CASE REPORT

Inflammatory Pseudotumor of the Mesentery Causing Portal Venous Thrombosis and Cavernomatous Transformation

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Abstract

A 32-year-old man was admitted to our hospital complaining of abdominal pain in the left upper quadrant. A mass was palpable on the left side of the umbilicus. Laboratory data revealed anemia, elevated erythrocyte sedimentation rate, hypergammaglobulinemia, and prolonged prothrombin time. Computed tomography demonstrated a soft tissue mass in the mesentery of the jejunum, portal venous thrombosis, and cavernomatous transformation in the porta hepatis. The patient was eventually diagnosed by laparoscopic partial resection as having inflammatory pseudotumor of the mesentery. Four months later, all of his symptoms and abnormal laboratory findings completely disappeared without any therapy. Inflammatory pseudotumor should be kept in mind as a cause of portal venous thrombosis, and/or cavernomatous transformation although it is rare. (Internal Medicine 41: 633-637, 2002)

Key words: erythrocyte sedimentation rate, hypergammaglobulinemia, laparoscopic partial resection, portal hypertension, hepatopetal collateral vessels

Introduction

Inflammatory pseudotumor can occur at various sites in the body but rarely occurs in the abdomen. The etiology of inflammatory pseudotumor is still unknown, although some believe that it may arise in association with trauma, surgery, or infection (1). A review of the literature indicates that most authors agree on the benign nature of the condition (2).

Portal venous thrombosis (PVT), first described by Balfour and Stewart in 1869 (3), is also a rare condition. It occurs in both children and adults, and is often associated with a wide variety of precipitating factors. The thrombus becomes organized, and tortuous collateral vessels gradually develop around the blocked portal vein, a process termed “cavernomatous transformation” (4). Although a variety of conditions may lead to PVT, there has been, to our knowledge, no report to date describing inflammatory pseudotumor as a cause of PVT and cavernomatous transformation of the portal vein.

Here, we report the extremely rare case of a patient presenting with a combination of inflammatory pseudotumor of the mesentery, PVT, and cavernomatous transformation of the portal vein.

Case Report

A 32-year-old man was admitted to our hospital complaining of abdominal pain in the left upper quadrant, appetite loss, and weight loss. A mass measuring 5 cm in diameter was palpable on the left side of the umbilicus without tenderness. Laboratory data included a white blood cell count of 7,100 cells/μl with 67.9% neutrophils, 24.5% lymphocytes, 6.0% monocytes, 1.2% eosinophils, and 0.4% basophils; a red blood cell count of 3,330,000 cells/μl; a hemoglobin of 8.9 g/dl; a hematocrit value of 28%; and a prolonged prothrombin time of 14.4 seconds. The patient’s erythrocyte sedimentation rate (ESR) was 96 mm/h, and his C-reactive protein (CRP) was 1.90 mg/dl.

Liver function tests showed an alanine aminotransferase (ALT) level of 61 units/l, and γ-glutamyl transpeptidase (γGTP) level of 244 units/l. Immunological studies revealed increased levels of IgA (421 mg/dl), IgG (2,124 mg/dl), and IgM (265 mg/dl) (reference values 110–410 mg/dl, 870–1,700 mg/dl and 33–190 mg/dl, respectively). Both hepatitis B surface antigen (HBsAg) and anti-hepatitis C virus antibody (HCVAb) were negative.

Abdominal computed tomography (CT) and ultrasonography (US) demonstrated dilatation of the intrahepatic portal vein and peripheral ring-like enhancement on transverse sections, leading us to suspect PVT. In addition, cavernomatous transformation was present in the porta hepatis (Fig. 1). The diagnosis of PVT associated with cavernomatous transformation was further confirmed both by angiography, which showed the...
absence of the trunk of the portal vein and the presence of collateral vessels around the portal vein (Fig. 2), and by US using the pulsed duplex Doppler technique. These examinations indicated no signs of cirrhosis of the liver or portal hypertension. Upper gastrointestinal endoscopy detected neither esophageal varices nor portal hypertensive gastropathy.

Fluoro-deoxyglucose positron emission tomography (FDG-PET) showed distinct uptake in the left mid-abdomen. Based on abdominal CT scans, it was considered to be a homogeneous and slightly enhancing soft tissue mass in the mesentery.
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of the jejunum (Fig. 3), and was also identified by US as a low echoic mass.

We performed a laparoscopic partial resection of the tumor for histopathological confirmation. The mass of the mesentery was partially resected with 21 cm of the jejunum. The tumor was a hard, well-demarcated and encapsulated white mass. Pathologically, foamy histiocytes, mild lymphoplasmatic infiltration, and myofibroblasts were present, though no neoplastic

Figure 4. Pathologically, foamy histiocytes, mild lymphoplasmatic infiltration, and myofibroblasts are present [HE stain, ×100 (left side) and ×400 (right side)].

Figure 5. The thrombus in the intrahepatic portal vein has disappeared, but the collateral vessels around the portal vein still are seen in the abdominal CT 1 year after admission.
cells were seen (Fig. 4). Immunohistochemical staining was positive with vimentin and focally positive with α-smooth muscle actin, but negative with desmin, S-100 protein, keratin, and CD34. Periodic acid-Schiff and Ziehl-Neelsen staining revealed no microorganisms. We thus diagnosed the patient with inflammatory pseudotumor of the mesentery associated with PVT and cavernomatous transformation of the portal vein.

Four months later, to our surprise, the mass of the mesentery, the intrahepatic portal venous thrombosis, and all of the abnormal laboratory findings had completely disappeared without any therapy, although the cavernomatous transformation of the portal vein still remained. Moreover, at the 1-year follow-up examination, abdominal CT demonstrated no changes in any of these findings (Fig. 5), and no symptoms at all were observed.

**Discussion**

Inflammatory pseudotumor is a distinctive pseudosarcomatous inflammatory lesion that occurs in the soft tissue and/or viscera of children and young adults. The original descriptions of these lesions focused on their occurrence in the lungs, and many lesions were generated for the pulmonary lesion, which is better known and possibly more common than its extrapulmonary counterpart. A number of synonyms of inflammatory pseudotumor, such as plasma cell granuloma, xanthofibroma, xanthogranuloma, and inflammatory myofibroblastic tumor, depending on the predominant cell type, have been used to describe the lesion. Its exact pathogenesis remains unclear. The clinical presentation and radiological features of inflammatory pseudotumor may mimic those of a malignant neoplasm, and the condition can be overtreated with radical surgery or chemotherapy, yet most of these tumors behave in a nonaggressive fashion (5). Therefore, histopathological confirmation is essential for the correct diagnosis. The clinical findings in the present case revealed anemia, a prolonged prothrombin time, an elevated erythrocyte sedimentation rate, and hypergammaglobulinemia, all of which are typical findings of inflammatory pseudotumor which have been previously reported (6, 7) together with pathological findings.

Interestingly, our case indicates that PVT associated with cavernomatous transformation could be caused by inflammatory pseudotumor of the mesentery, which had completely disappeared without any therapy 4 months after laparoscopic partial excision. In autopsy reviews, the overall incidence of portal vein occlusion ranges from 0.05% to 0.5% (8, 9). The most prevalent cause of PVT in adults is liver cirrhosis, which accounts for 24% to 32% of patients with PVT (8, 10). The second cause of PVT in adults is a variety of neoplastic diseases. In two studies, neoplasms were present in 21% and 24% of patients with PVT (10, 11). The PVT observed in these patients may occur as a consequence of direct invasion of the portal vein by the tumor, extrinsic compression of the portal vein, or periportal fibrosis following surgery or radiotherapy (10–14). A hyper-coagulable state secondary to malignancy additionally predisposes these patients to the development of PVT (9, 15). Infection is also a potential cause of PVT. One report demonstrates that approximately 10% to 25% of PVT cases arise due to sepsis in patients without cirrhosis or malignant diseases (4). Intra-abdominal inflammatory diseases may represent a small fraction of adults cases of PVT; in fact, pancreatitis, appendicitis, cholecystitis, alcoholic hepatitis, and perforated duodenal ulcer have been reported as causes of PVT (15–19). On the other hand, Webb and Sherlock do not mention noninfectious inflammatory causes of PVT in their review of 97 patients (20). Considering these previous reports, we propose that inflammatory pseudotumor of the mesentery should be included as a possible cause of PVT.

Nevertheless, the present case presents certain difficulties. The first is not only that the pathogenesis of inflammatory pseudotumor and intrahepatic portal venous thrombosis is unclear, but also why these findings disappeared without any therapy. Laboratory data indicated that the patient had started to improve since being admitted, and that his condition had normalized before laparoscopic partial resection. Therefore, the use of antibiotics after surgery can not reasonably be considered a possible explanation for the patient's remarkable recovery.

The second problem is the absence of portal hypertension. The thrombus may have arisen first within the superior mesenteric vein close to the inflammatory pseudotumor, and extended to the intrahepatic portal vein via the trunk of the portal vein. Simultaneously, hepatopetal collateral vessels may have grown and blood flow toward the liver may have been well compensated, so portal hypertension would not have appeared in this case.

In summary, we report here a case of inflammatory pseudotumor of the mesentery, which was associated with PVT and compensatory cavernomatous transformation of the portal vein. Although it may be extremely rare, inflammatory pseudotumor should be included in the differential diagnosis of diseases causing PVT.

**References**

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