Inflammatory Pseudotumor of the Liver: An Unlikely Cause of Multiple Hepatic Lesions

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Inflammatory pseudotumor of the liver, also called inflammatory myofibroblastic tumor or plasma cell granuloma, was first described in 1953 by Pack and Baker [1]. It is generally regarded as a benign, reactive inflammatory condition although some cases of recurrence and/or metastases have been reported [2]. About 50 cases have been described in the literature worldwide, mostly in young males. The presenting symptoms are abdominal pain, vomiting, diarrhea, jaundice and intermittent fever. Leukocytosis, hyperglobulinemia and elevated erythrocyte sedimentation rate have been reported in about 50% of the patients [3]. Other laboratory manifestations include elevated C-reactive protein levels and slightly deranged liver function tests [1].

Macroscopically, the lesion resembles a tumor, single or multiple, measuring up to 25 cm. Microscopically, the lesion is characterized by spindle-shaped cells proliferation, and myofibroblasts mixed with inflammatory cells composed of plasma cells, lymphocytes and, sporadically, histiocytes [4]. Sclerosis with a whorled appearance has also been noted. The lesion usually does not increase in size or may even undergo spontaneous regression. However, it may cause symptomatic biliary obstruction or portal hypertension according to its anatomic position, and then it should be removed whenever possible [3].

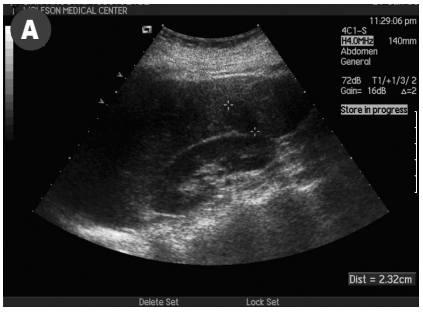
Patient Description

A 79 year old woman was admitted with right upper quadrant abdominal pain. In addition she complained of weakness, anorexia and weight loss of 10% during the previous 6 months. Her medical history revealed ischemic heart disease, two events of myocardial infarction, and risk factors such as smoking and hypercholesterolemia. She had undergone coronary artery bypass graft and percutaneous transluminal coronary angioplasty.

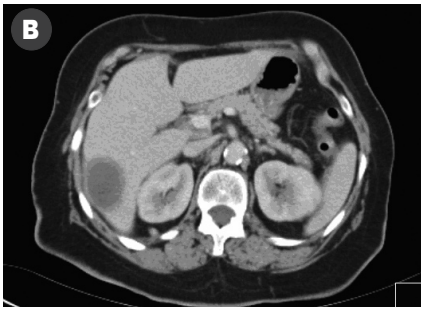
Physical examination on admission demonstrated diffuse abdominal tenderness, mainly on the right upper quadrant, without signs of peritonitis. The liver was not palpable. Laboratory tests revealed leukocytosis (white blood cells 15,400), deranged hepatic function tests (alanine transaminase 73 U/L, aspartate transaminase 79 U/L, alkaline phosphatase 124 U/L, total bilirubin 0.91 mg/dl), hyperglobulinemia (globulin 4.4 g/dl) and hypoalbuminemia (albumin 3.1 g/dl, albumin-globulin ratio 0.7). During her hospital stay she underwent other clinical tests: blood cultures, parasitology for Echinococcus, stool culture for E*ntamoeba histoliytica* and the serum tumoral marker levels (alpha-fetoprotein, carcinoembryonic antigen, and CA 19.9, CA 15.3, CA 125), which proved to be negative.

An ultrasound examination of the abdomen revealed the presence of two hypodense lesions of about 2.1 cm and 3.0 cm in diameter in the right lobe of the liver, without an increase in hepatic volume [Figure A]. An abdominal computed tomography scan confirmed the presence of two masses, with diameters of 4.8 x 3.6 cm and 2.0 x 2.3 cm in the right lobe of the liver. The smaller lesion showed contrast enhancement. No other pathological findings were reported [Figure B].

At this stage fine needle aspiration from the liver mass was performed. A purulent exudate with no bacterial growth was obtained from the large hepatic lesion and a core liver biopsy was obtained from the small lesion. The sample, 1.5 cm long, 5 µm tissue sections, fixed in 4% buffered formaldehyde solution, was paraffin embedded and the sections were stained with hematoxylin & eosin. Until the specific culture results arrived broad-spectrum antibiotic therapy was administered. A repeat ultrasound revealed enlargement of the purulent lesion, which had expanded and comprised three septate masses, 5.5 x 7.5 cm in size. A



[A] Ultrasonography of the right lobe of the liver, demonstrating the small mass, around 2 cm in diameter, well defined, hypoechoic and hypovascular.



[B] CT scan showing a round, low density lesion with regular borders in the right lobe of the liver, size 4.8 x 3.6 cm.

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repeat CT scan showed the hypodense lesion, 4 x 3.8 x 6.5 cm in size, with a rim contrast enhancement, surrounded by edema. Under CT guidance, drainage of the main lesion was performed, yielding a purulent material – again with no bacterial growth. This time a drain was left in the lesion.

Histopathological examination of the liver core sample revealed a lesion composed of cellular spindle cell tissue with tendency to whorled arrangement, several dense collagenous bundles, and moderate inflammatory cell infiltrate rich in plasma cells. Foamy macrophages were also noted focally. The lesion cells demonstrated a bland appearance, and were positive for CD68 (monocyte differentiation), focally positive for actin (cytoskeleton) and negative for CK-MNF-116 (cytokeratin), CD34 (pluripotent stem cell), S-100 (undifferentiated malignancies and neuroectodermal tumors), and P53 (cell cycle-regulating protein) immunostains. KI-67 (cell proliferation marker) was positive in less than 5% of the lesion cells. These findings are consistent with the diagnosis of pseudotumor of the liver.

Following enteral broad-spectrum antibiotic treatment and drainage, the patient's condition improved and a significant regression in the size of the lesions was noted on follow-up CT scan. The patient was discharged without clinical symptoms 2 weeks after admission.

Comment

Inflammatory pseudotumor of the liver is a rare disease. Its etiology is unknown, although several theories have been raised, including bacterial spread from food, chronic appendicitis, cholecystitis or Epstein-Barr virus infection [5].

Accurate diagnosis is important prior to surgery. The differential diagnosis includes granulomatous lesions (sarcoidosis and tuberculosis), and malignant lesions (lymphoma, malignant fibrous histiocytoma, hepatocellular carcinoma or metastatic spread) [3]. The inflammatory pseudotumor might be clinically asymptomatic, though at times it might present with alarming characteristics, imitating a malignant neoplasm. Imaging studies supporting the latter diagnosis might direct the course of treatment towards a markedly vigorous one [5].

A consensus regarding the optimal management of this lesion has not yet been reached. Percutaneous biopsy, whenever feasible, should be performed to obtain histological confirmation and verify the diagnosis. Patients are usually treated surgically with hepatic resection due to biliary or other obstructive symptoms, although there have been a few reports of pseudotumors regressing spontaneously [5].

We report our experience in the management of a woman with two inflammatory pseudotumours. Drainage and enteral antibiotics were administered. The smaller pseudotumor regressed spontaneously, and the larger lesion decreased following drainage. Therefore, we suggest considering a conservative management for these patients prior to an attempt at hepatic resection.

References

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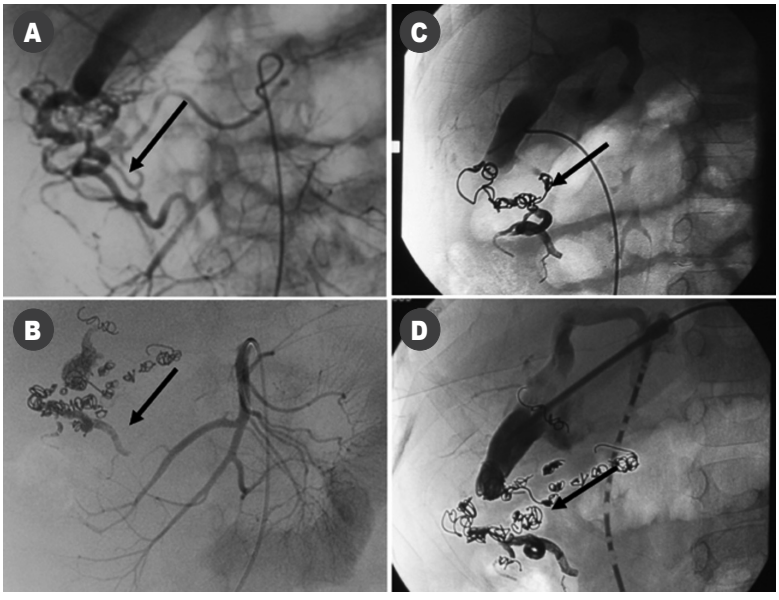
Erratum

In the case report "Arteriovenous fistula and portal hypertension in a child with Down syndrome" by Landau et al. (November issue: 2007;9:825–6), the figures were incorrectly marked. Figure B should be C, and Figure C should be B. The correctly marked figures appear below.



Transfemoral arteriography demonstrating the arteriovenous fistula (arrow) between the superior mesenteric artery – through the pancreaticoduodenal vessels – and the portal vein, before **[A]** and after **[B]** the procedures. Transhepatic catheterization demonstrating the portal vein and the arteriovenous fistula (arrow) during the first procedure **[C]** and after the procedures **[D]**.