CLINICAL CASE

Idiopathic mesenteric venous thrombosis: Report of a case

Thrombose veineuse mésentérique idiopathique : à propos d’un cas clinique

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Received 17 December 2007; accepted 19 February 2008
Available online 18 April 2008

KEYWORDS
Superior mesenteric venous thrombosis; Hypercoagulable state; CT abdominal scanner

Summary
Introduction. — Idiopathic mesenteric venous thrombosis is a rare entity. An early diagnosis and thrombolytic and anticoagulant therapy are very important.
Patient and methods. — We report a case of a patient, without any specific known risk factor, with small intestinal ischemia secondary to superior mesenteric vein thrombosis (SMVT).
Results. — In our case, only a computed tomography (CT) abdominal scan permitted the diagnosis of SMVT. The patient was successfully treated by resection of the infarcted bowel with primary anastomosis and immediate postoperative anticoagulation.
Conclusions. — Diagnosis of intestinal ischemia from mesenteric venous thrombosis (MVT) is often delayed because the symptoms are nonspecific. Moreover, when there is not any known predisposing factor, the diagnosis may become even more difficult with significant morbidity and mortality. CT abdominal scan done early in case of nonspecific abdominal pain, since the patients had a previous history of venous thrombosis, may not require a surgical treatment of MVT.

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MOTS CLÉS
Thrombose veineuse mésentérique supérieure ;
Introduction

Idiopathic superior mesenteric vein thrombosis (SMVT) is defined as a thrombosis of the superior mesenteric vein without any other related disease or etiologic factor. This pathology is an uncommon type of intestinal ischemia associated with significant mortality and morbidity because of its delay in diagnosis. In contrast to mesenteric infarction due to arterial occlusion, infarction due to venous thrombosis is subacute in its presentation and involves a limited, usually well-demarcated segment of bowel [1,2].

The diagnosis of SMVT is difficult because the symptoms are nonspecific, often associated with a spontaneous, intense and continued abdominal pain without any other specific symptom.

The imaging, abdominal ultrasound and CT are difficult to interpret for the exiguity of the signs of this pathology [2,3]. In early diagnosis, the thrombolytic and anticoagulant therapy, using unfractionated heparin or low molecular weight heparin, is really important as a first approach. If there are signs of peritonitis due to bowel necrosis, immediate surgical exploration is necessary.

We present an unusual case of a 51-years-old patient presenting with acute SMVT without any known predisposing disorder. This case expresses the really nonspecific clinical presentation of the idiopathic MTV.

Case report

A woman, 51 years old, was admitted to our hospital with a persistent and continuous abdominal and lumbar pain with tenderness. Laboratory tests showed a modest increase of white blood cells (WBC) count and of the D-dimer levels. The abdominal x-ray resulted negative, while the abdominal color flow Doppler sonography (CFDS) demonstrated a modest thickening of the appendix’s wall with signs of inflammation and the absence of free fluid in the Douglas’s space. The patient was treated by intravenous antibiotic therapy which led to a quick recovery with normalization of the sonographic report. After seven days, patient was discharged, but three days later, she was again hospitalised for the recurrence of the abdominal symptoms. CT abdomen scans showed the thrombosis of the superior mesenteric vein (Fig. 1). Thus, an emergency laparoscopy was performed, which revealed segmental intestinal infarction caused by the thrombosis in the SMV. We found a necrotic intestinal tract of approximately 50 cm of length and we performed a segmental resection of the involved bowel with primary anastomosis.

The postoperative period was good. The patient was given intravenous 24,000 units heparin sodium per day started immediately after the operation. On seventh postoperative day, she has started on sodium warfarin, and the heparin was stopped when a therapeutic level was achieved. After eight days, the patient was discharged.

A search for a precipitating condition revealed no evidence of a hypercoagulable state, myeloproliferative disorder, malignancy or a previous history of venous thrombosis.

The patient was subjected to a thrombophilia screen consisting of protein C, S, antithrombin levels, lupus anticoagulant, antiphospholipid antibodies, fibrinogen levels, factor VIII levels, factor V Leiden gene mutation and paroxysmal nocturnal hematuria screen. All coagulation tests resulted normal.

His immediate family was screened for thrombophilia and no evidence of any known predisposing disorder was found. All family members tested had normal coagulation.

On examination one year after the operation, the patient is well and has no sign of venous thrombosis. She will remain on lifelong anticoagulation with sodium warfarin.

Discussion

The idiopathic MTV is a rare cause of acute abdomen and it is defined as thrombosis of a SMV without any known predisposing factors in hypercoagulable condition due to heritable or acquired disorders of coagulation.

The most common hypercoagulable disorders are deficiencies in the intrinsic plasma anticoagulant system such as a resistance to activated protein C, prothrombin mutations, protein S, protein C and antithrombin III deficiencies and presence of factor V Leiden gene mutation [4–8]. Other causes of prothrombotic states are inflammatory bowel diseases, thrombophilia, cirrhosis, portal hypertension, oral-contraceptive use [9], abdominal neoplasm, paroxysmal nocturnal hemoglobinuria, essential thrombocytosis, polycytemia vera, intra-abdominal processes, trauma, inflammatory bowel disease and the postoperative state [1–3,10–12].

MVT is classified as either primary or idiopathic when any etiologic factor is found or secondary.

The proportion of patients with primary MVT continues to decline as our ability to diagnose inherited thrombotic
suspected acute superior mesenteric occlusion [18]. In our case, blood chemistry revealed a leukocytosis with a modest increase of D-dimer levels but these may be also high in patients with inflammatory disease or in patients with familial thrombosis history was not present. Past medical and family history is essential in making the correct diagnosis. However, in our patient, personal and familial thrombosis history was not present.

SMV thrombosis can induce ischemia or infarction of the small and large bowel. Bowel infraction due to SMVT can present as an acute abdominal disease, requiring urgent laparotomy with resection of the intestinal segment affected [16]. However, the clinical diagnosis of this event remains difficult and invariably requires specific imaging investigations in order to be able to treat the condition as soon as possible [17]. The clinical manifestations depend largely on the extent of the thrombus, the size of the vessel or vessels involves and the depth of bowel-wall ischemia. Abdominal pain, nausea, vomiting, bloody diarrhea and gastro-intestinal bleeding are most common symptoms at presentation [2,5,6,8,10]. The initial physical findings may be entirely normal. Fever, midabdominal pain, rebound tenderness develop later and indicate progression to bowel infarction and peritonitis [5].

The presence of increased D-dimer concentration levels and metabolic acidosis may serve to identify patients with suspected acute superior mesenteric occlusion [18]. In our case, blood chemistry revealed a leukocytosis with a modest increase of D-dimer levels but these may be also high in patients with inflammatory disease or in patients with intestinal occlusion.

Abdominal films are abnormal in 50 to 75% of patients but have findings specific for bowel ischemia in only 5% [5]. CFDs may demonstrate thrombus in the mesenteric veins [19], but CT is the test of choice for suspected cases of MVT [6,15,20,21]: an acute thrombus is evident as a central lucency in the mesenteric vein (Fig. 1). Other CT findings are enlargement of the SMV and a sharply defined vein wall with a rim of increased density. In our patient, only a CT abdominal scan has permitted the diagnosis of MVT, while all the other exams were not reliable.

In patients who are suspected of having MVT, CT is recommended. CT shows the mesenteric vessels and may define the extent of affected bowel, while it rules out other conditions that can cause abdominal pain.

Magnetic resonance imaging (MRI) has excellent sensitivity and specificity for the diagnosis of MVT [5,12,22,23], but its use is cumbersome and the equipment is not universally available. The symptoms are nonspecific and there is not any explicit radiological sign.

Once MVT has been confirmed, patient should be screened for hereditary or acquired thrombophilia. A bone marrow examination will be useful if a myeloproliferative disorder is suspected. In selected patients, once the acute symptoms have subsided, endoscopy and barium studies may be helpful to rule out the possibility of inflammatory bowel disease.

The treatment of MVT involves anticoagulant therapy alone or in combination with surgery.

The early initiation of anticoagulation using unfractionated heparin or low molecular weight heparin could minimise the serious complication such as peritonitis due to bowel necrosis requiring emergency surgery.

In our case, the delay in the diagnosis has not allowed to do a prompt anticoagulant and thrombolytic therapy even if their effectiveness has contrasting experiences [24—27]. As soon as the diagnosis of MVT is confirmed intraoperatively, immediate treatment with anticoagulants should be initiated. Subsequent management is dictated by the surgical findings, which range from a segmental infarction of small bowel to necrosis of entire bowel. On rare occasions, thrombectomy can be accomplished successfully when the thrombus is recent and is restricted to the SMV [5,28,29]. The optimal duration of anticoagulant therapy has not been defined; however, it is recommended that anticoagulation should be continued indefinitely, because it reduces the incidence of recurrence from 30—40% to 3—5% [24,25].

The addition of thrombolytic therapy to the treatment of MVT may enhance the clearance of thrombus and hasten the clinical improvements. Thrombolytic infusion may be done via an operatively placed mesenteric vein catheter [26,27]. MVT can safely be managed without surgery if there is no evidence of bowel infarction [7]. Immediate anticoagulation with heparin early in the course of the disease, even intraoperatively, clearly increases survival and significantly decreases the risk of recurrence [13]. Some authors have had occasional success using transhepatic portography to instill urokinase or tissue plasminogen activator directly into the thrombus in selected patients [30,31]. The use of thrombolytic agents is limited by the risk of hemorrhage and the...
low rate of success in cases in which the diagnosis has been delayed [29].

The mortality rate among patients with acute MVT ranges from 20 to 50% [14,15]. Survival depends on multiple factors, including age, the presence or absence of coexisting conditions and the timing of the diagnosis and surgical intervention.

Conclusion

SMVT is a rare but important clinical entity. The mortality rate is high because of delay in the diagnosis. A CT abdominal scan done early in the case of nonspecific abdominal pain, since the patients had a previous history of venous thrombosis, may not require a surgical treatment of MVT.

In patients with MVT, a screening of thrombophilia and of other coagulation disorders must always be carried out because they represent the most frequent cause of the venous thrombosis, in particular if there is lain thrombosis. Early diagnosis and prompt anticoagulation is the mainstay unless there are signs of peritonitis that necessitate surgical resection of the infarcted bowel tract [32].

Improvements in imaging techniques have led to early diagnosis, but there is often a considerable delay in the diagnosis because of a low degree of suspicion on the part of clinicians and the nonspecific clinical presentation. Early diagnosis and the immediate use of anticoagulation can improve the outcome. Surgery should be limited to patients with peritonitis or perforation. The objective of surgical management should be to conserve as much bowel as possible. In patients with inherited thrombotic disorders and those in whom a cause cannot be identified, lifelong anticoagulation is suggested.

References

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