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Case Report
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HENOCH-SCHÖNLEIN PURPURA: A CASE REPORT OF AN EIGHT-YEARS-OLD BOY

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Abstract
Henoch-Schönlein Purpura (HSP) is one of the most common causes of small vessel vasculitis mediated by immunoglobulin (Ig) A deposition in children. We present a case of HSP in an eight years old boy with cutaneous, abdomen and joint manifestation of the disease. He had history of upper respiratory tract infection couple weeks before. Physical examination showed erythematous, palpable, purpuric rashes on his both legs and buttocks. Mild bilateral non pitting edema was present over both the legs. Complete blood count (CBC) test revealed thrombocytosis and slightly leukocytosis with neutrophil predominate. The case was treated with oral methylprednisolone for seven days and showed a good outcome.
Keywords: Children, Henoch- Schönlein purpura, HSP

INTRODUCTION

Henoch-Schönlein Purpura (HSP) is a systemic small vessel vasculitis with the deposition of immune complexes containing immunoglobulin (Ig) A that occurs commonly in children with a peak age between four and eight years and rarely presents in adults. It is characterized by the involvement of the skin (non-thrombocytopenic palpable purpura that mostly located on the dependent parts like lower extremities and buttocks), synovia (arthralgia/ arthritis), gastrointestinal tract (bowel angina) and kidneys (haematuria/proteinuria). HSP is regarded as a specific immune-mediated entity induced by environmental factors, particularly a history of preceding infections (especially upper respiratory tract infection with β-hemolytic *Streptococcus*, in up to 75% of individuals) and certain drugs (e.g., penicillin, quinines, erythromycin). HSP is currently diagnosed based on symptoms and signs and histopathological findings since there are no disease-specific laboratory abnormalities. Because the disease course is usually benign and self-limited, treatment is supportive.[1,2,3,4]

Case Report

An eight years old boy presented with the complaints of rashes over both of the legs associated with pain of joints in upper and lower limbs for seven days. He developed swelling in both legs subsequently few days after appearance of rashes starting from feet and progressing to buttocks. He also complained of pain in the abdomen was localized around the umbilicus followed by worsening pain of all of the body's joint. He had history of upper respiratory tract infection couple weeks before.

On physical examination, the general condition of the patient was good and vitals were stable. The abdomen was soft and non-tender. Mild bilateral non pitting edema was present over both the legs. There was presence of non-tender, non-blanching, palpable purpuric rashes over both legs and buttocks.

Complete blood count (CBC) showed thrombocytosis and slightly leukocytoses with neutrophil predominate. Serum urea and creatinine were within normal limits. Urinalysis was negative for protein and blood. There was no blood detected on stool analysis.

The patient was diagnosed with HSP according to The American College of Rheumatology (ACR) 1990 criteria and European League against Rheumatism/ Paediatric Rheumatology International Trials Organization/Paediatric Rheumatology European Society (EULAR/PRINTO/PRES) 2010 criteria. Patient was treated with intravenous fluids drip, oral methylprednisolone 12 mg/day divided in three doses for seven days and other supportive management. Patient experienced significant improvement and then he was discharged after seven days of hospitalization.

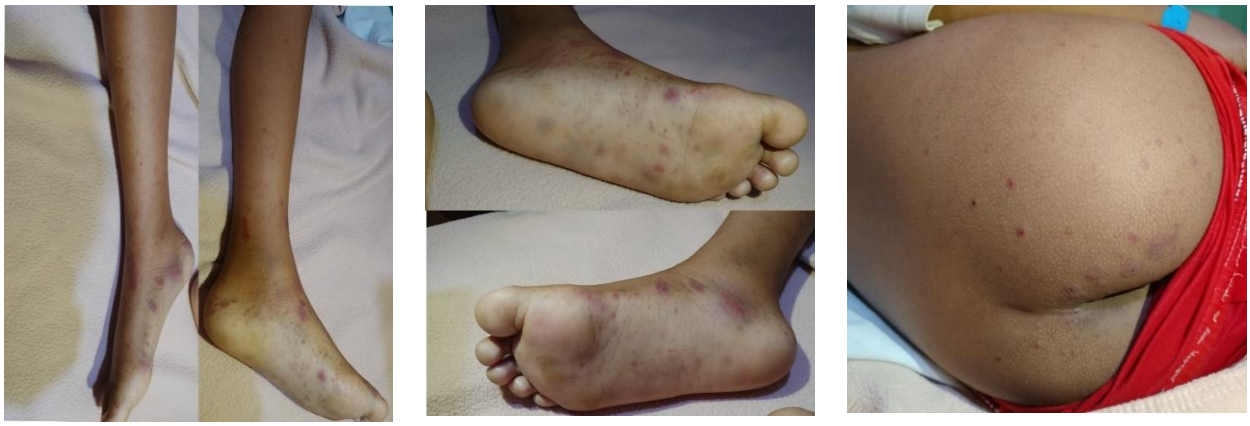

Figure 1: Palpable Purpura on Patient's Legs and Buttocks

Discussion

William Heberden first described HSP in 1801. Later, Schönlein described HSP as triad of purpuric rash, arthritis, and abnormalities of the urinary sediment in 1837. Whereas in 1874, Henoch described the association of purpuric rash, abdominal pain with bloody diarrhoea and proteinuria.[4,5] HSP occurs primarily in male children, with a peak age between four and eight years, however, adults may also be affected.[3] It has been proposed that various triggers such as a viral infection or streptococcal pharyngitis, vaccinations, drugs, and autoimmune mechanisms may result in the formation of an antigen and antibody complex and the deposition of such formed immune complex in the small vessels may activate the alternate complement pathway leading to neutrophil aggregation which results in inflammation and vasculitis. This process that including extravasation of erythrocytes, infiltration of tissues with neutrophils, and deposition of degenerating neutrophil fragments are well known as leukocytoclastic vasculitis (LCV). [3,6]

The clinical evaluation is critical in separating those cases of benign cutaneous vasculitis (usually following an infection or induced by a medication) from those cases associated with more serious underlying disease or which have significant systemic involvement.[3]

HSP is characterized by purpura, arthralgias, abdominal and renal disease. The extravasation of blood leads to the pathognomonic purpura of the skin, while inflammation and edema leads to swelling which is usually palpable. Palpable purpura in the absence of thrombocytopenia is typically suggestive of HSP as it found in almost all patients. Typically, mottled purpura appears on the extensor aspects of the extremities and areas under pressure such as the buttocks, which become haemorrhagic within a day and start to fade in about five days. New lesions may appear over a period of a few weeks. Urticarial lesions, vesicles, necrotic purpura, and haemangioma like lesions may also be present at some stages. Other symptoms of HSP include arthralgias, abdominal and renal disease manifestation, though not universally present. In about 40% of cases, the cutaneous manifestations are preceded by mild fever, headache, joint symptoms, and abdominal pain for up to two weeks. Arthralgia progressing to arthritis produces periarticular swelling around the knees and ankles. Abdominal pain and gastrointestinal bleeding may occur at any time during the disease. Severe abdominal pain may even suggest an acute surgical abdomen. Paralytic ileus may occur. Vomiting, rebound tenderness, and distension are other manifestations. Gastrointestinal radiographs may show spiking or a marbled cobble-stone appearance. Renal involvement manifests as microscopic or even gross haematuria and may occur in 25% or more of patients. Progressive glomerular disease and renal failure may develop in a small percentage, so that careful follow-up is necessary for those with hematuria.[3,7]

Screening laboratory tests may help to elucidate the underlying cause or extent of organ involvement. When the history suggests a recent drug and the patient is clinically well, nothing more than a urinalysis may be required. CBC, urinalysis, throat culture, hepatitis B and C serologies, antinuclear antibody (ANA) and rheumatoid factor (RF) are a reasonable initial screen for patients with no obvious cause for their vasculitis. The CBC test shows a mild leukocytosis with neutrophil predominate. Urinalysis must be performed in order to evaluate hematuria

and proteinuria. Stools should be examined to evaluate visible or occult bloods. Elevated serum IgA levels have been associated with HSP. Inflammatory markers such as erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels are often elevated. Serum protein electrophoresis, serum complements, anti-neutrophil cytoplasmic antibodies (ANCAs), and cryoglobulins may be required in some cases. In patient with severe abdominal pain, an ultrasound examination is helpful to delineate whether an intussusception is present. Endoscopy and/or colonoscopy play a major role in helping diagnosis of the patients with the gastrointestinal involvement as their initial presentation[3,7]

A skin biopsy should be obtained from the lesion less than 24 hours to confirm the diagnosis of LCV. The most frequent pathology observed is a mesangial or endocapillary proliferative glomerulonephritis. IgA, complement component C3, and fibrin depositions have been demonstrated in biopsies of both involved and uninvolved skin by immunofluorescence techniques. Since IgA immune complexes persist longer in vasculitis, they are easier to identify than the IgG/IgM containing immune complexes in other forms of LCV. Renal biopsy should be performed in case of uncertain diagnosis or severe renal impairment such as nephrotic syndrome. The age of onset, presence of renal impairment and hematuria at the onset, abdominal pain as an initial presentation, persistent eruption, renal pathology with fibrinoid necrosis and the number of sclerotic glomeruli is significant predictors of renal disease.[3,7]

In this case, an eight years old boy had the symptoms of palpable purpuric rash (over both lower legs and buttocks), swelling in both legs, arthralgia which indicates joint involvement and abdominal pain. However, there was no sign of renal involvement. CBC test revealed thrombocytosis and neutrophil predominate.

Diagnosis of this case was made by using ACR 1990 criteria and EULAR/PRINTO/PRES 2010 criteria. ACR classification consists of four criteria and shall be diagnosed with HSP if at least two of these criteria are present. The presence of any two or more criteria yielded a sensitivity of 87.1% and specificity of 87.7%. From EULAR/PRINTO/PRES classification, patient shall be diagnosed with HSP if there are a present of nonthrombocytopenic palpable purpura (mandatory criteria) together with at least one of additional criteria. The sensitivity and specificity of these classification criteria were 100% and 87% respectively.[4]

Table 1: The ACR 1990 Criteria for HSP Diagnosis[4]

Criteria	Definition
Palpable purpura	Slightly raised palpable haemorrhagic skin lesions, not related to thrombocytopenia
Age ≤ 20 years at disease onset	Patient 20 years or younger at onset of first symptoms
Bowel angina	Diffuse abdominal pain, worse after meals, or the diagnosis of bowel ischemia, usually including bloody diarrhoea
Wall granulocytes on biopsy	Histologic changes showing granulocytes in the walls of arterioles or venules

Table 2: EULAR/PRINTO/PRES 2010 Criteria for HSP Diagnosis[4]

Criteria	Definition
Mandatory criteria	Palpable purpura, not related to thrombocytopenia
Additional criteria	Diffuse abdominal pain
	Histopathology: typical LVC with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits
	Arthritis or arthralgia
	Renal involvement (proteinuria: >0,3 g/24h or >30 mmol/mg of urine albumin to creatinine ratio on a spot morning sample; and/or hematuria, red blood cell casts: >5 red cells per high power field or ≥2+ on dipstick or red blood cell casts in the urinary sediment)

Most acute HSP cases resolves spontaneously,[8] often without therapeutic intervention.[9] The basic principles of HSP management consist of supportive care and symptomatic therapy.[10] Patient often need hospitalization to control the symptom. Bed rest may be required in patient with arthralgia.[9] Adequate hydration must be maintained orally or sometimes need intravenous fluid.[10]

The use of glucocorticosteroids (GCS) for HSP is still controversial. This treatment cannot be recommended in all patient because majority cases will improve spontaneously.[10] Oral GCS are indicated if the patient has severe rash, severe colicky abdominal pain, involvement of renal, scrotal, or terticular,[7] and in cases of severe joint pain resulting in immobilization.[11] Arthralgia or soft tissue edema with pain sensation usually get clinically improvement with nonsteroidal anti-inflammatory drug or acetaminophen, but 1 to 2 mg/kg/day oral prednisone may be useful to shorten duration of pain.[8] However, there is no agreement on the use of GCS for HSP which explain variable steroid dosage, steroid type and duration of treatment.[11]

Our patient had severe arthralgia and he had walking difficulty. We treated our patient oral methylprednisolone 12 mg/day divided in three doses for seven days and the other supportive management. This patient experienced significant improvement after hospitalization. Patient must be monitored for rare but severe complications such as hemorrhagic involvement of the kidney, lung, gastrointestinal, genitourinary and central nervous systems.[8]

Conclusion

HSP is the most common vasculitis in children but there remains uncertainty regarding optimal monitoring and therapy. HSP can affect multiple organ systems. Although majority HSP cases are self-limited, the possibility of serious complications must be monitored. We got a significant clinical improvement in our HSP patient who was treated with oral methylprednisolone and the other supportive management.

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