Inflammatory pseudotumor of the liver

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Abstract

Inflammatory pseudotumor of the liver (IPL) is a rare benign neoplasm. It often masquerades as a malignancy, either primary or metastatic. We describe the case of a 71-year-old man who presented with fever and malaise. Workup revealed a cecal mass, as well as a lesion in the left lobe of the liver. Pathologic examination showed a cecal neoplasm and a hepatic inflammatory pseudotumor. The patient recovered uneventfully. This case highlights the ability of IPL to masquerade as a malignant hepatic neoplasm, and emphasizes the difficulties in diagnosis.

Key words Inflammatory · Pseudotumor · Tumor · Liver · Neoplasm

Introduction

Inflammatory pseudotumors are rare, benign tumors most commonly found in the lung,1 but also described in the central nervous system,2 salivary glands,3 larynx,4 bladder,5 breast,6 pancreas,7 spleen,8 lymph nodes,9 skin,10 and liver.11 It is perhaps the hepatic inflammatory pseudotumor that is most clinically relevant, since it can masquerade as a malignant liver neoplasm.

Case Report

A 71-year-old man was admitted from an outside hospital to the transplant surgery service for treatment of a presumed liver abscess and pulmonary embolism. On admission the patient was febrile, with blood cultures positive for Streptococcus viridians. Ultrasound examination demonstrated a large intrahepatic mass, confined to the left lobe, 13 × 14 × 8.7 cm in size. A computed tomography (CT) scan showed a liver mass (Figs. 1 and 2) and a possible cecal lesion. The liver biopsy specimen was nondiagnostic. Positron emission tomography (PET) imaging demonstrated intense uptake in the liver and cecum, raising the suspicion of a primary colon malignancy with hepatic metastases. A subsequent colonoscopy found a single, large, fungating, irregular, friable mass in the cecum, consistent with colon cancer. An alpha-fetoprotein level of 2 ng/ml (normal range, 0–10 ng/ml) argued against a liver primary tumor, and a carcinoembryonic antigen (CEA) level of 4.9 ng/ml (normal range, 0–3 ng/ml for nonsmokers) argued for colon cancer metastasis. After having discussed the case and obtaining informed consent, the patient underwent a right colon resection with primary anastomosis and a left lateral segmentectomy of the liver. Pathologic examination diagnosed an infiltrating moderately differentiated adenocarcinoma (T2N0Mx) of the cecum and an inflammatory pseudotumor of the liver (Fig. 3).

Discussion

Inflammatory pseudotumors of the liver (IPLs) are also referred to as inflammatory myofibroblastic tumors. They are three times more common in men than in women. They are more frequently seen in the nonEuropean population, and have an average age at presentation of 35 years.12 Sixty-one percent of the lesions are located in the right hepatic lobe, near the gallbladder or related to the biliary tree.13 IPLs typically present with vague constitutional symptoms such as fever (66%), abdominal pain (51%), and weight loss (21%).14 This vague and nonspecific symptomatology makes preoperative diagnosis difficult. Imaging studies often show large tumors that are diffuse, infiltrative, and associated with portal vein occlusion. All of these findings are also encountered in malignant neoplasms.
Upon gross examination at the time of laparotomy, IPLs are sharply circumscribed. They are typically adherent to surrounding organs, have a whorled cut surface, and occasionally have areas of calcification or focal hemorrhage.\textsuperscript{15} Histologically, IPLs are composed of densely hyalinized collagenous tissue infiltrated predominately by plasma cells and plump spindle cells, with occasional lymphocytes and histiocytes.\textsuperscript{16} In addition, medium- and large-sized veins demonstrate inflammation involving the vessel wall.\textsuperscript{12}

The pathogenesis of IPLs remains largely unknown. Many theories exist, however, including intraparenchymal hemorrhage and necrosis, infection, occlusive phlebitis of intrahepatic veins, immune reaction, or secondary reaction to an intrahepatic rupture of a biliary radicle.\textsuperscript{17} Perhaps the most popular theory suggests an infectious source as a cause. Some have hypothesized that microorganisms from certain conditions, such as appendicitis, seed the hepatic parenchyma through the bloodstream of the portal vein, creating an inflammatory reaction with obliterating phlebitis and granulomata formation.\textsuperscript{18}

Despite their rarity, IPLs remain clinically important because of their differential diagnosis with both benign and malignant hepatic masses. Clinical, radiologic, and histologic data can help distinguish between IPLs and other liver diseases. Specifically, confirmation of the diagnosis can sometimes be made with core biopsy. Fine needle aspiration should be avoided, as the procedure leads to a high incidence of misdiagnoses.\textsuperscript{12} Hepatocellular carcinomas (HCCs) are frequently associated with cirrhosis and viral infections, and have elevated alpha-fetoprotein levels. Both HCC and IPL can cause portal vein occlusion. However, while HCCs create occlusion with intraluminal tumor invasion, IPLs, on the other hand, cause occlusion by creating inflammation and thickening of the wall of the vein.\textsuperscript{12} Histology is important in distinguishing IPL from other spindle-cell tumors. Both angiosarcomas and leiomyosarcomas, when compared to IPLs, have fewer inflammatory cells and higher amounts of cellular atypia and frequent mitoses.\textsuperscript{16} IPLs have an excellent prognosis.\textsuperscript{12,14}