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Original Research Article

A unique and rare presentation of Henoch-Schonlein purpura: a case report

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ABSTRACT

Henoch-Schönlein purpura (HSP), or IgA vasculitis, is a common small vessel vasculitis in children, typically presenting with palpable purpura, abdominal pain, arthritis, and renal involvement. However, atypical presentations can delay diagnosis and management. A 12-year-old girl presented with multiple episodes of loose stools and abdominal pain. The initial diagnosis focused on acute gastroenteritis, and she was treated symptomatically. Subsequently, she developed urticarial rashes with breathing difficulty, complicating the diagnosis. Despite symptomatic treatment, she experienced severe abdominal pain, hematemesis and melena. Further investigations revealed antral gastritis, duodenitis, and cystitis. On the 14th day of admission, the patient developed characteristic palpable purpura on the lower limbs. Skin biopsy confirmed leukocytoclastic vasculitis with IgA deposits, consistent with HSP. The patient recovered with supportive care and remained symptom-free on follow-up. This case underscores the importance of considering HSP in pediatric patients with atypical presentations, such as gastrointestinal symptoms and urticaria without initial purpura. Early recognition and appropriate management are essential to prevent complications and ensure favourable outcomes. The variability in HSP presentations necessitates a high index of suspicion for timely diagnosis and treatment.

Keywords: HSP, IgA vasculitis, Pediatric vasculitis, Gastrointestinal symptoms, Urticaria, Palpable purpura

INTRODUCTION

IgA vasculitis, formerly known as HSP, is a complex immune-mediated vasculitis involving small blood vessels in various organ systems.[1] The diagnosis of HSP is best determined by the presence of purpura or petechiae (usually palpable) with a lower limb predominance in addition to 1 or more of the following four findings: abdominal pain (diffuse and colicky), arthritis or arthralgia, renal involvement (proteinuria >0.3 gm in 24 hours, morning urine albumin or creatinine levels of >30 µmol/L, or positive dipstick results for hematuria and positive histopathologic findings (leukocytoclastic vasculitis with predominant IgA deposits on skin biopsy, or proliferative glomerulonephritis with predominant IgA deposit on kidney biopsy).[2] The exact cause of the disease has not been identified to this date but various etiologies have been suggested such as exposure to varieties of pathogenic organisms, drugs, vaccinations, or malignancies.[3] HSP is usually self-limiting, however, severe fatal complications may occur. Early recognition of life-threatening complications is crucial to achieve a better outcome.[4] We are reporting a peculiar case of HSP with initial presentation of urticaria and gastrointestinal symptoms preceding palpable purpura which delayed the diagnosis of HSP.

CASE REPORT

A 12-year-old girl presented to the pediatric emergency with complaints of multiple episodes of loose stools, and abdominal pain for one day. No other significant history was noted. She was hemodynamically stable and was

started on treatment for acute gastroenteritis. On investigations, a complete blood picture showed neutrophilic leukocytosis with normal platelet count, liver function test, kidney function test, serum electrolytes, and blood sugar were normal. On the night of admission, the patient developed multiple itchy erythematous urticarial rashes all over the body associated with breathing difficulty. Repeat blood investigations showed neutrophilic leucocytosis but the chest x-ray was normal. She was treated symptomatically (antihistamines and steroids) and improved clinically. On the 5th day of admission, the patient started having severe abdominal pain, and multiple episodes of hematemesis with melena for which she was further evaluated. Her coagulation profile was normal, upper GI endoscopy showed antral gastritis with duodenitis, stool for occult blood was positive and ultrasound of the abdomen showed cystitis. She was started on intravenous antibiotics and urine samples were sent for routine examination and culture and sensitivity. Her urine routine examination report showed microscopic hematuria but repeated urine routine examination was normal. Stool examination for occult blood was repeated every day and after four days, it was reported as negative. Blood, urine, and stool cultures were sterile. Repeat liver function test, kidney function test, and serum electrolytes were normal.

On the 14th day of admission, the patient developed petechiae, and palpable purpura on bilateral lower extremities. Her vital signs were within normal limits and she was haemodynamically stable throughout her stay in hospital. A punch biopsy of skin was sent and the dermis showed a perivascular mixed inflammatory cell infiltrate composed predominantly of neutrophils and lymphocytes; inflammatory cells are seen in the vessel wall with leukocytoclasis. Direct immunofluorescence (DIF) study for Ig A shows immune deposits in the vessel wall. Ig G, Ig M, and C3 were negative. The patient recovered fully with symptomatic and supportive measures. Subsequent follow-ups didn't show any abnormality.

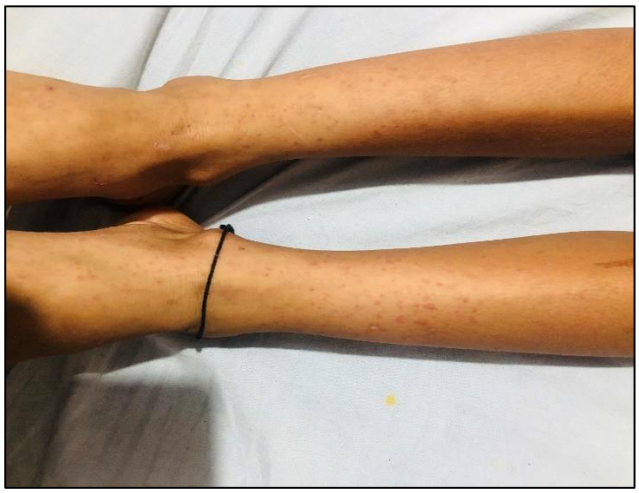

Figure 1: Palpable purpura on patient's lower limbs.

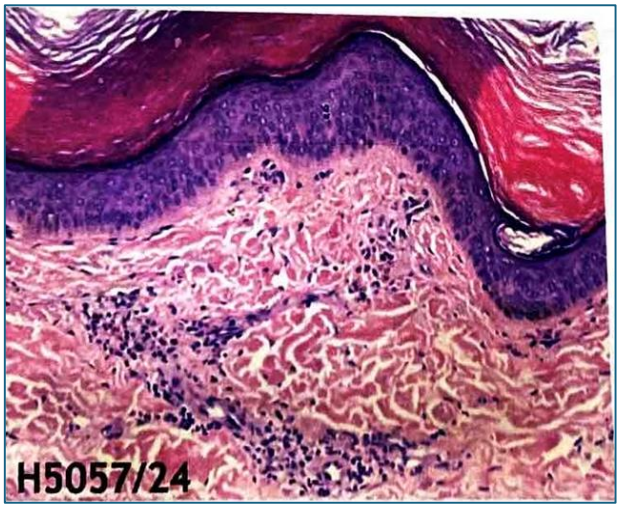

Figure 2: Punch biopsy of skin from right foot.
Dermis shows perivascular mixed inflammatory cell infiltrate composed predominantly of neutrophils and lymphocytes. Some inflammatory cells are seen in the vessel wall with minimal. leukocytoklasia.

DISCUSSION

HSP, or IgA vasculitis, is primarily a disease of children, characterized by small vessel inflammation predominantly affecting the skin, gastrointestinal tract, joints, and kidneys. The classic presentation includes palpable purpura, abdominal pain, arthritis, and renal involvement. This case is notable due to the atypical initial presentation with urticaria, neutrophilic leukocytosis with normal eosinophil count and gastrointestinal symptoms preceding the development of palpable purpura, which led to a delay in diagnosing HSP.

In this case, the 12-year-old patient initially presented with gastrointestinal symptoms, including loose stools and abdominal pain, which are not uncommon in HSP. However, the diagnostic challenge was due to the absence of purpura at the initial presentation and the development of urticarial rashes with breathing difficulty. CBC showed neutrophilic leukocytosis with normal eosinophil count. Gastrointestinal symptoms flared up after stopping steroid therapy for urticaria. It is important to consider that gastrointestinal symptoms can precede the characteristic rash in up to 30% of HSP cases, making early diagnosis difficult.[5,6]

The patient's progression to hematemesis, severe abdominal pain, and Malena suggested significant gastrointestinal involvement, which is a known complication of HSP. Gastrointestinal bleeding can be severe and is seen in about 10-30% of patients.[7] The presence of cystitis, microscopic hematuria and positive stool occult blood further indicated systemic involvement, which eventually led to the consideration of HSP upon the appearance of the characteristic purpuric rash.

Histopathological findings support a definitive diagnosis of HSP. In this case, the punch biopsy of the skin lesion revealed leukocytoclastic vasculitis with predominant IgA deposits, which is pathognomonic for HSP. DIF confirmed the presence of IgA deposits in the vessel walls, while IgG, IgM, and C3 were negative, consistent with the diagnosis.[8]

The patient was managed symptomatically and with supportive measures, including intravenous antibiotics for presumed bacterial cystitis and close monitoring of gastrointestinal symptoms and renal involvement. The resolution of bleeding manifestations is positive outcome in this case. Renal involvement, manifesting as hematuria or proteinuria, can occur in 20-60% of children with HSP and is associated with long-term morbidity.[9,10]

CONCLUSION

This case underscores the importance of considering HSP in the differential diagnosis of pediatric patients presenting with gastrointestinal symptoms and skin rashes, even in the absence of the classic purpuric rash. Early recognition and appropriate management are crucial in preventing complications and improving patient outcomes. The atypical presentation with urticaria and gastrointestinal symptoms highlights the variability of HSP and the need for heightened clinical suspicion to avoid delayed diagnosis and treatment.

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