Mesenteric Panniculitis as Cause of Abdominal Pain in Henoch-Schönlein Purpura

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Introduction

Henoch-Schönlein purpura (HSP), also known as anaphylactoid purpura [1], is the most common form of systemic vasculitis in children [2,3]. Most cases occur between the ages of 4 and 7 years [1]. HSP has several clinical and laboratory manifestations. Palpable purpura can be identified in practically all patients, most of whom develop polyarthralgia with a lack of frank arthritis. Gastrointestinal manifestations occur in 70% of children and mainly consist of colicky abdominal pain, primarily due to intestinal submucosal bleeding and edema that can lead to intussusception. Renal involvement takes place in 10-50% of cases and is characterized by glomerulonephritis and proteinuria. In 50% of patients, serum IgA level is found to be elevated. A skin biopsy might be needed in patients with unusual presentation, and whenever performed, it shows the classical leukocytoclastic vasculitis in postcapillary venules with IgA deposition [1]. Other less frequent causes of abdominal pain in patients with HSP include: bowel ischemia, intestinal perforation, acute pancreatitis, gallbladder involvement, acute appendicitis, massive GI bleeding [2,3], among others.

Martin-Sune et al. reported the first case of mesenteric panniculitis associated with HSP in a 37 year-old patient at La Paz Hospital, Madrid, on October 2010 [4]. This report describes the 2nd case of mesenteric panniculitis in patient with HSP that responded to corticosteroid therapy and colchicine.

Case Report

In September 2013, a 50 year-old man came to our emergency department because of severe colicky abdominal pain that started few hours prior to his presentation. Note that the patient was admitted ten days before to the internal medicine ward with lower limbs purpura (Figures 1 and 2), arthralgia and abdominal pain, his IgA level was found to be above normal (4.68 g/dL), and skin biopsy confirmed the diagnosis of HSP by showing the pathognomonic leucocytoclastic vasculitis. He was treated with steroids at a dose of 0.7 mg/Kg (70 mg daily) initially then tapered slowly, before being discharged once he started to improve, without further investigations.

At his second presentation, the patient only had minimal purpura but associated with severe abdominal pain. On physical examination, peri-umbilical and right lower quadrant tenderness were noted. Laboratory tests showed elevated white cells count (17'600/mm3), probably due to steroids therapy. Otherwise, the patient had normal liver function tests and normal pancreatic enzymes levels (amylase and lipase). An upper gastrointestinal tract endoscopy was performed to investigate his abdominal pain, but it showed normal findings (e.g. no submucosal bleeding or swelling, no gastritis, no ulcers). Finally, a Computed Tomography (CT) scan of the abdomen showed increase fat density in the mesentery centrally at the level of the umbilicus (Figures 3 and 4) leading to the diagnosis of mesenteric panniculitis.

Prednisone dose was increased to 1 mg/Kg (100 mg daily), and colchicine was added at a dose of 1 mg daily. The condition of the patient started to improve rapidly. His abdominal pain resolved along with his purpura. The patient was evaluated two months later with a follow-up CT scan of the abdomen which showed total resolution of the mesenteric panniculitis that was present in previous images.

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Discussion

There is only one previous case in the literature reporting mesenteric panniculitis as cause of abdominal pain in patient with HSP. That patient was a 37 year-old female who presented to La Paz hospital in Madrid with abdominal pain, fever, and arthralgia. She developed purpuric lesions on her lower extremities two days later. HSP was diagnosed by biopsy of one of these lesions, and a CT scan of the abdomen showed mesenteric panniculitis. The patient was successfully treated by oral corticosteroid therapy at a dose of 1 mg/Kg [4].

Three observations are worth-noting in our case report. First, the age of the patient is remarkable, because 90% of HSP occur in children, and the remaining 10% are divided between infants and adults [1-3]. Second, the patient had mesenteric panniculitis, a rare condition causing chronic inflammation involving the adipose tissue of the mesentery, usually complicating an abdominal surgery, or an abdominal malignancy; and is very rarely associated with vasculitis [4]. Third, the patient did not respond to moderate dose of steroids alone (0.7 mg/Kg), but his condition required colchicine (1 mg daily) and a full dose of prednisone (1 mg daily).

HSP is relatively a frequent condition, but mesenteric panniculitis is quite rare and it is an exceptional cause of abdominal pain in patients with HSP (only a single case previously reported) [4]. Mesenteric panniculitis is usually secondary to certain intra-abdominal stimuli (abdominal surgery or trauma [5-7], malignancy [4], mesenteric thrombosis [8], inflammatory bowel diseases [4], thermal or chemical injuries [8], retained suture material [8], bile or urine leakage [8], among others). Previous studies showed a male gender predominance (with a male/female ratio of 2.3:1), and some reports found that its incidence increases with age, most probably because adults have much more mesenteric fat compared to children [8]. This fact could explain why mesenteric panniculitis occurred in two adult patients with HSP, since the age is the only common feature between our patient and the case reported by Martin-Sune et al. [8] In 90% of cases, the inflammation of mesenteric panniculitis occurs in the small bowel mesentery [8]. The patient can be asymptomatic, or may have abdominal pain, nausea, vomiting, anorexia, and altered bowel habits [8], among other symptoms. The condition can progress slowly over months and can lead to fibrosis, scarring, retractions and obstructive symptoms [8]. Definite diagnosis is obtained by biopsy of the mesentery but it is rarely needed nowadays because of the presence of high-resolution CT scans and Magnetic Resonance Imaging (MRI) [5,9-11]. Treatment is reserved for symptomatic cases only, but no consensus has been established yet [8]. Some clinicians use prednisone only at a dose of 30-50 mg daily [5], others use one of the following agents with or without steroid therapy: colchicine (1 mg/day) [12,13], azathioprine (30-100 mg/day) [14,15], cyclophosphamide (2 mg/Kg) [16], progesterone or tamoxifen (10 mg/day) [17,18].

The patient in our case report received high dose prednisone (100 mg daily) in addition to colchicine (1 mg daily), and once his condition improved, colchicine was stopped and we started down tapering his prednisone dose over a period of 6 months before stopping it completely.

Conclusion

Mesenteric panniculitis has been added to the long list of etiologies of abdominal pain associated with HSP. In specific cases, when diagnosis is obscure, early usage of radiographic and invasive tests can lead to earlier definite diagnosis and permit earlier initiation of adequate therapy, thus preventing possible complications.

References


