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Case Report

Acute appendicitis as the first manifestation of Henoch-Schönlein purpura

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ABSTRACT

Henoch-Schönlein purpura (HSP) is a small vessel systemic vasculitis. Typical symptoms include palpable purpura, joint pain, and abdominal pain. Most cases improve after a few weeks, not requiring any treatments other than symptom control. Acute abdomen resulting from vasculitis is very rare and should be treated as a surgical emergency. We report the case of a 9-year-old boy with acute gangrenous appendicitis as the first manifestation of HSP.

Keywords: Acute abdomen, Appendicitis, Henoch-Schönlein purpura

INTRODUCTION

Henoch-Schönlein purpura (HSP) is a systemic idiopathic vasculitis related to deposits of immune complexes, mainly containing immunoglobulins A (IgA) on the walls of small vessels. The disease is characterized by the association of cutaneous, joint, and gastrointestinal signs.[1] Skin involvement is almost constant and is the first sign of HSP in more than two-thirds of the cases. The course of this illness is usually self-limited, with a good prognosis in most cases.[2] Digestive manifestations can be moderate, spasmodic pain, but they can be severe, leading to a surgical outcome.[3] We report the case of a 9-year-old male patient with acute gangrenous appendicitis as the first manifestation of HSP.

CASE REPORT

A 9-year-old male patient was taken to the emergency room due to a complaint of abdominal pain for a week. It had started as a feeling of discomfort in the abdominal region associated with nausea, anorexia, and unverified fever. The mother reported that the child had just had the flu and she suspected that the abdominal pain could be part of the infectious disease. However, the boy began to feel a lot of pain in the region of the right iliac fossa, and his parents sought for medical help. Physical examination revealed that he was pale, dehydrated, and afebrile, with pain from diffuse palpation of the abdomen and showed defensive reflexes of the abdominal muscles. The boy underwent a laparotomy that evidenced acute gangrenous appendicitis complicated by purulent peritonitis [Figure 1].

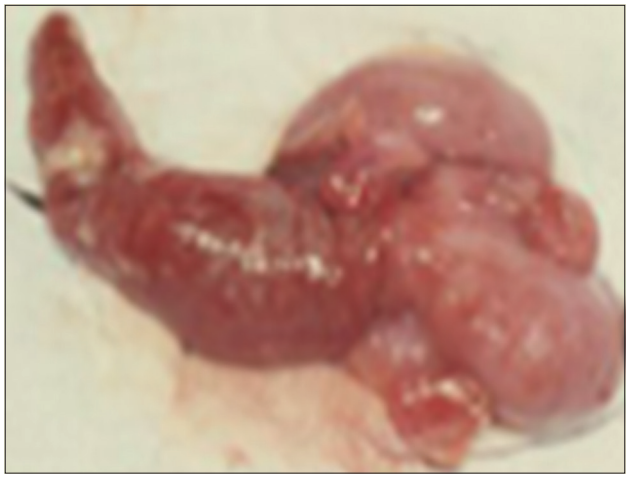

Figure 1: Surgical specimen showing an appendix with inflammation, hyperemia, and signs of necrosis.

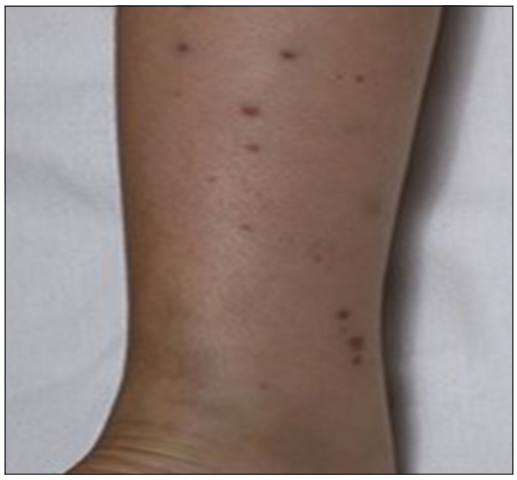

Figure 2: Palpable purpura in the lower limb.

On the 2nd post-operative day, he remained dehydrated and presented with tachycardia, tissue perfusion of 4 s, diffuse abdominal pain, decreased breath sounds (hydrothorax), arthralgia in the wrists, and presence of palpable purpura in the lower limbs and gluteal region [Figure 2].

Laboratory tests showed leukocytosis with the left shift associated with a platelet count of 690,000, and the imaging tests revealed no changes. The anatomical pathology examination confirmed the diagnosis of acute gangrenous appendicitis [Figure 3].

We decided to treat the patient postoperatively with a combination of ceftriaxone and metronidazole, associated with methylprednisolone, resulting in significant improvement of the condition. The patient showed good evolution, was discharged uneventfully 2 weeks after the surgery, and was followed up at the outpatient clinic.

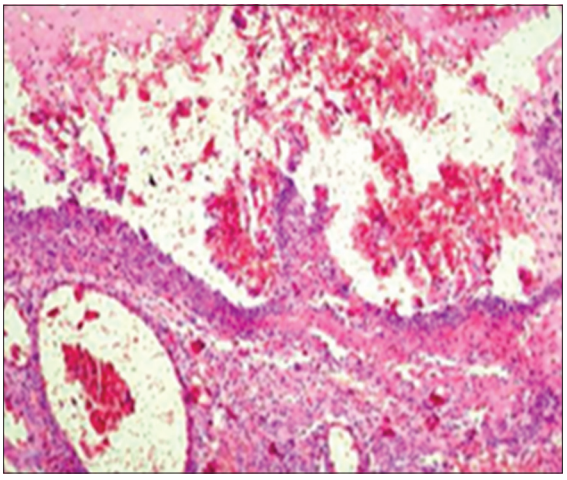

Figure 3: Acute appendicitis with fibrinopurulent exudate in all layers of the appendix.

DISCUSSION

Most cases of HSP are preceded by an upper respiratory tract infection, suggesting a potential infectious trigger.[4] The most characteristic pathological feature is a deposition of immune complexes containing IgA on the vessel walls of the affected organs and the renal mesangium.[5]

The main manifestations are palpable purpura, generally painless and non-pruritic, located predominantly in the lower limbs and gluteal region. Enteric vasculitis, leading to ischemia, edema, and hemorrhage, can occur in two-thirds of HSP cases, and the major symptom is abdominal pain of varying intensity. The acute abdomen condition may occur concurrently or preceding the skin rash and be associated with vomiting, transient paralytic ileus, ischemia, and necrosis. Intestinal perforation, massive intestinal bleeding, pancreatitis, pseudomembranous colitis, appendicitis, and fistula formation are more rare.[3]

The diagnosis of HSP is based on clinical criteria, i.e., palpable purpura (mandatory criterion) and at least one minor criterion: (1) Diffuse abdominal pain; (2) arthritis or arthralgia; (3) renal involvement; and (4) biopsy showing leukocytoclastic vasculitis with a predominant deposit of IgA or proliferative glomerulonephritis with a predominant deposit of IgA. The sensitivity of clinical criteria is 100%, and the specificity is 87% when applied to children.[6] Applied to adults, the diagnostic sensitivity is 99.2%, and the specificity is 86%, supporting this use in all patients with HSP.[7]

The treatment is controversial and depends on kidney involvement. In patients with the absence of renal involvement, the treatment is purely symptomatic. Pain medications, rehydration therapy, and surgery should be employed in specific cases.[8]

CONCLUSION

Gastrointestinal manifestations of HSP may precede other clinical manifestations of HSP by days, and although acute appendicitis is a very rare complication, health professionals should be aware of this possibility.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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