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Hepatic inflammatory pseudotumor: A case series

Guilherme D. Calomeni*, Elaine B. Ataíde, Ricardo R. Machado, Cecília A.F. Escanhoela, Larissa B.E. Costa, Ilka F.F. Boin

Rua Carlos Chagas, 420 (Setor de Transplante Hepático) – Cidade Universitária, Campinas 13083-878, SP, Brazil

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ABSTRACT

INTRODUCTION: Inflammatory pseudotumor (IPT) is a rare lesion consisted of inflammatory and myofibroblastic cells. These lesions may be found in different organs. There are less than 300 described cases.
PRESENTATION OF CASE: Case 1. 64-year-old cirrhotic male with a palpable epigastric mass. CT showed a lesion in liver segments 2 and 3 and left hepatic artery aneurism. Percutaneous embolization and wide spectrum antibiotics were tried, however the lesion grew. Left lateral hepatectomy was performed, and HIPT diagnosed. The patient died due to multiple organ dysfunction. *Case 2.* 30-year-old male with abdominal pain and fever. CT showed a hepatic hilar lesion. Surgical resection was performed after an ineffectual antibiotic trial, and HIPT was confirmed. The patient is doing well. *Case 3.* 73-year-old female with abdominal pain and fever. CT showed a 7 cm lesion in the left liver lobe. Unrewarding cancerous screening was performed, and unsuccessful antibiotic course was tried. Resection was performed, and HIPT diagnosed. The patient is doing well. *Case 4.* 50-year-old cirrhotic male with abdominal pain. CT showed a segment 6 lesion and portal vein thrombosis. Considering cancer as the first hypothesis and the MELD score of 9, segmentectomy was performed. HIPT was the final diagnosis. The patient died due to abdominal sepsis.
DISCUSSION: HIPT is a lesion with a vast list of differential diagnosis. Antibiotics are the first line of therapy, although surgery is often necessary. Overall prognosis is good, although comorbidities may worsen it.
CONCLUSION: HIPT is a rare and misleading entity.

1. Introduction

Inflammatory pseudotumor was first described in the lungs by Brunn in 1939.[1] It is a rare lesion consisted of inflammatory and myofibroblastic spindle cells. Albeit more common in the lung and orbits, these lesions are also found in the heart, liver, pancreas, spleen, stomach, small intestine, thyroid, meninges, and urinary bladder.[2]

Hepatic inflammatory pseudotumor (HIPT) consists of a rare lesion, with less than 300 cases described in medical literature.[3] It affects more commonly young adults, but can occur at any age. It is 3 times more common in males and is associated with other inflammatory lesions, as sclerosing cholangitis, retroperitoneal fibrosis and autoimmune diseases.[4] Its diagnosis and treatment are still a medical controversy. Here we describe 4 cases of HIPT.

2. Presentation of cases

2.1. Case 1

A 64-year-old Brazilian male with a past medical history of cirrhosis due to primary sclerosing cholangitis (PSC) was referred to our service because of a palpable epigastric mass and hemobilia. During initial evaluation, a painless, mobile, hard mass was palpable in the epigastric region. Laboratory findings consisted of hemoglobin 6 mg/dl, leukocyte count 6520/ml, total bilirubin 1.2 mg/dl, INR 1.44. Hepatic transaminases and renal function were normal. There was no serologic evidence of HBV and HCV infection. Upper gastrointestinal endoscopy identified esophageal varices without bleeding signs. Abdominal sonography showed a cirrhotic liver, a heterogeneous 62 mm × 50 mm lesion located in Couinaud's segments 2 and 3 in close relation to a left hepatic artery aneurism and mild intra-hepatic biliary dilation.

In face of these findings, the abdominal computadorized angiotomography was performed and showed a cystic lesion of 69 mm × 38 mm in segment 2 containing a branch of the left hepatic artery with a 12 mm × 10 mm aneurism with recent bleeding signs (Fig. 1). Furthermore, intra-hepatic biliary dilation and cholangitis related microabscesses could be seen.

* Corresponding author. Tel.: +55 1996611257.
E-mail addresses: guilhermecalomeni@yahoo.com.br (G.D. Calomeni), elainebataide@yahoo.com.br (E.B. Ataíde), ricardomachado@yahoo.com.br (R.R. Machado), cafesca@hotmail.com (C.A.F. Escanhoela), larrisa_eloy@yahoo.com.br (L.B.E. Costa), ilkaboin@yahoo.com (I.F.F. Boin).

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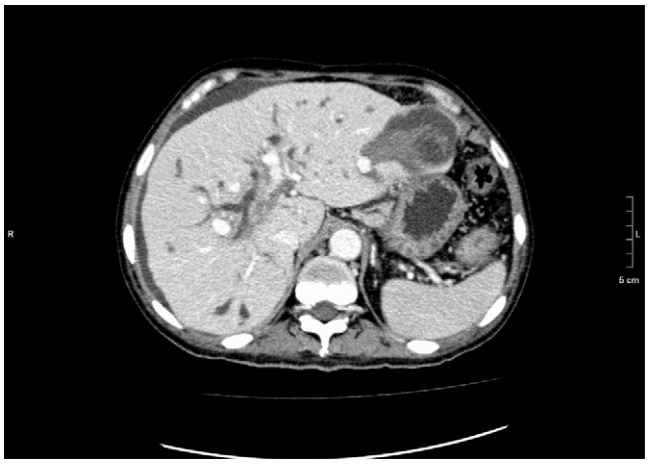

Fig. 1. Abdominal CT showing a cystic lesion in segment 2 and a left hepatic artery aneurism (patient 1).

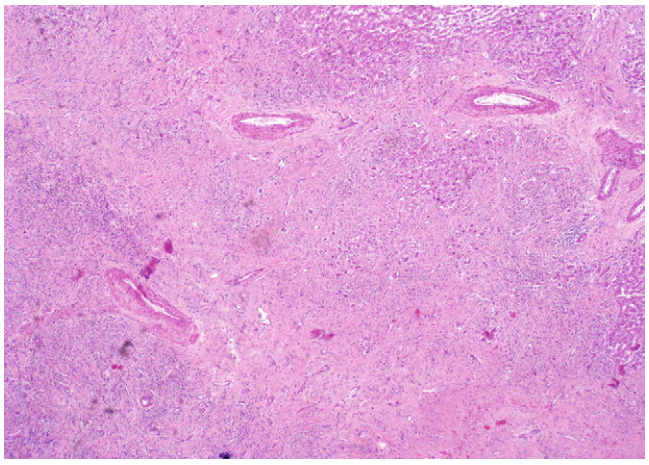

Fig. 3. Mixed inflammatory infiltrate and spindle cells are observed in this section (patient 2).

This patient was then submitted to percutaneous embolization of the left hepatic artery aneurism, which solved the intestinal bleeding. Nevertheless, daily fever persisted. Wide spectrum antibiotics were prescribed, but there was no improvement, which led to surgical exploration. A cirrhotic liver with biliomas in segments 2 and 3 associated with purulent discharge was found. A left lateral hepatectomy with cholecistectomy was performed. Histologic evaluation made the diagnosis of HIPT (Fig. 2).

After surgery, this patient presented with worsened hepatic function and cholangitis related sepsis. Blood cultures did not yield positive results, although leukocytosis with prominent left shift was found in different complete blood counts (CBC). Despite intensive care, he died on the 10th postoperative day, victim of systemic multiple organ dysfunction.

2.2. Case 2

A previously healthy 30-year-old Brazilian male was referred to our service because of complaints of epigastric pain, daily low grade fever and mild weight loss in the last 5 months. On physical evaluation, he was anicteric, well-nourished and experienced mild right upper quadrant pain with no palpable masses. Laboratory findings consisted of Hb 11.4 g/dl, normal hepatic transaminases and total bilirubin, and negative HBV and HBC serologies.

Investigation was carried out with abdominal CT that revealed a heterogeneous 5 cm hepatic hilar lesion. There was no biliary ductular dilation or vascular involvement. Differential diagnosis included lymphoproliferative disease and abscess.

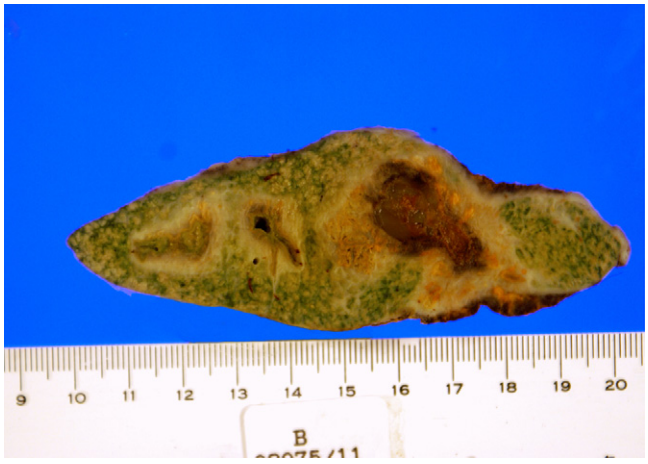

Fig. 2. Heterogeneous lesions in liver segment 2 (patient 1).

After an unsuccessful trial of wide spectrum antibiotics, surgical resection was performed. A right hepatectomy was carried out due to right hepatic lobe invasion by the lesion. Histologic and immunohistochemical analysis confirmed the diagnosis of HIPT (Fig. 3). This patient had no surgical complication whatsoever and is followed regularly, being kept asymptomatic.

2.3. Case 3

A previously hypertensive 73-year-old Brazilian female was referred to us due to right upper quadrant pain and daily fever in the past 10 days. She denied jaundice and weight loss. On physical examination, painful hepatomegaly was remarkable. Laboratory findings consisted of Hb 12 mg/dl, leukocyte count 6700/dl without left shift, slight increase in hepatic transaminases, and normal alkaline phosphatase and total bilirubin.

Abdominal sonography showed a solid lesion of approximately 130 cm^3 occupying segments 2 and 3. Abdominal CT confirmed this finding. Suspecting of a metastatic lesion with associated infection, antibiotics were prescribed and investigation with upper endoscopy and colonoscopy performed, yielding no abnormal findings. The patient clinical status worsened, therefore, surgery was indicated. A left hepatectomy was performed.

Histopathological analysis showed a nodular, firm, whitish lesion measuring 5.5 cm × 5.5 cm x 4.5 cm. There was intense plasmacytoid round cells proliferation, interspersed by small lymphocytes and scarce fusiform cells. The first hypothesis was lymphoplasmacytic lymphoma, which led to investigation of some immune markers. The final diagnosis was HIPT.

This patient had no postoperative complication, was discharged from hospital in the 6th postoperative day and is followed regularly in an outward fashion, being kept asymptomatic.

2.4. Case 4

A 50-year-old Brazilian male with past medical history of compensated cirrhosis (Child–Pugh A) due to HCV was referred to our service due to abdominal pain and increased abdominal volume. During physical examination, ascites was noted, but there were no palpable lesions. Laboratory findings consisted of total bilirubin 2.1 mg/dl, AST 75 U/L, ALT 85 U/L, creatinine 1.89 mg/dl, INR 1.34, Hb 9.9 g/dl, and leukocyte count 11,740/ml without left shift.

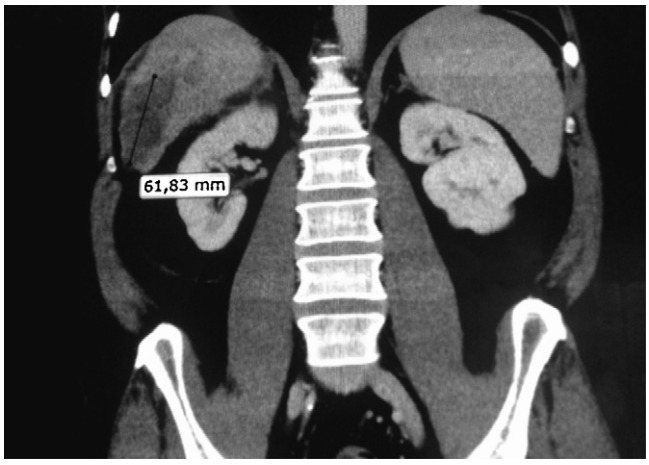

Fig. 4. Abdominal CT showing a heterogeneous lesion in segment 6 of a cirrhotic liver (patient 4).

Abdominal CT showed evidence of chronic liver disease, a heterogeneous lesion of 6.1 cm in segment 6, and portal vein thrombosis (Fig. 4). The main diagnostic hypothesis was hepatocellular carcinoma.

After clinical support and physiologic hydration, patient's renal function improved and his Model for End Stage Liver Disease (MELD) score was 9. Considering the contraindications to liver transplantation and arterial chemoembolization, the final decision was to perform surgical resection. A segmentectomy of segment 6 was carried out. Immunohistochemical analysis diagnosed HIPT.

The immediate postoperative evolution was satisfactory, with hospital discharge occurring on the 3rd day. However, 20 days later, this patient was readmitted because of abdominal sepsis (bacterial peritonitis). Paracentesis fluid analysis demonstrated elevated leukocyte count (over 1500/mm^3), with neutrophilic prevalence. Cultures presented proliferation of multiple bacteria. Wide spectrum antibiotics and intensive care were instituted, but acute renal failure supervened, and he died.

3. Discussion

HIPT was first described by Pack and Baker in 1953.[5] It is a rare, although well established, entity. It occurs more frequently in men around their thirties, but it can affect any age group. Its association with other inflammatory and autoimmune diseases is well know, including PSC, as occurred with our first patient. HIPT etiology is still unclear. Some authors believe it to be a low grade fibrosarcoma with inflammatory cells. However, malignant progression is not common. Others claim it to be an inflammatory response to surgical aggression or trauma. Finally, some say its origin is infectious.[6]

Right upper quadrant abdominal pain, intermittent fever, jaundice, and weight loss are the main signs and symptoms presented by patients with HIPT. Laboratory investigation normally does not yield clarifying results, with radiologic investigation been more elucidative. Nevertheless, image findings are not specific, and differential diagnosis with other hepatic lesions, including malignant ones, is difficult. This problem occurred in case 4. HIPT may present with different enhancement patterns on CT or MRI, as the lesion may be hypo or hypervascular.[7] We do not know any other description of HIPT associated with hepatic artery aneurism.

Considering the lack of specificity of imaging studies, HIPT diagnosis is a real medical challenge. Many advocate the use of radioguided percutaneous biopsy, as it would allow diagnosis through the demonstration of spindle cells, myofibroblast and mixed inflammatory infiltrate (plasmacytes, lymphocytes, and histiocytes). However, the biopsy itself is not harmless. There are descriptions of bleeding, malignant seeding in needle path (in the case of cancerous lesions), and infections secondary to the procedure.[8]

Since diagnosis is confirmed, clinical treatment with wide spectrum antibiotics and/or anti-inflammatory drugs is accepted as adequate. Absolute regression is the primary goal.[9] Surgery is deemed necessary when symptoms continue despite clinical treatment, lesion increases in size, vital structures are compressed by the tumor, or there is diagnostic uncertainty.[10] In our first 3 cases, the absence of patient improvement with antibiotic therapy made surgery necessary. On the other hand, diagnostic uncertainty demanded surgery in case 4.

Surgical results described in medical literature are encouraging, as occurred in cases 2 and 3. Nevertheless, liver cirrhosis increases surgical risks. In fact, we had two early deaths in our series, both occurring in cirrhotic patients. They faced complications in the early postoperative period, and their management was impaired because of liver dysfunction. This difficulty was predictable, but it did not make surgery less necessary. They had high surgical risk due to their baseline health problems, although surgical resection was indispensable.

4. Conclusion

IPT is a rare entity that can occur in the liver in association with other inflammatory diseases. Its symptoms are nonspecific, and its diagnosis is intriguing. Clinical treatment is desirable, although not always sufficient. Surgical resection is indispensable in many patients. The limited number of described cases contributes to the great amount of controversy regarding this medical problem.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Written informed consent was obtained from patients 2 and 3 for publication of this case report and accompanying images. For patients 1 and 4, written informed consent was obtained from their next of kin for publication of this case report and accompanying images.

Authors' contributions

G.D.C. analyzed patient data, interpreted results, and wrote the article. E.B.A. analyzed patient data, and reviewed the article. RRM collected patient data. C.E. collected patient data, and contributed to immunohistochemical analysis. L.B.E.C. contributed to immunohistochemical analysis. I.F.F.B. designed the study, and reviewed the article. All authors read and approved the final manuscript.

References

1. Umiker WO, Iverson LC. Post inflammatory tumor of the lung: report of four cases simulating xanthoma, fibroma or plasma cell granuloma. *The Journal of Thoracic Surgery* 1954;**28**:55–62.
2. Kim YW, Lee JG, Choi JS. Inflammatory pseudotumor of the liver treated by hepatic resection: a case report. *Yonsei Medical Journal* 2006;**47**:140–3.
3. Ntinas A, Kardassis D, Vrochides D. Inflammatory pseudotumor of the liver: a case report and review of the literature. *Journal of Medical Cases Reports* 2011;**5**:196.

4. Torzilli G, Inoue K, Midorikawa Y, Hui A, Takayama T, Makuuchi M. Inflammatory pseudotumors of the liver: prevalence and clinical impact in surgical patients. *Hepato-Gastroenterology* 2001;**48**:1118–23.
5. Pack GT, Baker HW. Total right hepatic lobectomy; report of a case. *Annals of Surgery* 1953;**138**:253–8.
6. Narla L, Newman B, Kolli R. Inflammatory pseudotumor. *RadioGraphics* 2003;**23**(3):719–29.
7. Kato H, Kanematsu M, Kondo H, Osada S, Goshima S, Yamada T, et al. Inflammatory pseudotumor of the liver: ferumoxide enhanced MR imaging as a tiebreaker. *Journal of Magnetic Resonance Imaging* 2004;**20**:501–5.
8. Takamori R, Wong LL, Dang C, Wong L. Needle-tract implantation from hepatocellular cancer: is needle biopsy of the liver always necessary? *Liver Transplantation* 2000;**6**(1):67–72.
9. Koea JB, Broadhurst GW, Rodgers MS, McCall JL. Inflammatory pseudotumor of the liver: demographics, diagnosis, and the case for nonoperative management. *Journal of the American College of Surgeons* 2003;**196**:226–35.
10. Akatsu T, Wakabayashi G, Kitajima M. Inflammatory pseudotumor of the liver masquerading as hepatocellular carcinoma after a hepatitis B virus infection. *Surgery Today* 2006;**36**:1028–31.