ORIGINAL RESEARCH PAPER

General Medicine

KEY WORDS:
Henoch–Schönleinpurpura, Children between 2-15 yrs,Renal involvement.

HENOCH SCHONLEIN PUPURA: A RARE CASE REPORT

B .Nadia Shulamite Department of Pharm.D, CMR College of pharmacy, kandlakoya (v) Hyderabad, 501401, Telangana state , India

Dr. G. Ramya Bala Prabha* Department of Pharm.D, CMR College of pharmacy, kandlakoya (v) Hyderabad, 501401, Telangana state , India *Corresponding Author

Sharadha Srikanth Department of Pharm.D, CMR College of pharmacy, kandlakoya (v) Hyderabad, 501401, Telangana state , India

Konde Abbulu Department of Pharm.D, CMR College of pharmacy, kandlakoya (v) Hyderabad, 501401, Telangana state , India

ABSTRACT

INTRODUCTION: Henoch Schonlein purpura (HSP) is the most common vasculitis in children, in whom prognosis is mostly Dependent upon the severity of renal involvement.HSP mainly affects children between 4 to 15 years and has an incidence of 14 in 100,000 population with peak incidence at 5years. It usually involves skin, gut, joints, and kidneys.
Nephritis is observed in about 30% of children with HSP. Renal damage eventually leads to chronic kidney disease in upto 20% of children.HSP nephritis and IgA nephropathy are related diseases resulting from glomerular deposition of aberrantly glycosylated IgA1.
Discussion: A fourteen year old boy ,was diagnosed with Henoch Scholein purpura, IgA vasculitis with leucocytoclistic vasculitis. The patient complained about swelling of both legs since two days, and there was pain in joints , both the knees, ankles, wrist, metacarpal Since two days, rashes over both the legs , the rashes first started over feet ascending over legs and buttocks within a day, history of pain in abdomen, and burning type of pain in epigastrium region, increased frequency of urination. Patient experienced similar complaints four years back. A Clinical diagnosis of Henoch Scholein purpura, IgA vaculitis, leucocyttoclistic vasculitis was done. During the hospital administration, patient was managed symptomatically for pain control, skin rashesand electrolytes balance and corticosteroids were given to reduce the inflammation and thus treating rashes..
Conclusion: HSP is an autoimmune leucocytoclastic vasculitis of childhood which involves skin, gut, joints, and kidneys commonly.Most affected children have been between 2 and 11 years of age. Perioperative management of liver and kidney functions is important.

INTRODUCTION

Henoch Schonlein purpura (HSP) is the most common vasculitis in children, in whom prognosis is mostly dependent upon the severity of renal involvement. Nephritis is observed in about 30% of children with HSP. Renal damage eventually leads to chronic kidney disease in up to 20% of children with HSP nephritis in tertiary care centres, but in less than 5% of unselected patients with HSP, by 20 years after diagnosis.[1]

HSP mainly affects children between 4 and 15 years and has an incidence of 14 in 100,000 population with peak incidence at 5 years.

The main symptom is a rash with numerous small bruises, which have a raised appearance, over the legs or buttocks. It is more common in boys than girls [2]

The symptoms of HSP usually begin suddenly. In addition to the characteristic red spotting of the skin (most often on the buttocks and backs of the legs), they may include headache, loss of appetite, and/or fever.

The skin typically becomes red (diffuse erythema). Cramping abdominal pain may occur and is usually most severe during the night. Blood may be present in the stool and abnormal bleeding (hemorrhaging) from the gastrointestinal tract can cause bloody diarrhea. Joint pain (arthralgia) may develop in any joint of the body, especially the knees and ankles. Some people with HSP experience vomiting and diarrhea; others may have severe constipation and unusually dark stool (melena).

Individuals with HSP typically develop small red or purple spots (petechiae) on the skin, especially on the legs. These purpura spots are caused by small hemorrhages under the skin and are not associated with abnormally low levels of platelets (nonthrombocytopenic) as is common with some other forms of purpura. Other skin lesions associated with this condition include large hives (urticarial wheals) or ulcers (necrotic), especially on the buttocks and legs. [3]

Swelling may occur in the face and neck due to abnormal fluid accumulation in the soft tissues of these areas (angioneurotic edema). In rare cases, swelling and edema in the throat can cause breathing difficulties that can lead to life-threatening respiratory problems.

The exact cause of HSP is not known, although research suggests that this disease may be caused by immune system dysfunction (i.e., increased IgA immune complexes).

Autoimmune disorders are caused when the body's natural defenses against "foreign" or invading organisms (e.g., antibodies) begin to attack healthy tissue for unknown reasons.

In some cases, it has been suggested that this disorder may be an extreme allergic reaction to certain foods, such as chocolate, milk, eggs, or beans. Various drugs (e.g., nifedipine, diltiazem, cefuroxime, diclofenac, etc.), bacteria (e.g., Streptococcus), and insect bites have also been indicated as possible causes in some cases. [4]

In children, the initial symptoms typically begin after the age of 2 years and usually last for about 4 weeks and the disease usually has a somewhat mild course. About 50 percent of affected children experience one or more recurrences, usually within months. The rate of recurrence seems to be higher among those children whose initial disease was more severe.[5]

Most affected children have been between 2 and 11 years of age. In the USA, about 14 cases occur per 100,000 school-aged children. It is generally a benign (non-threatening) disorder appears in most instances to cure itself (self-limiting).

The diagnosis of HSP may be difficult, especially in adults. The

disease is frequently confused with other forms of vascular inflammation (see Related Disorders section of this report). Routine laboratory tests are usually not definitive for the disorder. The platelet count is typically normal although white blood cell and sedimentation rates may be elevated.

The disorder is diagnosed by a combination of the presence of skin lesions and/or joint tenderness, combined with a confirmed test for blood in the urine (urinalysis), and a skin biopsy that shows inflammation of the arterial and venous capillaries [6].

Therapy

If individuals are thought to have HSP as the result of an allergic reaction, they must strictly avoid the offending substance (e.g., food or drug). When evidence of streptococcal infection is present, antibiotic therapy is prescribed. Mild childhood cases of the disease often improve spontaneously with advancing age. There is no specific treatment, however, in most patients, the disease has a limited course and the outlook for recovery is good.

If non-steroid anti-inflammatories fail to relieve symptoms, some patients may be treated with glucocorticoids (steroid) drugs such as prednisone. These drugs may be useful to help control acute abdominal and joint pain. In some cases, swelling of soft tissues (angioedema) may be helped with steroid drugs. Dapsone may be prescribed when prednisone is contra-indicated or fails to relieve symptoms. The use of steroids to treat this disorder remains a matter of controversy in the medical literature. Some research indicates that steroids do not shorten the length of the illness or reduce the frequency or recurrence of symptoms. Other studies indicate that early steroid treatment may help to reduce the risk of kidney damage. [7][8]

Patients with HSP who have advanced kidney disease and renal failure will probably benefit from mechanical cleansing of the waste products from the blood (hemodialysis). Aggressive and supportive care may be necessary during acute kidney crisis. Some patients with severe kidney disease have undergone kidney transplantation. However, the disease can recur in the transplanted kidney. [9]

CASE PRESENTATION

A fourteen year old boy, was diagnosed with Henoch Scholein purpura, IgA vasculitis with leucocytoclistic vasculitis. The patient complained about swelling of both legs since two days, and there was pain in joints, both the knees, ankles, wrist, metacarpal Since two days. And patient also complained about having painful rashes, red skin, lesions over both the legs ,multiple lesions of variable sizes, over both upper and lower limbs, abdomen associated with nocturnal itching. The rashes first started over feet ascending over legs and buttocks within a day. The patient had a history of pain in abdomen, and burning type of pain in epigastrium region. And also patient experienced increased frequency of urination. Patient experienced similar complaints four years back. The patient was admitted in general medicine ward in tertiary care hospital. The laboratory investigations revealed that there was pus cells (3-4 hpf) and trace albumin and the urine was yellow color. And ultra sonography scan of abdomen was done the impression was found to be mild ascites and the complete blood picture was found to be normal. (Albumin has an important role in binding calcium, bilirubin and many drugs. A reduction in serum albumin will increase free level of agents which are normally bound and adverse effect can result if the "free" entity is not rapidly cleared from the body will cause edema [9].Mild ascites was seen which will be seen generally when there is less albumin level and further with Ultrasonography was confirmed. Serum Creatinine levels were slightly decreased, T.S.Bilirubin, D.Bilirubin, Alk phosphatase levels were increased indicating there was hepatic involvement. A Clinical diagnosis of Henoch Scholein purpura, IgA vaculitis, leucocytoclistic vasculitis was done. During the hospital administration, patient was managed symptomatically for pain control, skin rashes. For supportive care the patient protected from secondary bacterial infection, maintained proper nutrition, fluid and electrolytes balance and corticosteroids were given to reduce the inflammation and thus treating rashes. Proper skin dressing for fast wound healing and Budesonide cream, 5% permite cream was used for lesions topically. For systematic treatment, Methyl prednisolone 1gm IV OD was administered. Later plan for oral steroids was estabiliahed. Vitamin B.complex and Topical Fusidic acid cream was added. Finally the patient was discharged after 20 days and 5% permite cream once overnight application should be given, tablet naproxen 500 mg BD.

DISCUSSION

Henoch-Schonlein purpura (HSP) is the most common form of acute small-vessel vasculitis primarily affecting children. It is recognized as a systemic vasculitis involving skin, gut, kidneys, and joints. HSP is an autoimmune acute leucocytoclastic vasculitis of childhood, was first described in 1837. It is initiated by deposition of immune complexes as responses to infections such as group A streptococci, mycoplasma, Epstein-Barr, and Varicella virus. [10]

HSP mainly affects children between 4 and 15 years and has an incidence of 14 in 100,000 population with peak incidence at 5 years. It usually involves skin, gut, joints, and kidneys, but may rarely have systemic manifestations seen as hepatosplenomegaly and may rarely develop fatal complications like pulmonary hemorrhage and myocardial infarction. [11],[12]. It presents classically with a unique distribution of the rash to the lower extremities and the buttock area. [13] Joints are frequently involved especially knees, ankles, and elbows; but the disease is not known to leave any permanent deformity and there is evidence to suggest that extrarenal manifestations respond well to immunosuppressive therapy. Gastrointestinal (GI) symptoms occur in up to 85% of the patients. Renal involvement is manifested by hematuria and proteinuria. Severe renal and central nervous system disease may lead to life-threatening conditions, and immunosuppressive agents and plasmapheresis may be needed.

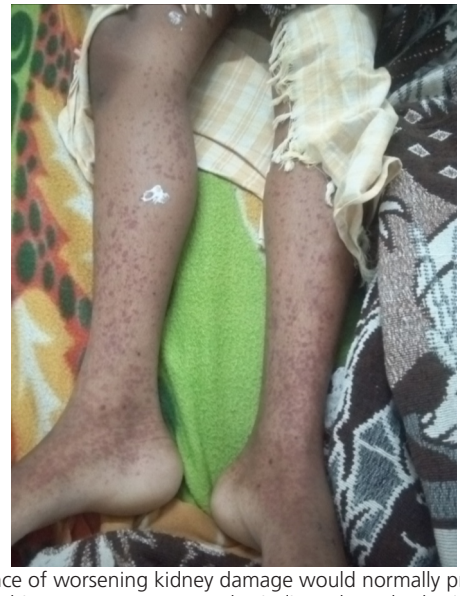
The American College of Rheumatology published diagnostic criteria for HSP in 1990, including age less than or equal to 20 years at disease onset, the presence of palpable purpura, GI bleeding, and a biopsy showing granulocytes in the walls of small arterioles or venules [14].

As it is nown that HSP has a strong involvement with renal system, there may be loss of small amounts of blood and urine. This can may even be preceeded to CKD. A systematic review didn't find any evidence that steroid treatment is effective at decreasing the likelihood of developing long term kidney diseases. Treating renal involvment conditions equally as the other systemic symptoms can prevent long term damage of renal systems, As in this case the primary concern was given only to the cutaneous involvement, pedal odema and increased frequency in urination which are symptoms related to renal system where left untreated. As this condition occurs majorly in children between 4-11, which can leave them effected in there most of there productive age.

As of 2017, the optimal way to treat Henoch Schonlein purpura remains controversial. Analgesics may be needed for the abdominal and joint pains. Wound care is warranted if the skin death and ulcerations occur. It is uncertain as to whether HSP needs treatment beyond controlling the symptoms. Most people do not receive therapy because of the high spontaneous recovery rate. Experts disagree on whether to routinely use corticosteroids as treatment for HSP. However, if they are given early in the disease episode, the duration of symptoms may be shortened, and abdominal pain can improve significantly [15]. Moreover, the chance of severe kidney problems may be reduced.[16]

A systematic review did not find any evidence that steroid treatment (prednisone) is effective at decreasing the likelihood of developing long-term kidney disease

There is no good evidence that treating children who have HSP with antiplatelet agent prevents persistent kidney disease. There is also no evidence that treating children or adults with cyclophosphamide prevents severe kidney disease.Heparin treatmentis not justified[17].



Evidence of worsening kidney damage would normally prompt a kidney biopsy. Treatment may be indicated on the basis of the appearance of the biopsy sample; various treatments may be used, ranging from steroids by mouth to a combination of intravenous methylprednisolone (steroid), cyclophosphamide and dipyridamole followed by prednisone. Other regimens include steroids/ azathioprine, and steroids/cyclophosphamide (with or without heparin and warfarin). Intravenous immunoglobulin (IVIG) is occasionally used [18].

Patients with HSP who have advanced kidney disease and renal failure will probably benefit from mechanical cleansing of the waste products from the blood (hemodialysis). Aggressive and supportive care may be necessary during acute kidney crisis. Some patients with severe kidney disease have undergone kidney transplantation. However, the disease can recur in the transplanted kidney. [19]

CONCLUSION

HSP is an autoimmune acute leucocytoclastic vasculitis of childhood which involves skin, gut, joints, and kidneys commonly. Most affected children have been between 2 and 11 years of age. . It further identifies independent prognostic factors, such as initial renal failure, the level of proteinuria, and the histologic quantification of interstitial fibrosis and glomerular sclerosis. Perioperative management of liver and kidney functions is important. Sufficient Intravenous fluid administration is necessary. Attention should be paid to decrease the risk of tissue compression.

REFERENCES

1) Haycock, G. B. In Oxford Textbook of Clinical Nephrology 1st edn, Ch. 4 (eds Cameron, S. et al.) 595–612 (Oxford University Press, 1992).
2) Stephanie S. Gardner, American Academy of Family Physicians: "Henoch-Schonlein Purpura." MD on January 15, 2018
3) Ballinger S. Henoch-Schönlein Purpura. Curr Opin Rheumatol. 2003;15:591-94.
4) Crowson AN, Mihm MC Jr, Magro CM. Cutaneous vasculitis: a review. J Cutan Pathol. 2003;30:161-73.
5) Ozen S. The spectrum of vasculitis in children. Best Pract Res Clin Rheumatol. 2002;16:411-25.
6) Nadeau SE. Neurologic manifestations of systemic vasculitis. Neurol Clin. 2002;20:123-50.
7) Davin JC, Weening JJ. Henoch-Schonlein Purpura nephritis: an update. Eur J Pediatr. 2001;160:689-95.
8) Rostoker G. Schönlein-henoch purpura in children and adults: diagnosis, pathophysiology and management. Biodrugs. 2001;15:99-138.
9) Rogerwalker, Catewhittlesea. Drug induced skin disorders SJS. Textbook of Clinical pharmacy and therapeutics 4th edition 2007. Churchill Livingstone Elsevier, Pg.no.811.
10) Kraft DM, McKee D, Scott C. Henoch-Schonlein Purpura: A review. Am Fam Physician 1998;58:408-8, 11
11) Vats KR, Vats A, Kim Y, Dassenko D, Sinaiko AR. Henoch Schonlein purpura and pulmonary hemorrhage: A report and literature review. Pediatr Nephrol 1999;13:530-4. [13]Agraharkar M, Gokhale S, Le L, Rajaraman S, Campbell GA. Cardiopulmonary manifestations of Henoch-Schonlein purpura. Am J Kidney Dis 2000;35:319-22. [14]Miller ML, Pachmanin L. Vasculitis M. Syndromes. In: Behrman, editor. Nelson Textbook of Pediatrics, 17 th ed. Philadelphia: W.B. Saunders Company: 2004. p. 167; 826-8.
12) Blanco R, Martinez-Taboada VM, Rodriguez-Valverde V, Garcia-Fuentes M, Gonzalez-Gay MA. Henoch-Schonlein purpura in adulthood and childhood: Two different expressions of the same syndrome. Arthritis Rheum 1997;40:859-64
13) Hetland, LE; Susrud, KS; Lindahl, KH; Bygum, A (November 2017). "Henoch-Schönlein Purpura: A Literature Review". Acta Dermato-venereologica (Review). 97 (10): 1160–66. doi:10.2340/00015555-2733.
14) Weiss PF, Feinstein JA, Luan X, Burnham JM, Feudtner C (2007). "Effects of corticosteroid on Henoch-Schönlein purpura: a systematic review". Pediatrics. 120 (5): 1079–87. doi:10.1542/peds.2007-0667. PMC 3525094.
15) Hahn, Deirdre; Hodson, Elisabeth M.; Willis, Narelle S.; Craig, Jonathan C. (2015-08-07). "Interventions for preventing and treating kidney disease in Henoch-Schönlein Purpura (HSP)". The Cochrane Database of Systematic Reviews (8)
16) Rai A, Nast C, Adler S (1 December 1999). "Henoch-Schönlein purpura nephritis". Journal of the American Society of Nephrology. 10 (12): 2637–44.
17) Saulsbury FT. Henoch-Schonlein purpura. Curr Opin Rheumatol. 2001;13:35-40.