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A CASE REPORT ON HENOCH- SCHONLEIN PURPURA.

Maheshwari V.*, Syam Prakash K. R.[1], Priscilla Mary J.[2], S. Haja Sherief[2] and T. Sivakumar[3]

[1*]Pharm D. Interns, Department of Pharmacy Practice, Nandha College of Pharmacy, Erode, Tamil Nadu, India.

[2*]Department of Pharmacy Practice, Nandha College of Pharmacy Erode, Tamil Nadu, India.

[3*]Principal, Nandha College of Pharmacy, Erode, Tamil Nadu, India.

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***Corresponding Author**
Maheshwari V.
Pharm D. Interns,
Department of Pharmacy Practice, Nandha College of Pharmacy, Erode, Tamil Nadu, India.

ABSTRACT

Henoch-Schonlein Purpura (HSP) is a form of cutaneous small vessel vasculitis that can involve visceral organs and is associated with deposition of immunoglobulin A (IgA)-containing immune complexes. HSP may appear after a remote history of infection (often an upper respiratory tract infection) as a rash with palpable petechiae or purpuraprimarily in the legs and buttocks.[1] Vessel wall inflammation resulting in necrosis and tissue ischemia explain's the clinical presentation of gastrointestinal hemorrhage and glomerulonephritis. A 40 years old male patient presented with B/L lower limb pain, rashes associated with an history of abdominal pain, elbow pain, ankle pain with palpable purpura on and off. These all are the main signs and symptoms of the disease. The case was diagnosed as HSP and was treated with injection Dexamethasone 2cc and T. Wysolone 20mg OD. The patient was also diagnosed with Type 2 diabetes mellitus, this HSP –triggered immunological abnormalities leading to insulitis may have lead to the development of diabetes.[2] Inflammation damages the blood vessels in the skin, intestine and kidney. Early diagnosis and treatment favor the better outcome in cases without any renal complications.

KEYWORDS: Dexamethasone, Glomerulonephritis, Henoch-SchönleinPurpura, Immunoglobulin A, Palpable petechiae, Wysolone.

INTRODUCTION

Henoch-Schonlein Purpura (HSP) is a disorder causing inflammation and bleeding in the small blood vessels, which is self-limited, systemic, nongranulomatous, autoimmune complex, small vessel vasculitis, with multiorgan involvement. Henoch -Schonlein Purpura was first described by William Heberden in 1801. Later scholein described the association between purpura and arthritis, whereas Henoch reported a case with gastrointestinal symptoms along with renal involvement. Henoch-Schonlein Purpura (HSP), also known as IgA vasculitis, is a small vessel vasculitis with IgA1 dominant immune deposits predominantly on capillaries, venules, or arterioles. It often involves skin and gastrointestinal system and may also cause arthritis. The annual incidence of Henoch Schonlein Purpura in children is around 14-20 per 1, 00, 000population.[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.

It's etiology is unclear but is associated with infections (bacterial, viral, parasitic), medications, vaccinations, tumors (non-small cell lung cancer, prostate cancer, and hematologicalmalignancies), alpha-1-antitrypsin deficiency.

PATHOPHYSIOLOGY

Antigen and antibody complexes, mostly IgA, form as a result of bacterial and viral infections, vaccinations, drugs, and autoimmune mechanisms. These antigen antibody complexes deposit in the small vessel walls and activate the alternate complement pathway which leads to neutrophil accumulation resulting in inflammation and vasculitis without a granulomatous reaction. This can involve multiple systems including skin, gastrointestinal tract, kidney, and joints but it can involve any organ system. Vasculitis causes extravasation of blood and its components into the interstitial spaces resulting in edema and hemorrhage.

TREATMENT

The goals of treating HSP are typically to (1) ameliorate acute symptoms, (2) mitigate short-term morbidity (such as abdominal complications that require surgery), and (3) prevent chronic renal insufficiency. Because HSP is characterized by leukocyte infiltration of the blood vessel walls along with immunoglobulin A deposition (with resulting vascular injury and necrosis)and corticosteroids inhibit inflammatory processes, early treatment with corticosteroids has been postulated to be effective for all 3 therapeutic goals.[4] The prognosis

is good, with exception of renal involvement that may need the follow-up till six months or longer.

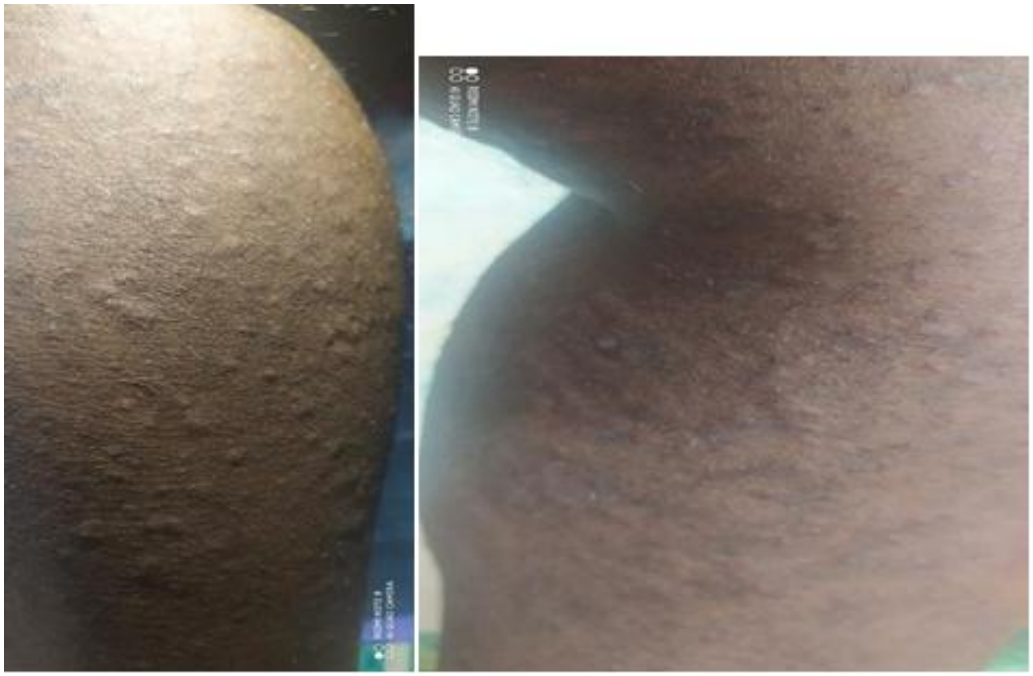
CASE REPORT

A 40-year-old male patient presented with the complaints of B/L lower limb pain and rashes, swelling for past 16 days. History of abdominal pain, brownish stools, vomiting associated with bilateral lower limb burry sensation with new onset lesions. Then he also developed palpable purpura on and off followed by development of bilateral ankle pain. On examination, the general condition of the patient was fair and vitals were stable. The abdomen was soft and mild ascites were noticed or observed. On USG examination, hepatomegaly with mild ascites was confirmed and the patient was newly diagnosed with Type II DM.

Laboratory tests showed that blood urea level 12mg/dL(10-50 mg/dl) slightly decreased, fastingblood glucose level got increased 225mg/dL as well as PPBS also increased 355mg/dL, HbA1c is 9.5% which indicate patient is diabetic. Totalred blood cell count is 4.89 million/mm^3. ESR level is increased to 54mm/hr. C-Reactive protein shows positive (13.63mg/dL).[5,6] On his urine analysis, proteinuria was observed, which is one of the confirmation test for of Henoch- Schonlein Purpura.[7] On biochemistry report, SPOT protein level is increased 54.90mg/dLserum creatinine level is also increased 132.mg/dL. Protein creatinine ratio (PCR) in urine is 0.41mg/dL. Through this confirmatory tests for Henoch-Schonlein Purpura was confirmed.

He was treated with Inj.dexamethasone 2cc in OP department and T. Wysolone20mg OD . For diabetes, T. metformin 500mg TDS, T. Glimepiride 1mg BD. T.Calcium ascorbate and Zinc gluconate 250/2mg BD. T. CPM 4mg OD. T. DUCAINE 400mg.

The recovery from purpura and bilateral swelling of the legs was observed after treatment in the second and third week.



DISCUSSION

Henoch-Schonlein Purpura (HSP) was first described by William Heberden in 1801. Later, Schonlein recognized the association between purpura and arthritis, whereas Henoch reported a case that also included gastrointestinal symptoms along with the renal involvement.[9]

The patient generally presents with the complaints of B/L lower limb pain and rashes, abdominal pain, brownish stools and vomiting and associated with bilateral lower limb burry sensation with new onset of lesion. Then he also developed Palpable purpura on and off followed by the development of bilateral ankle pain. These all are the signs and symptoms of Henoch- Schonlein Purpura. The joint involvement is generally characterized by the pain and swelling of the joints, most affecting the knees and ankles. The abdominal pain followed by vomiting and intestinal bleeding is the dominant features involving the gastrointestinal system. Microscopic hematuria and albuminuria are the prominent renal findings.

Our case have the symptoms of lower limb pain and rashes, abdominal pain. Laboratory finding shows elevated ESR Level, elevated creatinine level, presence of proteinuria and presence of brownish stools. Here this patient was diagnosed newly with diabetes which may be due to HSP, this HSP –triggered immunological abnormalities leading to insulitis may lead to the development of diabetes.[2] (hence his glucose level was under monitoring). Hematuria (microscopic or gross) is the most common renal manifestation. Proteinuria is either seen along with hematuria. This patient was having protenuria but there is no

hematuria. Persistent proteinuria and hematuria predicts the development of ESRD (End Stage Renal Disease). Although most of the patients develop renal involvement within 3 months of skin manifestations, they should be followed for a year with urine analysis. Renal involvement is the most important prognostic factor in determining morbidity and mortality from HSP.

In 94 percent of children and 89 percent of adults, HSP care illustrates its self limiting nature. Symptomatic care for symptoms such as rash, and arthritis should be appropriate. Acetaminophen and anti inflammatory drugs which are non steroidal may be used. Typically prednisone or methyl prednisolone should be started for one to two weeks at 10-20 mg/kg per day, tapering to o.5 mg/kg/day for the next week, and then 0.5mg/kg every other day for another week. If the patient tolerates oral steroids, intravenous (iv) steroids can be administered. Early steroid therapy decreases gastrointestinal symptoms in patients within 2 days compared with 12 days without steroids, and can decrease HSP or gastrointestinal recurrence and decrease progression. Steroids may avoid in significant complications including gastrointestinal compressions, bleeding or intussusceptions suggested high dose iv plus steroids in nephritic- proteinuria patients and mesenteric vasculitis. Immunosuppressive drugs (cyclophosphamide, azathioprine, cyclosporine A, and mycophenolate mofetil) in combination with high-dose IV plus steroids are recommended if there is no benefit from steroids alone. This is usually recommended in rapidly progressive glomerulonephritis (RPGN) and hemorrhagic involvement of the lungs and brain. Steroids are more often used for the relief of abdominal pain, joint pain, and skin disease. Alternatively, methotrexate and dapsone have been quite effective (steroid sparing agent) for the treatment of chronic abdominal pain and skininvolvement.[6] The role of the corticosteroids in preventing the long-term outcome of renal complications is controversial. In general, prednisolone is the commonly used steroid for the treatment of HSP. In our case, T.Wysolone 20mg OD was prescribed. Several cases and studies have been reported resulting in better outcomes on treatment with dexamethasone.

CONCLUSION

HSP being one of the most common vasculitis of the children and its classic presentation of palpable purpura, arthritis, abdominal involvement, and renal features makes the diagnosis quite easier. Early initiation of treatment with steroids will help in symptomatic relief to achieve positive outcomes. The renal disease may need long-term follow-up; otherwise, the

diseases have favorable prognosis. Corticosteroids, given early in the course of illness, seems to produce consistent benefits. Early corticosteroid treatment significantly reduced the odds of developing persistent renal disease benefits for several major clinically relevant Henoch-Schonlein purpura outcomes.

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