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Case Report

Severe Acute Pancreatitis and Behcet's Disease (Case Report)

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Received: June 22, 2023; **Published:** July 28, 2023

Abstract

Behcet's disease is a vasculitis of unknown etiology that affects blood vessels of all sizes, and in which venous lesions are remarkably frequent. Patients with this condition experience recurrent symptoms, whose severity and frequency decrease over time. However, complications may be severe. Although the mortality rate - mainly due to major vessel disease - is relatively low, Behcet's disease is associated with high morbidity rates. Diagnosis is based on certain characteristic symptoms, primarily oral or genital-oral ulcerations, as well as other systemic manifestations. Pancreatitis is yet another but rare manifestation, scarcely mentioned in literature. We report the case of a patient with Behcet's disease who presented an acute pancreatitis.

Keywords: *Acute Pancreatitis; Behcet; Organ Dysfunction*

Introduction

Behcet's disease is a chronic systemic vasculitis that affects all different sizes of arterial and venous blood vessels [1-3]. Its cause is yet unknown. Its evolution is marked by unpredictable relapse periods associated with high morbidity and significant mortality rates, depending on the organ systems involved and the patient's comorbidities. Behcet's disease is seen mainly in the third decade of life, with at least a 10-year difference between the first symptoms and the diagnosis which is essentially clinical [4]. Although the incidence of gastrointestinal manifestations varies widely, the pancreas is rarely involved. We report the case of a young male patient with Behcet's disease presenting a severe acute pancreatitis.

Case Report

A 17-year-old man was admitted to the emergency room for severe epigastric pain radiating to the back and incoercible vomiting for four hours straight. The patient had history of interauricular defect surgery and clinical diagnosis of Behcet's disease at a young age. His disease was controlled regularly and treated with corticosteroids and azathioprine. In the initial evaluation, the patient was found conscious with a GCS at 15/15, eupneic with a blood oxygen saturation level of 99%, a heart rate at 126 bpm and a blood pressure at 120/80 mmHg. His temperature was at 37,2ºC and his blood sugar level at 1,5 g/L. The abdominal examination found no guarding or rigidity. Initial investigations found a hemoglobin level at 11,2 g/dL, a leukocyte count at 13000/mm^3, a urea level at 0.65 g/L, a creatinine level at 13,7 mg/L, AST level at 32 UI/L, ALT level at 26 UI/L and an elevated lipase level at 926 UI/L. An abdominal CT scan showed a pancreatitis Balthazar Grade E, with pancreatic necrosis. A complementary abdominal echography revealed no further abnormalities. No remarkable inflammatory syndrome was detected as C-reactive protein level was at 96 mg/l and procalcitonin level was normal. Treatment with aza-

thioprine was stopped, and corticosteroids were started. The rest of the treatment included the insertion of a nasogastric tube, analgesic and antiemetic drugs, gastroprotection with a PPI, thromboembolism prophylaxis, multivitamins supplementation and parenteral nutrition. The patient rapidly improved both clinically and biologically, and was transferred shortly, after a period of 4 days in the ICU, to the Internal Medicine Department.

Discussion

Behcet's disease is a chronic systemic inflammatory disease first described by Turkish dermatologist Hulusi Behcet in 1973 who recognized its three characteristic symptoms which are oral and genital ulcerations and uveitis with hypopyon [1-3]. The cause is yet unclear, however, several factors have been thought to contribute including genetic, infectious and immune factors. It is a vasculitis resulting in occlusion of venous vessels rather more frequently that arterial vessels. The mean age of onset is between 10 and 45 years old, and males are generally more involved than women [4]. This disease, however diagnosed in many countries around the world, is most common in the Middle East, Asia and Mediterranean countries [4]. Diagnosis is mainly clinical, based on constantly reviewed guidelines, essentially stating Mason and Barnes [5], Japanese [6], O'Duffy [7], Hamza [8] and The International Study Group for Behcet's Disease criteria [9]. The latter has defined 5 items in the ISG criteria for the diagnosis of the Behcet's disease: mucosal lesions (oral and genital aphthous ulcers), skin lesions (erythema nodosum, pseudofolliculitus), ocular manifestations (anterior uveitis, posterior uveitis, retinal vasculitis) and a Pathergy reaction. More recently, Davachi has proposed ICBD criteria with a higher sensibility compared to ISG criteria, and a reasonable specificity. Thus, these new criteria have been adopted as a guide for the diagnosis and classification of Behcet's Disease. They include, on top of the 5 ISG items, vascular manifestations (superficial phlebitis, deep vein thrombosis, arterial thrombosis and aneurysms).

Other systems can also be affected in Behcet's disease: articular manifestations were reported in 39,4% of patients and mainly included arthralgia, monoarthritis and polyarthritis, neurological manifestations, also called Neuro Behcet's Disease, were reported in 10,6% of patients, cardiovascular manifestations were reported in 0,6% of patients and included pericarditis, myocarditis, endocarditis, mitral valve prolapse, valvular lesions, intracardiac thrombosis, endomyocardial fibrosis, cardiomyopathies (myocardial infarction), and coronary artery lesions (coronary aneurysms), renal manifestations were scarcely reported and mainly included amyloidosis (the most frequent although rare) and glomerulonephritis, and last but not least, gastrointestinal manifestations were also reported, essentially caused by a vasculitis affecting venules or, alternatively, by an unspecific inflammatory syndrome.

Gastrointestinal manifestations, also known as entero-Behcet, were reported in 7% of patients with Behcet's Disease. They appear generally 4 to 6 years after first oral ulcerations, and are more frequent in Japan than in Mediterranean countries and The Middle East. They are due to mucosa lesions and ulcerations that can be found in various parts of the digestive tract, especially in the ileocecal region. The esophagus, ascending colon and transverse colon are the least affected regions. Main symptoms include anorexia, vomiting, dyspepsia, diarrhea, melena, abdominal pain and, less frequently, gastrointestinal perforation that requires immediate medical care. Whereas the incidence of gastrointestinal manifestations may vary, pancreatitis is a rather rare one, hardly mentioned in literature. However, the possibility of an underdiagnosed association of Behcet's Disease with pancreatitis has been suggested, for in an autopsy series of 170 cases in Japan, 5 cases of pancreatitis were found without being priorly diagnosed. Characteristic symptoms include epigastric pain with back radiation, incoercible vomiting, gastrointestinal transit abnormalities such as diarrhea and weight loss. In our case, the patient had presented epigastric pain and vomiting. Biological exploration must include inflammatory markers such as leukocyte count and C reactive protein. Pancreas imaging using an abdominal CT-scan may also be helpful in diagnosis. In the case of our patient, diagnosis was mainly clinical, in addition to an elevated lipase level and a suggestive abdominal CT-scan. No clear cause of the pancreatitis has been identified, but several potential etiologies were taken to account: autoimmune cause (search for IgG may be helpful), biliary origin (a hepatobiliary echography is used to eliminate such a possibility), drugs (azathioprine may be responsible of an acute pancreatitis 3 to 5 weeks after its introduction, as been reported from 2 patients in literature and who improved after stopping the therapy), and other risk

factors (alcohol use, smoking). Our patient was neither alcoholic nor a smoker, had no history of previous infections, and had a normal calcium and triglycerides levels. All these arguments have led the decision of the authors to stop azathioprine and start corticosteroids, hence allowing a better clinical and biological evolution. It is important to note that the precedent observation, reporting the 2 patients with azathioprine-induced pancreatitis, revealed that both had pseudoaneurysms of the coeliac and superior mesenteric arteries. These observations suggest that the main cause of intestinal lesions could be a vascularitis, so its contribution to an acute pancreatitis may be possible. No consensus is actually made for treatment of an acute pancreatitis associated with Behcet's Disease, so a treatment of the main gastrointestinal symptoms may be efficient [10-26].

Conclusion

Although acute pancreatitis is a rare manifestation of Behcet's Disease, it should always be considered as a differential diagnosis if a patient with this disease presents abdominal pain and/or vomiting. A cohort study including a larger group of patients is necessary for a better understanding and for more refined therapy strategies regarding this pathology, hence allowing an improvement in its morbidity and mortality rates.

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Volume 6 Issue 9 September 2023