Case Report

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Case Report on Henoch-Schonlein Purpura Vasculitis

Madhu M[1], Robin George[*2], Manaswi N[1], Ashik P T[1], Thasvin U R[1]

[1]Department of pharmacy practice, Sri Adichunchanagiri hospital and research centre, Mandya, India
[2]Department of Pharmacy Practice, Sri Adichunchanagiri College of Pharmacy, Mandya, Karnataka, India

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Abstract

Henoch-Schonlein purpura is a type III hypersensitivity leukocytoclastic small vessel vasculitis caused by the IgA-immune complex that primarily affects children and has the potential to escalate to many complications. A female patient of age 17 years came with the complaint of asymptomatic red rashes over their left lower leg since 3 weeks back. It soon involved the other leg within 3-4 days. Also Associated ankle pain. on clinical examination Pulse Rate-86/min, BP-120/80 mm/Hg, RR-20/minute, non-pitting edema and palpable purpura of bilateral lower legs and few over the thighs. skin biopsy was done, also received fresh tissue for IF, which shows 1-granular IgA along upper dermal vessels, she was given Tab. Augmentin 625mg, Tab. Colchicine 500 mcg, Mupimet ointment and other supportive measures for two days. When the patient's condition improved, discharged with medication Tab. Augmentin 625mg, Tab. pantoprazole 40mg, Mupimet ointment each for 4 days and Tab. colchicine 500mcg for 7 days with advice to review in dermatology OPD and rheumatology OPD with prior appointment after one week. Continued collaboration and knowledge-sharing within the medical community will ultimately improve patient outcomes and contribute to the advancement of care for individuals affected by this rare vasculitis disorder.

*Corresponding Author

Name: Robin George
Phone:
Email: robin.george793@gmail.com

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INTRODUCTION

Henoch-Schonlein purpura is a type III hypersensitivity leukocytoclastic small vessel vasculitis caused by the IgA-immune complex that primarily affects children and has the potential to escalate to chronic renal impairment. The rash typically affects the buttocks and lower extremities and starts as symmetric erythematous macules that progress into purpuric papules. The skin, kidneys, gastrointestinal tract, and joints are the most often impacted organs, while the involvement of the lungs, genitourinary tract, and central nervous system is uncommon [1, 2]. According to the most recent classification criteria, there must be palpable purpura along with at least one of the other clinical or pathological characteristics listed below: Symptoms of renal involvement, such as hematuria or proteinuria, include diffuse abdominal discomfort, arthritis or arthralgia, a biopsy indicating IgA predominate deposition [2].

According to estimates, the disease affects between 3.4 and 14.3 cases per million adults and 15 cases per 100,000 children per year [3]. HSP can be divided into two major categories: HSP without renal involvement and HSP nephritis [2]. Recently, vasculitides have been related to the severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) and its offspring coronavirus disease 2019 (COVID-19). The lab reports to detect HSP vasculitis are Antistreptolysin-O titers, Basic metabolic panel (e.g,

electrolytes, blood urea nitrogen, creatinine), Blood culture, Coagulation profile (PT/PTT), Complete blood count, IgA levels, Skin or renal biopsy, Urinalysis, hCG, amylase, liver function tests, abdominal CT scan, Skin biopsy, ESR, echocardiography, bone marrow biopsy, CSF analysis and culture, creatinine, urinalysis etc [4].

Analgesics or non-steroidal anti-inflammatory drugs are used as first-line treatment for the relief of arthralgia and antihistamines for the treatment of purpuric rash. Plasma exchange and immunosuppressive medications including cyclophosphamide, azathioprine, and cyclosporine have also been mentioned as treatments for refractory GI problems in HSP patients [5]. Dapsone, at a dose of 1-2 mg/kg once a day for a week, can be beneficial for chronic IgA vasculitis when cutaneous signs last longer than 6 weeks [6]. If the patient is unable to tolerate oral steroids, intravenous (IV) steroids can be delivered. Typically, prednisone or methylprednisolone can be begun at 1 to 2 mg/kg per day for one to two weeks, tapering down to 0.5 mg/kg/day over the following week, and then to 0.5 mg/kg every other day for one more week. Early steroid therapy reduces gastrointestinal symptoms in patients within 2 days as opposed to 12.3 days when they do not receive steroids, and it may also slow the progression of renal disease or HSP [7, 8]. While IVIG (intravenous immunoglobulin) provides an alternate therapy for steroid-resistant illness, corticosteroids have remained the conventional therapy for individuals with moderate to severe GI involvement [9].

There are serious side effects, including myocarditis, mumps, and involvement of the brain system (such as encephalopathy and pulmonary bleeding) as well as the respiratory and urinary systems (such as scrotal and penile involvement) [10]. So, here we present a 17 years female patient diagnosed with HSP vasculitis with the complaint of red rashes on lower limbs.

Case Study

A female patient of age 17 years came with the complaint of developed asymptomatic red rashes starting over the left lower leg since 3 weeks back. It soon involved the other leg within 3-4 days. There's also some sort of pain in the ankle. Two weeks ago, patient was having a history of fever. The patient was given one dose of Amoxicillin and topicals for 1 day, after consulting a doctor in the area. History of atopy present and there is No history of abdominal pain, urinary symptoms, recurrent mouth ulcers, photosensitivity, no new drug intake in the past 3 months. A history of similar lesions in her brother is present. On clinical examination Pulse Rate-86/minute, BP-120/80 mm of Hg, Respiratory rate-20/minute, non-pitting edema of bilateral lower legs present, Multiple palpable purpura over bilateral lower legs and few over the thighs. Single skin punch 0.3 x 0.2 x 0.2 cm hairline Received in formalin, as a specimen labeled "Left leg LM skin biopsy". Entire specimens are deposited in Cassette I. Fresh tissue is also received for IF. Microscopically Sections of the skin-epidermis. IF shows granular IgA along the upper skin vessels, the external appearance of the patient's lower extremity skin is shown in Figure 1. She started oral antibiotics TAB. AUGMENTIN 625mg TDS, Tab. Colchicine 500mcg BD with Mupimet ointment for L/A biopsy site and other supportive measures and continued for two days.

An ophthalmology consultation was referred to the patient's complaints of blurred vision. There were no signs of vasculitis or an infection in her eye and she started to tear drops with regular follow-up advice for six months. The patient improved clinically during this period; Laboratory parameters were normal. Therefore, the patient was discharged in hemodynamically stable condition. For hospital discharge, Augmentin Tab. 625mg p/o 1-1-1 x 4 days (after meals), pantoprazole Tab. 40mg p/o 1-0-1 x 4 days (before meals), Mupimet topical ointment L/A biopsy site 1 -0-1 x 4 days, colchicine Tab. 500mcg p/o 1-0-1 x 7 days with advice to be considered at OPD dermatology and OPD rheumatology with prior appointment after one week.

DISCUSSION

HSP is an acute systemic small vessel inflammation characterized by classical palpable purpura, arthritis, gastrointestinal and renal manifestations [11]. It is the most common vasculitis in children and usually occurs in children aged 3 to 10 years; however, it is known to affect young people as well. Many patients have a history of severe allergies and believe that the exact cause is unknown, but in our case it was due to family history.

patient Immune complex deposition on the skin causes palpable purpura and petechiae. This happens when blood vessels become inflamed, which can bleed into the skin, causing a red-purple rash (purpura). The most prominent feature of this form of vasculitis is a purplish red rash, often on the lower legs and buttocks. Henoch-Schonlein purpura can also cause abdominal pain and joint pain [12].

The pathophysiology of HSP is not fully understood; however, IgA plays an important role. IgA-antibody immune complexes result from antigen exposure due to infection or drug deposition in small ves-

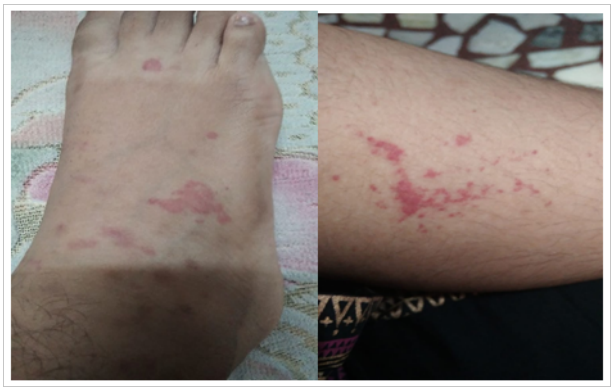

Figure 1: Red rashes on the lower limbs

sels of the skin, joints, kidneys, and gastrointestinal tract. Additional C3 receptor lymphocytes can bind to immune complexes and are located in the vessel wall contributing to the hyperinflammatory response [13]. In this case, the patient had gastrointestinal complications.

Although HSP is a clinical diagnosis, laboratory and imaging studies may be helpful in more atypical cases. In addition to routine laboratory studies, an extensive immunohistochemical panel may be required to assist in the diagnosis and rule out alternative pathophysiology, including levels of ANA, ANCA, RF, and factor VIII and XIII. Imaging may also be considered, such as a kidney or skin biopsy, which may play a role when the diagnosis is uncertain or to monitor for possible complications and systemic involvement [14]. In this case, all laboratory values were normal, but the skin biopsy showed HSP vasculitis. When viewed microscopically, the dermal-epidermal sections show reticulocysts with preserved granules and basket-shaped keratin. The dermis is hyalinized collagen that thickens and has features of small vessel inflammation with endothelial swelling, and extravasation of erythrocytes. leukocytoclastic, fibrinoid necrosis, and perivascular infiltrates of eosinophils, neutrophils, lymphocytes, and histocytes. This is also seen around the small circuits around the fatty S/c appendages that appear to be common. PAS shows inflammation, MF shows melanosis along the basal layer, Orcein shows elastic fibers in the dermis Tol blue shows interstitial mast cells. IF shows granular 1-IgA along blood vessels in the skin. Management of HSP is primarily supportive and involves a combination of analgesics, antiemetics, rehydration, and monitoring for complications. Treatment is aimed at relieving acute symptoms and preventing kidney failure. Skin lesions usually do not require treatment. Since HSP is characterized by IgA deposition and leukemic cell infiltration in the vascular wall, corticosteroids may play a role in suppressing this inflammatory process [15]. One study found that 1 mg to 2 mg per kg of prednisolone taken by mouth for two weeks was effective for abdominal and joint symptoms. Other studies investigating the role of corticosteroids in HSP have shown that although steroids do not prevent the onset of kidney damage, they help reduce symptoms (especially abdominal pain and joint pain).as was the case in our patient whose symptoms disappeared after steroid administration [16]. Since the patient is in the initial phase, there is no increase in laboratory values, corticosteroids are not prescribed. And re-examined IgA to prescribe steroids but the value was normal, so the patient continued to take Tab. Augmentin 625mg, Tab. Colchicin 500mcg and Cupimet ointment for the biopsy site.

CONCLUSION

In conclusion, this case report sheds light on the manifestation, diagnosis, and management of Henoch-Schönlein purpura vasculitis (HSP), which reports that the cases are also related to the outcome. testing increased. Management requires an individual approach to address the specific presentation and severity of the disease. In this case, a combination of supportive care, antibiotics, and topical medication is effective in controlling the patient's symptoms and preventing further complications. This report highlights the importance of early recognition and prompt intervention in HSP vasculitis to minimize the risk of long-term sequelae. As with any case report, the results presented here contribute to existing knowledge, providing valuable information that can aid in the diagnosis and management of future cases. future. However, further research and larger studies are needed to better understand the underlying pathophysiology, risk factors, and optimal treatment strategies for HSP vasculitis. Continued collaboration and knowledge sharing within the medical community will ultimately improve patient

outcomes and help improve care for those affected by this rare vasculitis disorder.

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Conflict of Interest

The authors declare there is no conflict of interest.

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