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Case Presentation
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Acute Pancreatitis Revealing Henoch-Schönlein Purpura

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Abstract

Henoch-Schönlein purpura (also known as IgA vasculitis) is a systemic vasculitis of small vessels. It mainly affects children. The clinical spectrum of the disease mainly includes cutaneous purpura, arthralgias and/or arthritis, acute enteritis and glomerulonephritis. The short-term prognosis of the disease depends on the severity of the digestive involvement and the long-term prognosis on the renal involvement. Pancreatic involvement is exceptional. The physiopathological mechanism seems to be vascular by analogy with the renal and cutaneous involvement. Treatment remains not clearly defined. We report the case of a patient with IgA vasculitis complicated by pancreatitis and successfully treated with corticotherapy.

Keywords: Henoch-Schönlein purpura; Pancreatitis; Digestive involvement; Intestinal involvement; IgA vasculitis

Introduction

Henoch-Schönlein purpura a systemic vasculitis of small vessels resulting from tissue deposits of immune complexes containing type A Immunoglobulins (Ig). It mainly affects children and rarely adults. It can affect the skin, joints, kidney, gastrointestinal system, more rarely the lung, heart and nervous system [1]. Diagnosis requires an anatomical and clinical analysis. The prognosis is variable and is mainly related to digestive and renal involvement [2].

Digestive involvement in Henoch-Schönlein purpura is varied and can occur as an initial manifestation. It includes intestinal intussusception, perforation, mesenteric ischemia and pancreatitis which is a rare and poorly described complication of this disease. A recent review of all cases of Henoch-Schönlein purpura-related pancreatitis showed a higher incidence in males and highlighted the importance of timely treatment with steroids to obtain a good outcome [3]. The pathophysiological mechanism is complex and poorly understood. It is explained by a vascular involvement of the capillaries, small arteries and small veins within pancreatic tissues, which increases vascular permeability and leads to pancreatic edema [4]. We report a patient with IgA vasculitis purpura complicated by acute pancreatitis.

Case Presentation

A 20 year old woman with a medical history of 3 episodes of spontaneously resolving purpura decline without associated digestive or respiratory signs. She consulted for a 4th episode of purpura of the lower limbs (Figure 1) associated with severe abdominal pain, of the epigastric region without vomiting or digestive hemorrhage, with inflammatory arthralgias of the elbows and knees. There was no precessive infectious episode. The clinical examination revealed purpura of the lower limbs reaching the thighs without ulceration nor necrosis nor nodules. The biology assessment showed an elevated CRP (110 mg/L) with neutrophil hyperleukocytosis (15.1 G/L including 11.7 G/L neutrophils) and hyperlipaemia (600 U/L). The liver test showed anicteric cholestasis (GT: 115 U/L) without cytolysis. Triglyceride levels and blood calcium levels were normal, glomerular filtration rate, urine sediment examination and 24-h proteinuria were normal. The immunological tests, including anti-double-stranded DNA, anti-SSA, and ANCA antibodies, were negative, and the CT scan performed three days after the onset confirmed Balthazar B pancreatitis without biliary tract lithiasis (Figure 2). Skin biopsy of purpura showed leukocytoclastic vasculitis. Immunofluorescence testing on a small fragment did not reveal immunoglobulin A deposits, but the diagnosis of Henoch-Schönlein purpura was made based on the association of infiltrated purpura, diffuse abdominal pain and arthralgias according to the ACR 1990 and EULAR/PRES/PRINTO criteria. The patient was treated with oral corticosteroid therapy (1 mg/kg/day) with methylprednisolone bolus. The purpura and abdominal pain progressively resolved within a few weeks without relapse with normalization

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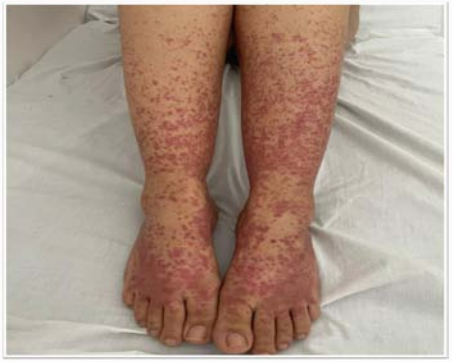

Figure 1: Purpura of the lower limbs.

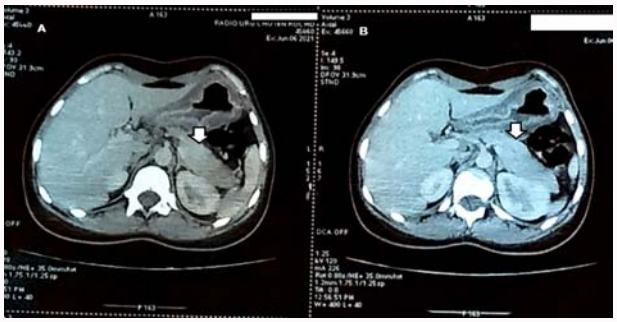

Figure 2: Enlarged and edematous pancreas: Balthazar stage B pancreatitis.

of CRP and lipasemia.

Discussion

We report an original case of Ig A vasculitis complicated by pancreatitis. Indeed, gastrointestinal involvement in IgA vasculitis is as common in children as in adults [2,5,6], with a prevalence in adults varying from 37% to 65%. Abdominal pain and intestinal bleeding were consistent in 31% of cases, diarrhea in 26% and nausea or vomiting in 19% [5]. The duodenum and small intestine are the two segments most frequently involved in IgA vasculitis with gastrointestinal involvement [7,8].

The most common complications of IgA vasculitis are gastrointestinal intussusception, perforation, obstruction or hemorrhage and pancreato-hepatobiliary involvement. The latter is unusual and includes pancreatitis, alithiasic cholecystitis and gallbladder perforation. The involvement of the pancreas is probably due to the presence of small vessel vasculitis lesions. It is diagnosed in patients with abdominal pain and an amylase or lipase level at least twice the upper limit of normal, in the absence of alcohol abuse, gallstones or the use of drugs that can cause pancreatitis [9]. Ultrasound and/or CT scans are beneficial. They reveal thickening of the intestinal wall, inflammation of the mesentery, sub-mucosal hematomas, pancreatic damage, and endoscopy is also useful for assessing lesions in the digestive tract, revealing petechial purpura, erosions and even areas of necrosis. The management of IgA vasculitis is particularly challenging due to the lack of correlation between the initial presentation and the potential risk of developing severe gastrointestinal involvement or progression to end-stage renal disease [10,11]. IgA vasculitis is managed primarily by bed rest, which limits the extension of the purpura to the skin, but does not affect the digestive or renal systems. The use of corticosteroids remains controversial. Their administration to reduce the rate of renal and gastrointestinal complications is the subject of several studies. Indeed, the authors failed to find a statistically significant difference in the risk of gastrointestinal and renal complications between the corticosteroid group and the control group [12]. A prospective study has shown that corticosteroids are beneficial in the treatment of abdominal pain [13] and although these results are consistent, they cannot be considered definitive proof. Similarly, a study evaluating the efficacy of glucocorticoids in preventing the obstruction of severe gastrointestinal complications such as perforation remains controversial, as the majority of patients improved spontaneously [14]. In adults, Pillebout et al. reported a randomized controlled trial evaluating glucocorticoids alone *vs.* glucocorticoids and cyclophosphamides in 54 patients with severe IgA vasculitis, including severe bleeding, ischemia, perforation or persistent abdominal pain not responding to standard analgesics. This study suggests that treatment of adults with severe IgA vasculitis with the addition of cyclophosphamides did not provide any benefit over glucocorticoids alone [15].

Conclusion

Henoch-Schönlein purpura is a systemic vasculitis with a good prognosis globally. Renal and digestive disorders are predictive of its severity abdominal symptoms in the context of IgA vasculitis should be monitored closely. The pancreatic enzymes should be checked systematically for elevation. In our case, early diagnosis of pancreatitis and initiation of steroids after excluding other causes of pancreatitis avoided unnecessary surgical interventions.

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