SHORT REPORT

Inflammatory pseudotumor of the liver associated with Crohn's disease

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Abstract

Inflammatory liver pseudotumor is a rare entity. Associations with several inflammatory conditions were reported but association with inflammatory bowel disease is unusual. We report the case of liver inflammatory pseudotumor occurring in the course of Crohn's disease in a 23-year-old woman and treated conservatively.

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1. Introduction

Inflammatory pseudo-tumor (IPT) of the liver is a rare benign disease with unclear etiology and pathogenesis.1,2 Because of its unusual clinical and radiological presentation a presumptive diagnosis of malignancy is frequently made and hepatic resection is usually performed.2,3 However, inflammatory pseudotumor of the liver is benign and may regress spontaneously or with medical treatment. Diagnosis must be established to avoid radical surgical procedure in some cases4. Association with Crohn's disease was reported only in three cases.5,6 We report a patient with Crohn's disease associated with a liver IPT.

2. Case

A 23-year-old woman presented with a one-month history of glairous and bloody diarrhea, and diffuse abdominal pain. Her temperature was 38° and the pulse rate was 92 bpm. On physical examination, she had mild upper abdominal tenderness. Laboratory tests revealed hemoglobin levels of 7.4 g/dl, white blood cell count 12,000/mm³ and albumin of 26 g/l. The erythrocyte sedimentation rate was 80 mm/h, and serum C-reactive protein levels were 167.1 mg/dl. Liver function tests were normal. Colonoscopy revealed severe colitis characterized by large linear and "well-like" ulcers and mucosal disbanding with skip areas. Lesions were more prominent in the sigmoid colon. Stool parasitology and culture were negative. Colonic biopsy did not show cytomegalovirus inclusions or bacteria. The diagnosis of severe Crohn's colitis was made. However, an ultrasonography revealed a hypoechoic heterogenous subcapsular space occupying lesion in the left lobe of the liver (segments 3 and 4), measuring 2 cm. A CT scan showed a hypodense and ill-defined liver nodule with a poor and delayed contrast enhancement (Figs. 1–3). Tumor markers, namely AFP, CEA, CA125, and CA19-9, were all negative.

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Hepatitis B surface antigen and antibodies to hepatitis C virus were negative. In the context of idiopathic inflammatory bowel disease, the diagnosis of IPT was suggested and a conservative approach was opted. The patient received initially corticosteroids and antibiotics, which resulted in resolution of clinical symptoms, apyrexia, and normalization of the serum CRP levels, followed by azathioprine maintenance therapy. Repeat ultrasonography and CT scans showed a complete regression of the tumor six months later.

3. Discussion

Inflammatory pseudo-tumor (IPT) is a rare benign disease characterized histopathologically by proliferating fibrous tissue infiltrated by inflammatory cells. IPT have been reported in the orbit, lung, intestine, pancreas, heart, brain, ovary and retro peritoneum. The liver is a rare site. To date, there have been fewer than 80 cases reported in the literature. The pathogenesis of liver IPT remains unclear. The inflammatory cells noted in IPT suggest an immune reaction of the host to bacterial infection resulting in occlusive phlebitis of intrahepatic veins. Many patients report a history of recent infection. IPT is also associated with chronic inflammatory condition, such as diverticular stenosis. IPTs occur predominantly in males. The age at diagnosis may range from 9 months to 83 years. Clinically, most patients present with nonspecific symptoms including fever, abdominal pain and weight loss. Less commonly, patients experience malaise, nausea, and vomiting. Only few IPTs are found incidentally at CT scan or laparoscopy. An inflammatory syndrome is frequently reported (60%). Liver function tests are sometimes abnormal (40%). Only three cases of IPT associated with Crohn’s disease have been reported (Table 1). In our case symptoms were dominated by glairy and bloody diarrhea which led to exploring the colon first. Evolution was marked by amelioration under corticosteroid therapy. Endoscopic ultrasonography may discover incidentally IPTs.

Radiologic findings have been reported to be nonspecific. Imaging techniques disclose a solid focal liver tumor most often located in the right liver lobe (53%). Ultrasonography shows frequently a hypoechoic nodule as well. CT scan shows a hypo- or iso-dense nodule to the remains in the liver parenchyma. Enhancement of inflammatory pseudo-tumor on delayed-phase CT scans has been frequently observed. It may be explained by the fact that IPTs may consist of various amounts of fibrous tissue along with cellular infiltrates. In our case, the tumor was shown as a solitary hypoechoic band in US. In CT scan, it appeared like a hypodense nodule occupying the left lobe of the liver with poor enhancement in the portal time. In the two cases reported by Georgios, there were a hilar liver mass with common bile duct stricture and dilatation of intrahepatic ducts. In the case of Amankoh, an inhomogeneous mass in the left hepatic lobe was shown. MRI features of IPT are rare and
which exhibits foamy histiocytes, plasma cells, and variable features of the lesion are characterized by connective tissue cellular carcinoma is different from that of IPTs. Histologic peripheral cholangiocarcinoma with abundant fibrosis can show typically show a central water density. Hepatic metastasis and abscess may be mistaken as IPTs because symptoms and laboratory data can be similar. However, liver abscess at CT scan may be mistaken as IPTs because symptoms and laboratory data can be similar. However, liver abscess at CT scan typically show a central water density. Hepatic metastasis and peripheral cholangiocarcinoma with abundant fibrosis can show delayed enhancement. The enhancement pattern of hepatocellular carcinoma is different from that of IPTs. Histologic features of the lesion are characterized by connective tissue which exhibits foamy histiocytes, plasma cells, and variable degrees of neutrophil and lymphocyte infiltration.4

4. Conclusion

Inflammatory pseudo-tumor of the liver associated with Crohn’s disease is rare. Our case is the fourth reported in the literature. Normal serum tumor marker levels and imaging techniques lacking the typical features of liver abscesses and enhancement in delayed-phase CT scan in this case of Crohn’s disease led to the diagnosis. It is likely that systemic inflammation due to Crohn’s disease is causally related to the pathogenesis of IPT. Therefore, anti-inflammatory treatment with corticosteroids and immunosuppressives resulted not only in resolution of symptoms of Crohn’s disease but also of IPT. Biopsy and surgical treatment would be considered in cases resistant to treatment with anti-inflammatory drugs. Although rare, a diagnosis of inflammatory bowel disease should be considered in patients with inflammatory pseudo-tumor of the liver and vice-versa.

Table 1 Reported cases of IPT in patients with Crohn’s disease.

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Activity and location of Crohn’s disease</th>
<th>Presentation of IPT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amankonah6</td>
<td>37-years-old Man</td>
<td>Ileum treated with mesalamine therapy Diagnosis established 2 weeks after IPT</td>
<td>Night sweats Abdominal pain Weight loss</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Moderately treated with mesalamine therapy Diagnosis established 2 weeks after IPT</td>
<td>Intermittent fevers Night sweats Abdominal pain Weight loss</td>
</tr>
<tr>
<td>Georgios5</td>
<td>68-years-old Man</td>
<td>Colon Severe treated with corticotherapy and mesalamine Therapy Diagnosis established after IPT</td>
<td>Vague abdominal pain Weight loss Jaundice</td>
</tr>
<tr>
<td>Georgios5</td>
<td>67-years-old Man</td>
<td>Stomach–duodenal–ileum Severe Treated with infliximab and azathioprine Diagnosis established 1 years after IPT</td>
<td>Intermittent abdominal pain Jaundice</td>
</tr>
<tr>
<td>Case reported</td>
<td>23-years-old Women</td>
<td>Colon Severe treated with corticotherapy and azathioprine concomitant Diagnosis established after IPT</td>
<td>Asymptomatic (found incidentally)</td>
</tr>
</tbody>
</table>

References