum, 1990; 33: 1114-21.
9. Shin JL, Park JM, Shin YH, et al. Predictive factors for nephritis, relapse and significant proteinuria in childhood Henoch-Schonlein Purpura. Scand J Rheumatol, 2006; 35: 56-60.
10. Calvo-Rio V, Hernandez LJ, Ortiz-Sanjuan F, Loricera J, Palmou-Fontana N, Gonzalez-Vela MC, et al. Relapses in patients with Henoch-Schonlein purpura: Analysis of 417 patients from a single center. Medicine, 2016; 95: 28(e4217).
11. Calvino MC, Llorca J, Gacia-Porrúa C et al. Henoch Schonlein Purpura in children from Northwestern Spain: A 20 year epidemiological and clinical study. Medicine (Baltimore), 2001; 80: 279-90.
12. Trapani S, Micheli A, Grisolia F, et al. Henoch-Schonlein purpura in childhood: epidemiological and clinical analysis of 150 cases over a 5 year period and review of literature. Semin Arthritis Rheum, 2005; 35: 143-53.
13. Nathwani D, Laing RB, Smith CC, et al. Recurrent post-infective Henoch-Schonlein syndrome: a genetic influence related to HLA B35? J Infect, 1992; 25: 205-10.