Case Report

Henoch-Schönlein Purpura Associated with Gangrenous Appendicitis: A Case Report

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Submitted: 23 Oct 2012
Accepted: 20 Mar 2013

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Abstract

Henoch-Schönlein Purpura (HSP) is a leucocytoclastic vasculitis of unclear aetiology characterised by symmetrical, non-traumatic, nonthrombocytopenic purpura mostly involving the lower limbs and buttocks, as well as arthritis, gastrointestinal manifestations, and occasional nephritis. A 35 years old male presented with purpuric rash on the lower extremities, abdominal pain, fever, arthralgia, and melaena. A diagnosis of HSP with appendicitis was made, which is an exceedingly rare phenomenon.

Keywords: *abdominal pain, appendicitis, purpura, purpuric, rash, Schonlein-Henoch, vasculitis*

Introduction

Henoch-Schönlein Purpura (HSP) is a systemic hypersensitivity disease of unknown cause that is characterised by a purpuric rash and systemic manifestations, such as colicky abdominal pain, polyarthralgia, and acute glomerulonephritis. HSP is predominantly a small-vessel vascular disease that most often affects the skin and internal organs (1). Common complications of HSP that lead to surgical intervention include intussusception, perforation, necrosis, and massive gastrointestinal bleeding. Acute appendicitis is rarely seen as a complication of HSP (2).

Case report

A 35 years old man presented with intense abdominal pain, fever, swelling of left ankle joint, and multiple itchy and erythematous rashes of one-week duration. His illness started with pain in both knee and ankle joints two weeks prior to the onset of the rash, for which he had taken Ayurvedic Indian medications. There was no history of dysuria, loose stools, or vomiting. On examination, palpable purpuric lesions were concentrated on the buttocks, lower extremities (Figure 1), and abdomen, with tenderness and guarding at McBurney's point.

Routine blood investigations were within normal limits except for mild leukocytosis and a mild rise in erythrocyte sedimentation rate (ESR). A skin biopsy taken from the purpuric lesions showed leucocytoclastic vasculitis (Figure 2). A stool benzidine test for occult blood was strongly positive. Platelet count, bleeding time, clotting time, serum amylase, lupus erythematosus (LE cell), hepatitis B surface antigen (HBsAg), liver function tests, and renal function tests were within normal limits, and an abdominal ultrasound revealed an inflamed appendix. Appendectomy

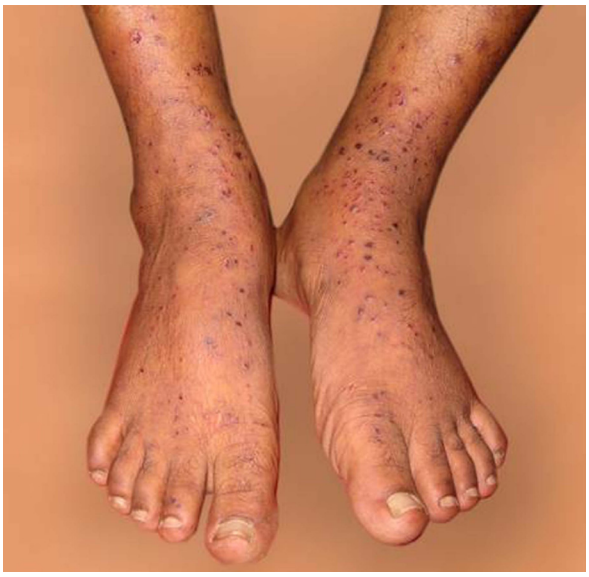

Figure 1: Rashes over bilateral lower extremities.

was performed, and the appendix was gangrenous upon histopathological examination. Following the operation, the colicky abdominal pain and fever were resolved, but the purpuric lesions persisted even after surgery, though with less severity. Complete cure did not occur until 16 days after surgery, and recurrence was noted once after two months, but patient recovered completely with symptomatic treatment.

Discussion

HSP is an IgA-mediated systemic small-vessel vasculitis commonly affecting joints, the gastrointestinal tract, and kidneys (3). These manifestations are due to the deposition of circulating immune complexes, comprised of polymeric IgA and complement component 3 (C3), in arterioles, capillaries, and venules throughout the body. It may develop in all age groups, from 4 months to 89 years (4). Abdominal symptoms and signs of HSP precede the emergence of the rash in 14–36% of cases, as seen in the present case, which makes initial diagnosis difficult and may even result in unnecessary laparotomy (1,5,6). Vascular involvement of the intestine may produce signs that mimic acute appendicitis and lead to an unnecessary appendectomy (2). Major complications of abdominal involvement develop in 4.6% of cases (range 1.3–13.6%); of these, intussusception is by far the most common (6). A few complications, like bowel ischemia and infarction, appendicitis, intestinal perforation, fistula formation, late ileac stricture, massive upper gastrointestinal haemorrhage, pancreatitis, hydrops of the gallbladder, and pseudomembranous colitis, are seen infrequently. Diagnosis of HSP is based on clinical manifestations and laboratory investigations considering the criteria described by the American College of Rheumatology (7) and the International Consensus Conference, 2006 (8,9).

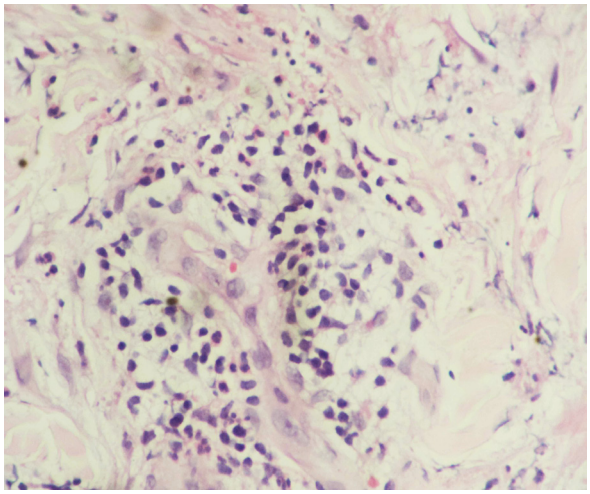

Figure 2: Haematoxylin and eosin stained section of purpuric lesion (400× magnification), showing leucocytoclastic vasculitis.

Conclusion

In conclusion, HSP is a multisystem vascular disorder with a number of complications, especially musculoskeletal, gastrointestinal, and renal. A few of the complications, particularly those that are gastrointestinal, require emergency surgery. Serial clinical assessment and ultrasonography are necessary to clarify the exact nature of the gastrointestinal involvement and to reduce the likelihood of unnecessary surgery.

Acknowledgement

None.

Conflict of Interest

None.

Funds

None.

Authors' Contributions

Conception and design, drafting of the article, critical revision of the article for the important intellectual content, final approval of the article, provision of study materials or patient and administrative, technical or logistic support: NKS, SA

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