

Inflammatory pseudotumor of the liver: A case report and literature review

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Summary

Reported here is the case of a 55-year-old man who had tarry stools for 3 days before he was seen at this Department. The man had weight loss and an intermittent fever for 3 months prior. Histopathology revealed an inflammatory pseudotumor of the liver. This case is reported here along with a review of the literature. Nine days after surgery, the patient passed bright red blood (150 mL) in the stool with no clear trigger. A colonoscopy a month later revealed no abnormalities. This is a rare report of an inflammatory pseudotumor featuring intractable bleeding. An inflammatory pseudotumor of the liver is a rare condition, and differentiating this pseudotumor from hepatic space-occupying lesions is crucial. An inflammatory pseudotumor of the liver may spontaneously regress and mimic other liver tumors. The treatment of choice for this pseudotumor is still surgical resection, and this is especially true for patients with severe symptoms or an indeterminate diagnosis.

Keywords: Inflammatory pseudotumor (IPT), liver, bleeding

1. Introduction

An inflammatory pseudotumor (IPT) is a rare condition that was first described in the lung in 1939 (1). IPT most commonly occurs in the lung, but it can be found in other locations including the central nervous system, major salivary glands, the kidneys, the liver, the omentum, the ovaries, the larynx, the urinary bladder, the breasts, the pancreas, the spleen, lymph nodes, skin, soft tissues, and the orbit of the eye (2). An IPT of the liver (IPTL) is a rare benign lesion characterized by chronic infiltration of inflammatory cells and an area of fibrosis that sometimes mimics a malignant tumor (3,4). An IPTL was first reported by Pack and Baker in 1953 (5).

The etiology and pathogenesis of IPTL remain unknown. Biologically, there are no specific symptoms or laboratory or radiologic findings that are useful at diagnosing IPTL. Differentiating between IPTs and

other focal hepatic lesions remains a major problem. The treatment of choice is still surgical resection, and this is especially true for patients with severe symptoms or an indeterminate diagnosis (6,7).

2. Case Report

This case involved a 55-year-old man who had an unremarkable medical history and tarry stools for 3 days before he was seen at this Department. The man also had weight loss (15 kg) and an intermittent fever for 3 months prior. He had no abdominal pain or night sweats and he presented without jaundice. On admission, a physical examination revealed no signs ("stigmata") of chronic liver disease or hepatomegaly. A previous computed tomography (CT) scan showed a well-defined heterogeneous mass 4.0 cm × 4.0 cm in size situated in the left hepatic lobe (Figure 1). On CT, the lesion featured central necrosis, a hyper-dense rim, and mild enhancement starting in the arterial phase, thus corresponding to a hepatic abscess.

Laboratory results revealed an aspartate aminotransferase level of 122 IU/L (normal, 5-40 IU/L), an alanine aminotransferase level of 89 IU/L (normal, 8-40 IU/L), an alkaline phosphatase (ALP) level of 162 IU/L (normal, 40-140 IU/L), a gamma-glutamyl transpeptidase (GGT) level of 159 U/L (normal, 7-40 U/L), an erythrocyte sedimentation rate (ESR) of 45

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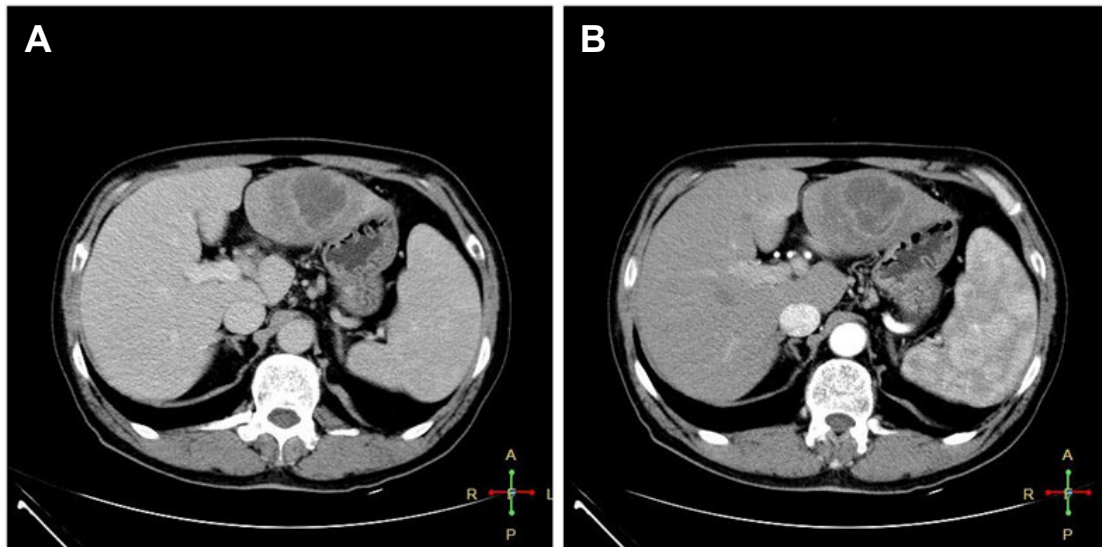


Figure 1. Computed tomography images of the abdomen. (A) A well-defined heterogeneous mass 4.0 cm × 4.0 cm in size was situated in the left hepatic lobe. **(B)** The lesion featured central necrosis, a hyper-dense rim, and mild enhancement starting in the arterial phase.

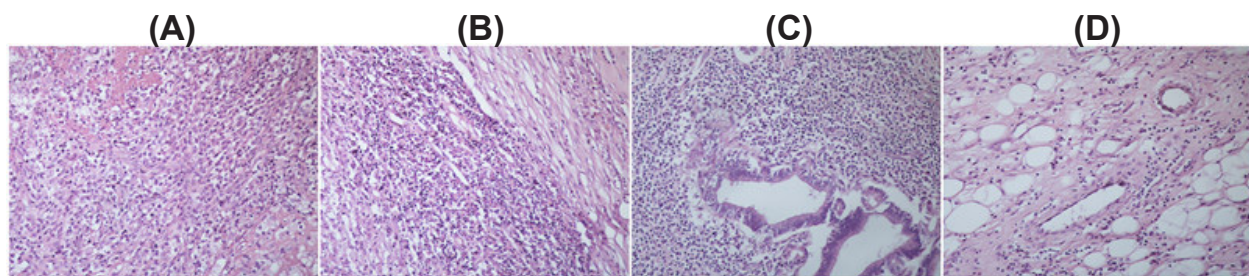


Figure 2. Histology of a surgical specimen. (A) Numerous lymphocytes and plasma cells are evident. **(B)** Cells mainly consist of inflammatory cells and eosinophils. **(C)** Shown are lymphocytes and macrophages. **(D)** Shown are inflammatory and xanthomatous changes. (hematoxylin and eosin, × 200).

mm/h (normal, < 7 mm/h), a CEA level of 1.34 ng/mL (normal, < 3.4 ng/mL), AFP level of 3.41 ng/mL (normal, < 7.02 ng/mL), and a CA 19-9 level of 6.64 U/mL (normal, < 39 U/mL). In addition, the patient had negative serologic results for hepatotropic viruses, cytomegalovirus, Epstein-Barr virus (EBV), and HIV 1 and 2. Standardized immunohistochemistry (Xijing Hospital, Fourth Military Medical University, Xi'an, China) showed monoclonal antibodies against broad-spectrum keratin AE/AE3(+), CD138(+), CK19(+), LCA(+), and Hep(-) (Dako, Glostrup, Denmark). Both antibodies reacted positively to cells from the lesion. An ultrasound-guided percutaneous liver biopsy with a trucut needle was performed and revealed hepatic tissue with proliferation of spindle-shaped cells mixed with an inflammatory infiltrate of histiocytes, suggesting a diagnosis of IPTL.

After surgery, a specimen of the liver parenchyma was examined histopathologically. The tumor had been completely removed. Microscopy revealed a host of benign cells, including numerous inflammatory cells, mature plasma cells, lymphocytes, eosinophils, and macrophages; most of these cells had xanthomatous

changes (Figure 2). The patient received no further treatment. Nine days after surgery, the patient passed bright red blood (150 mL) in the stool with no clear trigger. A colonoscopy a month later revealed no abnormalities. Two months later, the patient's condition was still satisfactory.

3. Discussion

IPT is also known as an inflammatory myofibroblastic tumor or plasma cell granuloma, a xanthomatous pseudotumor, and inflammatory fibrosarcoma (8). Someren classified IPTs into 3 groups according to histology: xanthogranuloma-type pseudotumors, plasma cell granuloma-type pseudotumors, and sclerosing pseudotumors (9). Macroscopically, the lesion may mimic a malignancy and may be alone or several lesions may be present. The lesion may be as large as 25 cm. Microscopically, IPT is characterized by spindle-shaped cells, myofibroblasts, and mixed inflammatory cells (plasma cells, lymphocytes, and sporadic histiocytes). IPTL most often occurs in childhood and early adulthood (10). In adults, the ratio of males to females

affected ranges from 1:1 to 3.5:1 (11). IPTL appear to be more common in non-European populations (12). These IPTs are most likely inflammatory or infectious in origin. The lesions often appear to develop from a healing abscess or an inflammatory condition (11). The most common symptoms of IPTL are abdominal pain, a fever, and weight loss. IPTL frequently resolves spontaneously with a good prognosis (13).

IPTL is quite rare and accounts for 8% of extrapulmonary IPTs. The incidence of IPTL is reported to be around 0.7% according to recent studies (14,15). The etiology and pathogenesis of IPTL remain unknown since a variety of tumorous inflammatory lesions lack the features of other IPTs, but IPTL is thought to involve an inflammatory reaction (16). Infectious agents that have been hypothesized to cause IPTL include infections, trauma, vascular causes, and autoimmune disorders (17). Numerous studies have noted that the microorganisms responsible for IPTL include *Bacteroides caccae*, *Actinomyces*, *Klebsiella*, *Escherichia coli*, Gram-positive cocci, and β -hemolytic *Streptococcus* (9). Other studies reported that hepatopancreatobiliary autoimmune diseases, such as IgG4 sclerosing cholangitis, could also cause IPTL (18).

The diagnosis of liver pseudotumors is obviously difficult. Ultrasound and CT scans are not specific, revealing variable patterns of echogenicity or a liver mass mimicking hepatocellular cancer or an abscess (19,20). A CT scan usually reveals lesions with variable contrast enhancement. IPTs may display a hypovascular pattern because of fibrosis and also display delayed enhancement, similar to metastatic liver tumors and cholangio-carcinomas (21). MRI may produce low signal intensity (hypointensity) on T1-weighted images with moderate to high signal intensity (hyperintensity) on a T2 sequence (16,21). In general, differentiating IPTs from malignant tumors with radiographic studies is difficult. A definitive diagnosis of IPT can be made based on needle biopsy findings and, occasionally, in-needle aspiration, as long as the pathologist is aware of this possibility.

Biologically, there are no specific symptoms or laboratory or radiologic findings that are useful at diagnosing IPTL. Despite recent increases in the diagnostic capability of radiologic studies, differentiating IPTL from other focal hepatic lesions remains a major problem. Unfortunately, clinical and radiologic features of IPTL can mimic other liver tumors like lymphoma, malignant fibrous histiocytoma, hepatocellular carcinoma, metastatic tumor, tuberculosis, and sarcoidosis and thus lead to surgery (16). If an atypical solid mass is found in the liver, IPTL should be considered as a potential diagnosis, particularly if the mass is accompanied by clinical evidence of an inflammatory process: a recent history of asthenia, malaise, vague upper abdominal discomfort, and/or an intermittent fever; the presence of stigmata of chronic

liver disease or splenomegaly; abnormal liver function test results; and a lack of specific imaging findings.

Although liver biopsy indisputably has a role in the investigation and management of liver metastases of unknown origin, its role is more contentious and possibly dangerous in cases of a solitary hepatic mass that is likely to be malignant (22). The main histopathological findings in all cases are the presence of myofibroblastic spindle cells, plasma cells, macrophages, and lymphocytes without cellular atypia or atypical mitotic figures (23). A biopsy of the tumor is not necessary when planning a surgical intervention for the liver.

Furthermore, an optimal treatment for IPTL and a method of determining its prognosis have yet to be established (24,25). Due to its diagnostic ambiguity, the lesion completely resolved in some patients that received antibiotics and/or corticosteroids, but some of these lesions recurred (25). In contrast, numerous studies have reported performing a hepatic resection, mainly due to evidence that the tumor is malignant according to preoperative radiography, after which IPTL never recurred (26).

Even though IPTL may spontaneously regress or regress following antibiotic treatment, the treatment of choice is still surgical resection, and this is especially true for patients with severe symptoms or an indeterminate diagnosis (6,27). Hepatectomy has become a safer option for non-cirrhotic patients over the past 20 years, with mortality converging to 0%. Therefore, the treatment of choice should be surgical resection in such cases (28). This approach is preferable because it minimizes the risk of a biopsy-related complication (dissemination in cases of malignancy) and because it eliminates the possibility of IPT recurring.

In conclusion, IPTL is a rare condition, and differentiating this pseudotumor from hepatic space-occupying lesions is crucial. IPTL may regress spontaneously and it may mimic other liver tumors. The treatment of choice is still surgical resection, and this is especially true for patients with severe symptoms or an indeterminate diagnosis.

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