Pylephlebitis and acute mesenteric ischemia in a young man with inherited thrombophilia and suspected foodborne illness

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We report on a young man who developed complicated pylephlebitis after foodborne illness. Despite antibiotics and resection of the focus of infectious colitis, he developed extensive small bowel infarction. He was treated with anticoagulation, local thrombolytic infusion, and resection of irreversibly ischemic small bowel. Thrombophilia workup demonstrated heterozygosity for factor V Leiden and the prothrombin G20210A mutation. The complications of pylephlebitis can be minimized by using systemic anticoagulation, thrombectomy, and/or local thrombolytic infusion along with antibiotics and surgical management of the infection. Evaluation for thrombophilic states should be considered, particularly if a patient does not respond to initial therapy. (J Vasc Surg 2012;55:1769-72.)

Pylephlebitis, or thrombophlebitis of the portal vein, is a potentially devastating complication of intra-abdominal infection. Antibiotics and resection of the focus of infection may not be sufficient to control thrombus progression, particularly in patients with thrombophilic disorders or late presentation.

CASE REPORT

An otherwise healthy 27-year-old man presented to an outside hospital with a 6-day history of gastrointestinal complaints after eating a spoiled casserole from a carry-out restaurant. His symptoms began with vomiting and watery diarrhea, followed by obstipation and right lower quadrant pain.

Computed tomography (CT) of the abdomen at the outside hospital was initially consistent with right-sided colitis. He was treated for presumptive infectious colitis with levofloxacin and metronidazole, but within 48 hours, his abdominal pain worsened and he developed pancreatitis (lipase >1000). Seventy-two hours after presentation, the final interpretation of his abdominal CT identified nonocclusive intrahepatic portal vein thrombosis and hypoperfusion of the right lobe of the liver. Repeat CT that day showed interval occlusive thrombus of the superior mesenteric vein (SMV) (Figs 1 and 2). He was then transferred to our center.

On arrival, he was hemodynamically stable with mild diffuse abdominal tenderness. A heparin infusion was started and antibiotics were continued (piperacillin/tazobactam and metronidazole). Colonoscopy that day demonstrated congested, ulcerated colonic mucosa and a 4-cm inflammatory mass in the proximal ascending colon (Fig 3). The patient was then taken to the operating room to resect the infectious focus that was believed to have precipitated the portomesenteric thrombus; a right hemicolectomy was performed. Anticoagulation was held briefly for surgery precipitated the portomesenteric thrombus; a right hemicolectomy was performed. Anticoagulation was held briefly for surgery and resumed 2 hours postoperatively. Blood and peritoneal fluid cultures were negative but Yersinia pseudotuberculosis colitis was clinically suspected based on history and tissue pathology.1 His stool tested negative for Clostridium difficile by enzyme-linked immunosorbent assay.

Ten hours postoperatively (hospital day 5), the patient developed frank peritonitis and shock (pH 7.23, base excess −8.4). The heparin infusion was held briefly for emergent re-exploration. The small bowel was deep purple 25 cm proximal to the ileocolonic anastomosis but potentially viable. The veins of the small bowel mesentery were engorged, and there was no antimesenteric Doppler signal. Our assessment was that the portomesenteric thrombus had propagated to the point of restricting small bowel perfusion. The patient went directly to interventional radiology for thrombolytic infusion and heparin was resumed 1 hour postoperatively.

Angiogram demonstrated an occluded SMV and slow flow in the superior mesenteric artery (SMA). Tissue plasminogen activator (TPA) infusion (1 mg/h) was started via a SMA catheter. Within 2 hours, however, his shock worsened, and dusky, edematous bowel was visible under the clear open abdominal dressing. A stat echocardiogram was consistent with acute pulmonary embolism (PE). The source of PE was suspected to be a femoral venous line, which was removed. He returned to the operating room emergently for peritonitis. A 112-cm segment of small bowel proximal to the ileocolonic anastomosis was grossly ischemic with no Doppler signal, this was resected. The heparin drip resumed 1 hour later, but TPA was discontinued. The next day, the patient underwent attempted placement of a thrombolytic catheter into the SMV by internal jugular approach, but the portal venous system could not be accessed. Heparin infusion was continued during all subsequent surgeries, which included a final resection of 10 cm of infarcted small bowel and eventual ileostomy.

An inherited thrombophilia was suspected in this patient, although he denied a personal or family history of venous thromboembolism (Table). He was found to be heterozygous for the factor V Leiden and prothrombin II mutations. Functional assays...
of antithrombin III and proteins S and C demonstrated decreased activity, most likely due to factor consumption. Resistance to heparin was not suspected because thromboelastography showed appropriate correction with heparinase. He is a carrier of the methylenetetrahydrofolate reductase C677T polymorphism, but homocysteine level was confirmed to be normal.

The patient was eventually transitioned to anticoagulation with warfarin, which will continue indefinitely. Nine months after his initial illness, his portal vein was radiographically patent, and his ostomy was reversed. He is now doing well.

DISCUSSION

Pylephlebitis should be treated aggressively and comprehensively with the objective of avoiding visceral ischemia, liver abscess, and chronic portal hypertension. Antibiotics are essential because uncontrolled infection, not visceral ischemia, is the primary threat to the patient’s life. Blood cultures should be obtained and broad-spectrum antibiotics started for any patient with acute portal vein thrombosis (PVT) and fever, regardless of whether an intra-abdominal infection has been identified. Cultures are frequently polymicrobial, but *Bacteroides fragilis* is most common and has unique prothrombotic effects. Antibiotics should continue for at least 4 weeks to cover early radiographically occult hepatic abscesses. Surgical resection of the primary infection should be considered in patients who are stable enough for surgery. Advances in genetic testing have demonstrated that noncirrhotic, nonmalignant portal vein thrombosis is frequently the result of an inherited thrombophilia superimposed on local risk factors such as infection, inflammatory bowel disease, or trauma. Among 100 case reports of pylephlebitis since 1971, approximately one-third of the patients who were screened for genetic thrombophilia were found to have an abnormality. Another series of 92 patients with acute PVT demonstrated a relative risk of 2.7 in patients with factor V Leiden mutation, 1.4 for prothrombin 20210 G/A mutation, and 4.6 for protein C deficiency. The prothrombin 20210 G/A mutation may account for many PVT cases that were previously considered idiopathic.
Table. Thrombophilia screening in our patient

<table>
<thead>
<tr>
<th>Laboratory test</th>
<th>Result</th>
<th>Interpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Factor V Leiden mutation</td>
<td>Genotype G/A</td>
<td>Heterozygous abnormal</td>
</tr>
<tr>
<td>Prothrombin II G20210A mutation</td>
<td>Genotype G/A</td>
<td>Heterozygous abnormal</td>
</tr>
<tr>
<td>Antithrombin III functional assay</td>
<td>36% activity</td>
<td>Low activity due to recent thrombosis and heparin infusion</td>
</tr>
<tr>
<td>Protein C functional assay</td>
<td>50% activity</td>
<td></td>
</tr>
<tr>
<td>Protein S functional assay</td>
<td>30% activity</td>
<td></td>
</tr>
<tr>
<td>Anticardiolipin antibody IgG</td>
<td>7 GPL units/mL</td>
<td>Normal (reference range, 0-23)</td>
</tr>
<tr>
<td>Anticardiolipin antibody IgM</td>
<td>2 MPL units/mL</td>
<td>Normal (reference range, 0-11)</td>
</tr>
<tr>
<td>Homocysteine</td>
<td>3 μmol/L</td>
<td>Normal (reference range, 5-15)</td>
</tr>
<tr>
<td>Methylene tetrahydrofolate reductase (MTHFR) C677T mutation</td>
<td>Genotype C/T</td>
<td>Heterozygous carrier</td>
</tr>
<tr>
<td>Methylene tetrahydrofolate reductase (MTHFR) A1298C mutation</td>
<td>Genotype A/A</td>
<td>Homozygous normal</td>
</tr>
<tr>
<td>JAK2 V617F mutation</td>
<td>Negative</td>
<td>Homozygous normal</td>
</tr>
</tbody>
</table>

CONCLUSIONS

This case demonstrates the importance of early treatment for pylephlebitis, to treat the hematologic and infectious pathology. Anticoagulation should be considered for all patients to facilitate recanalization of the portal vein, even if bowel ischemia is not suspected. Earlier and more aggressive anticoagulation may have avoided bowel infarction in this patient. Clinicians should also consider the presence of a thrombophilic disorder in a patient with atypical presentation or inadequate response to treatment.

REFERENCES


Submitted Oct 13, 2011; accepted Dec 27, 2011.