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Case Study

Case Report on Henoch-Schonlein Purpura: A Vasculitic Disorder

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ABSTRACT

Henoch-Schonlein Purpura (HSP) is a rare vasculitic disorder causing small vessels of the skin, alimentary canal, kidney, joints, and rarely lungs and central nervous system (CNS). HSP occurs decreasing with age. Here we report a case of a male child who presented with mild grade fever, leg pain, purpuric rashes, and non-pitting pedal edema. The head to foot examination shows restriction of movements presented in the lower limbs and palpable purpura presented in both upper and lower limbs associated with edema. Symptomatic therapy is enough for the treatment. Although he was successfully treated, mystery still veiled the etiology, pathophysiology, also line of management of this rare and enigmatic disease.

Key words: Henoch-Schonlein Purpura, Vasculitic disorder, Purpuric Rashes, Pedal edema

INTRODUCTION

Henoch-Schonlein purpura (HSP), a small vessel vasculitis, is the most common in children, with an incidence of around 10 cases per 100,000 a year. It is a self-limited, systemic, nongranulomatous, autoimmune disease with multiple organ involvement. The etiology is unknown, although it associated with infections (bacterial, viral, parasitic), medications, vaccination, tumors (non-small cell lung cancer, prostate cancer, and hematological malignancies), alpha-1antitrypsin deficiency and familial Mediterranean fever. It is the most common cutaneous vasculitis up to 90% of cases in children. Few laboratory tests are useful for diagnostics. Mild leukocytosis, eosinophilia are also associated with a platelet count[1,4].

The diagnosis in children based on cutaneous rashes that may cause urticaria or purpura, affecting primarily, the extender surfaces such as buttocks, and legs. Arthritis is a feature that often affects the larger joints, particularly the knees and ankles, and it is debilitating. Gastrointestinal involvement, which may

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include colicky abdominal pain, vomiting, gastrointestinal bleeding, pancreatitis, and gallbladder hydrops, occurs in 50-70%, but more often with the renal disease such as HSP nephritis. Nevertheless, steroids used to decrease GI symptoms such as chronic GI pain, as well as to decrease renal complications[5,6].

Generally, prednisolone or methylprednisolone started for one to two weeks at 1 to 2mg/kg daily tapering down to 0.5mg/kg/day for the next week, and then 0.5mg/kg another day of next week. When the patient does not tolerate oral steroids intravenous (IV) steroids can be administered. Immuno-suppressive medications such as (cyclophosphamide, azathioprine, cyclosporine A, and mycophenolate mofetil) prescribed and associated with high-dose IV steroids if steroids give their own activity. It is generally prescribed for rapid progressive glomerulonephritis (RPGN) and hemorrhagic involvement for lung and brain. Nonsteroidal anti-inflammatory drugs (NSAIDs), i.e. flurbiprofen (5 mg/kg daily) or naproxen (15 mg/kg daily), to reduce severe joint pain[2,7].

CASE REPORT

A 5-year-old male child admitted to the pediatrics department of the hospital presented with mild grade fever for 1 week, it was acute in onset. He was given oral antibiotics by his primary care doctor. Leg pain has developed for the last 2 days and associated with restricted movements. Pain has increased in joints (knee and ankle joints). Later the child developed purpuric rashes involving upper-lower extremities, buttocks, and non-pitting pedal edema (Figure1&2) is also present. The patient also had a history of intermittent abdominal pain. Laboratory tests showed Hb:10.7g/dl, WBC:8210cells/cumm, DLC-P:37.7%; L:49.0%; E:8.7%; M:4.2%, platelet:3.44lakh/cumm, MCV:70.7fl, PCV:29.1%, ESR : 12mm/hr, MCH : 26.0pg, MCHC : 36.8g/dl, on urinalysis reddish discoloration of urine due to calcium oxalate crystals, and no proteinuria present. From the systemic examination, CNS shows that the tone is normal in both upper and lower limbs. Occult blood stool is negative. Head to foot examination revealed that there was a restriction of movement in the lower limb and palpable purpura on the skin. Edema presented in the dorsal side of upper limbs and arthralgia is also present, the patient diagnosed with Henoch-Schonlein Purpura (HSP) as per the patient's symptoms and systemic examinations and treated with intravenous fluids, oral prednisolone 4ml three times a day, oral food to resolve his symptoms. His symptoms relieved and the patient was better on the fifth day of the hospital.

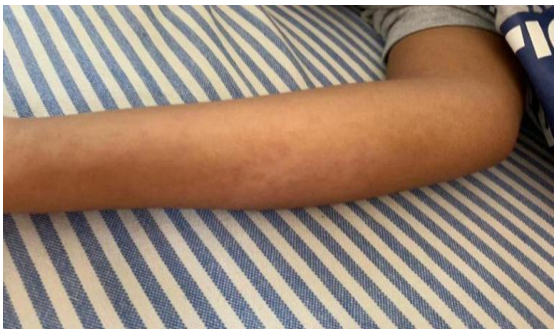

Figure 1: Clinical picture of palpable purpura involvement of upper extremity.

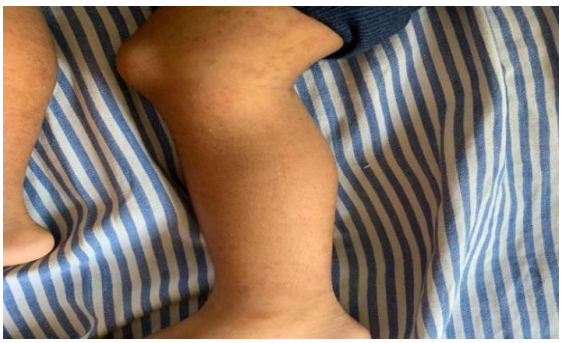

Figure 2: Clinical picture of palpable purpura involvement of lower extremity.

DISCUSSION

Henoch-Schonlein Purpura is usually seen in children between 3 and 10 years of age.

Males are twice as much affected than females. While it can occur at any age, HSP is predominantly a childhood disease. Prominent features of HSP include the epidermal part with skin lesions consisting of sub-epidermal haemorrhages and necrotizing vasculitis of small vessels of the dermis. IgA is present in these vessels as well. Vasculitis may also occur in multipleorgans, such as the gastrointestinal tract, kidney, rarely lungs, and CNS. HSP tends to have atypical clinical features[1,4].

Clinical symptoms of HSP include GI pain, nonthrombocytopenic purpura, arthritis, and nephritis. In infants below the age of 2 years, the severity tends to milder and worse in adults. Most patients are presented the disease condition from autumn to spring seasons and respiratory infection is often followed by HSP also been associated with a variety of pathogens, medicines, and other environmental exposures. Nephritis is also a characteristic feature of HSP that can have severe effects and the long-term prognosis depends on the severity of nephritis. The second most common clinical feature is arthritis/arthralgia[3,4,6].

IgG ANCA helps to diagnose these conditions. Cytoplasmic ANCA (c-ANCA) is most commonly associated with Wegener's granulomatosis and perinuclear ANCA (p-ANCA) with microscopic polyarteritis. Serology is again a distinguishing feature in systemic lupus erythematosus, which associated with similar vasculitic rashes. Other causes of purpura may include sepsis, coagulation disorders, or thrombocytopenia that can easily identify by hematological investigations[5].

Although the cause is unclear, it is clear that IgA plays a pivotal role in HSP pathogenesis. There are two IgA subclasses, IgA1 and IgA2 but HSP involves only IgA1. It is a self- limited disease that lasts an average of 4 weeks, in most severe cases. If the patients with recurring symptoms, after 4–6 months recurrences usually subside. HSP treatment is based on clinical guidelines. The updated criteria developed by EULAR/ PRINTO/ PRES published a revised set of criteria in 2010, and are the gold standard for the diagnosis of HSP (Table 1). When applied to infants, the sensitivity is100% and specificity is 87%. There are no specific bio-markers useful for diagnosing HSP. Some bio-markers show disease activity and prognosis but none have proved clinically useful. Skin biopsies are the gold standard treatment of skin vasculitis. IgA-predominant vascular deposits are characteristic of HSP but not relevant for the diagnosis of HSP as these deposits can be identified in other vasculitic syndromes, erythema nodosum, and venous stasis-related conditions. Symptomatic treatment for symptoms like rashes and arthritis include acetaminophen and anti-inflammatory non-steroidal agents[4,8].

In the present study, the patient diagnosed with Henoch-Schonlein Purpura on the basis of clinical symptoms, systemic examinations, and using certain criteria such as European League Against Rheumatism (EULAR), the Paediatric Rheumatology International Trials Organization (PRINTO) and the Paediatric Rheumatology European Society (PRES) published in 2010, with high sensitivity and specificity. The severity of the disease and organ involvement indicates the treatment. Usually, the treatment for HSP is symptomatic without renal involvement [8].

In this case study, the patient treated with IV fluids, NSAIDs, antiemetics, and prednisolone4mlthrice a day from the second day of the hospital for the condition. The following recommended dose and route of administration continued until the fifth day of the hospital.

Table 1: Diagnostic criterion for Henoch-Scholein Purpura (HSP), as developed by EULAR/PRINTO/PRES

Criterion	Description
Mandatory Criterion Predominance	Purpura or petechiae with lower limb.
Minimum 1 out of 4 Criterion	1. Diffuse abdominal pain with acute onset. 2. Histopathology showing leukocytoclastic vasculitic or proliferative glomerular nephritis, with predominant Immunoglobulin A (IgA) deposits 3. Arthritis or arthralgia of acute onset 4. Renal involvement in the form of proteinuria or hematuria

EULAR/PRINTO/PRES: The European League against Rheumatism/ the Pediatric Rheumatology International Trails Organization/ the Pediatric Rheumatology European Society[8].

CONCLUSION

Henoch-Schonlein Purpura is the most common vasculitic disorder in children[1]. The patient diagnosed according to symptoms, systemic examinations, and EULAR/ PRINTO/ PRES criterion. Most cases rectify with symptomatic treatment, but serious complications may occur for example renal failure. The patient in this study started recovering the symptoms (leg pain, purpuric rashes, non-pitting edema) while switching the steroid therapy. All symptoms revealed on the last day of the hospital.

Conflict of Interest: The authors have declared no conflict of interest.

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REFERENCES

1. Roberts P Fetal. Henonch-ScholeinPurpura: A review article. South Med J.2007; 100: 821-24.
2. Sohagia AB et al. Henoch-Schonlein Purpura-A Case Report and Review of the Literature. Gastroent Res Pract. 2010; 2010: 1-7.
3. Peru Hetal. Henoch-SchonleinPurpura in childhood: clinical of 254 cases over a 3-year period. Clin Rheumatol. 2008; 27: 1087-92.
4. Saulsbury FT. Clinical update: Henoch-SchonleinPurpura. Lancet. 2007; 369(9566):976-8.
5. Tizard EJ. Henoch-SchonleinPurpura. Curr. Pediatr. 2002; 12: 575-80.
6. Tarvin SE, Ballinger S. Henonch-ScholeinPurpura. Curr. Paediatr. 2006; 162: 259-63.
7. Trapani S et al. Henoch-SchonleinPurpurainchildhood: Epidemiological and Clinical Analysis of 150 Cases Over a 5-year Period and Review of Literature. Semin Arthritis Rheu. 2005; 35: 143-53.
8. Hetland LE et al. Henoch-Schonlein Purpura: A Literature Review. Acta Derm Venereol.2017;97:1160-66.