Inflammatory myofibroblastic tumour of the liver. Case report

Karina Olvera,[1] Josué Barahona-Garrido,[2,3] Alfonso Alfaro De-León,[1] Félix I Téllez-Ávila,[1] Mario Vilatobá,[4] Jesús K Yamamoto-Furusho[1]

[1] Department of Gastroenterology, Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán. Mexico City, Mexico.
[2] Unit of Gastroenterology and Digestive Endoscopy. Hospital Las Américas / Grupo Hospitalario Guatemala. Guatemala City, Guatemala.
[3] Instituto de Enfermedades Digestivas y Nutricionales. Guatemala City, Guatemala.
[4] Department of Surgery. Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán. Mexico City, Mexico.

Acta Gastroenterol Latinoam 2012;42:220-223

Summary

Inflammatory myofibroblastic tumour (IMT), also called inflammatory pseudotumour, is an uncommon neoplastic benign lesion histopathologically characterized by fibroblast and myofibroblast proliferation, with inflammatory cell infiltration. Systemic inflammatory response can appear although specific pathogens are rarely found. We present a case of a woman in whom liver abscess was initially suspected. The absence of microorganisms in the tissue obtained by needle aspiration biopsy and the lack of antibiotic therapy response indicated hepatic resection that lead to diagnosis of IMT of the liver, which has to be differentiated from abscess and abscessed tumours. The aim of this case report is to analyze the clinical presentation, course, diagnostic methods, therapies, and existing evidence of the possible pathogenic mechanisms leading to this neoplasm.

Key words. *Inflammatory pseudotumour, hepatic tumour, liver abscess, Mexico, Guatemala.*

Tumor miofibroblástico inflamatorio del hígado. Presentación de un caso

Resumen

El tumor miofibroblástico inflamatorio, también conocido como tumor seudoinflamatorio, es una neoplasia benigna infrecuente caracterizada histológicamente por infiltrado inflamatorio con proliferación de fibroblastos y miofibroblastos. Puede presentarse con signos de respuesta inflamatoria sistémica, aunque raramente se encuentran patógenos específicos. Se presenta el caso de una mujer en la que se sospechó inicialmente en absceso hepático. Ante la ausencia de microorganismos en el material obtenido de la biopsia por punción con aguja de aspiración y la inadecuada respuesta a la terapia antibiótica se indicó la realización de una resección hepática, lo que condujo al diagnóstico definitivo de un tumor miofibroblástico inflamatorio del hígado, lesión que debe diferenciarse del absceso hepático y de los tumores abscedados. El objetivo de este reporte de caso es analizar la presentación, el curso clínico, los métodos diagnósticos, y las alternativas terapéuticas, así como la evidencia que existe de los posibles mecanismos patológicos responsables de esta neoplasia.

Palabras claves. *Pseudotumor inflamatorio, tumor hepático, absceso hepático, México, Guatemala.*

The inflammatory myofibroblastic tumour (IMT) has been described as a rare benign lesion histologically characterized by proliferation of fibroblasts, myofibroblasts, and inflammatory cells.

Case report

A 23-year-old woman with history of surgical treatment for congenital type-3 esophageal atresia during infancy (disconnection of distal tracheoesophageal fistula, closure of the tracheal defect and primary anastomosis of the esophagus) was admitted to the emergency department because of systemic inflammatory response and a painful liver mass.

Correspondence: *Josué Barahona-Garrido*
Unit of Gastroenterology and Gastrointestinal Endoscopy
Hospital Las Américas. Grupo Hospitalario Guatemala
10 calle 2-45 zone 14, office 204. Guatemala City 01014, Guatemala.
Phone: 23854915, 23854916 Fax: (502) 23854913
E-mail: gastromedic@gmail.com

One week prior to admission patient initiated intermittent fever and two days later she presented right upper abdominal pain that was increasing until affected daily activities. Patient had no history of chronic liver disease but was chronically emaciated (body mass index of 15.4 kg/m²). At admission, she had blood pressure of 90/50 mmHg, body temperature of 39 °C, cardiac rate of 120 per minute, and respiratory rate of 27 per minute were noted. A right upper abdominal mass was evident by palpation. Liver function tests were within normal values, except for alkaline phosphatase of 199 IU/mL (normal upper limit: 132 IU/mL) and albumin of 1.4 g/dL. Magnetic resonance image showed a 10 cm encapsulated, lobulated, and radially septated lesion with a central scar (Figure 1). Alpha-fetoprotein was within normal levels and patient was negative for the enzyme-linked immunosorbent assay for anti-*Entamoeba histolytica*. A pyogenic abscess of the liver was initially suspected. Ultrasound-guided needle aspiration biopsy was performed in order to

Figure 1. *Axial (a) and coronal (b) magnetic resonance images show a 10 cm diameter lesion located in the right lobe of the liver, with heterogeneous and radially-septated appearance.*

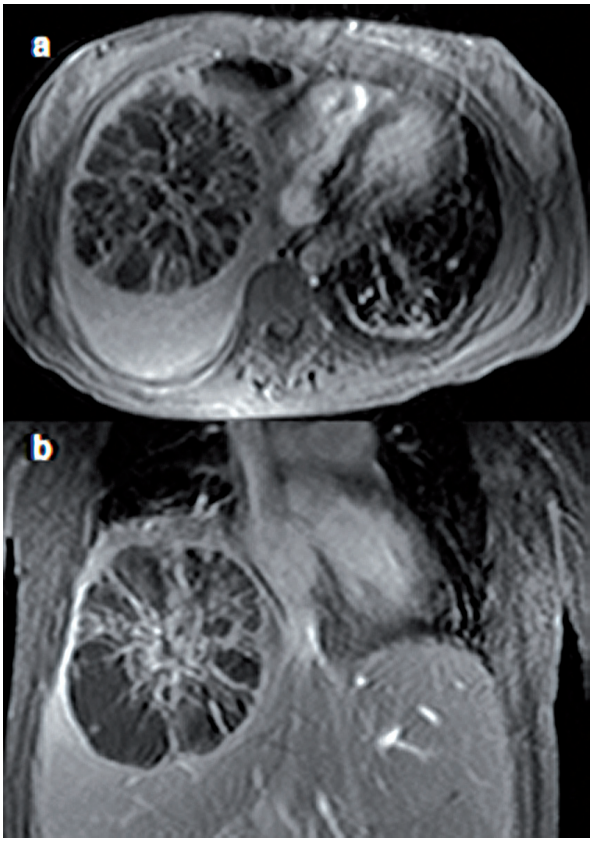
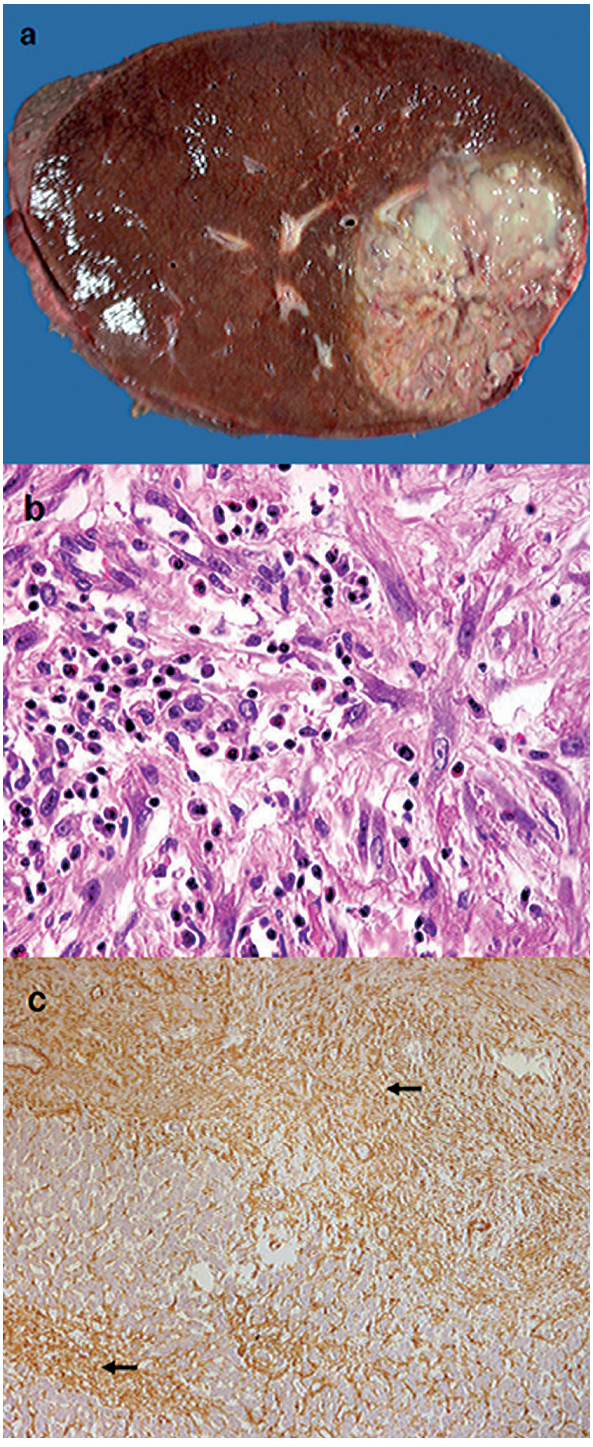


Figure 2. *Partial hepatectomy specimen showing a well-demarcated nodule with purulent material (a). The nodule was composed of spindle fibroblast cells and a dense inflammatory infiltrate of polymorphonuclear cells and foamy histiocytes (b). Immunoreactive staining for actin highlights the infiltration of myofibroblastic cells (arrows), characteristic finding of the inflammatory myofibroblastic tumour (c).*



obtain material for microscopic and bacteriological examination. Microscopic analysis of the sample showed only necrosis and inflammatory cells in absence of microorganisms. Tissue and blood cultures were negative. Ceftriaxone plus metronidazole were initiated without achieving clinical improvement. Surgical resection was indicated under suspicion of abscessed neoplasm. Intraoperative ultrasound did not bring additional information. Open right trisegmentectomy under selective hepatic vascular exclusion was performed. Hemorrhage was the only complication during surgery and 1,000 mL of packed red blood cell and 800 mL of fresh frozen plasma transfusions were given. Fever was noted after surgery but no infection was identified. Histopathological analysis showed a lesion composed of stellated spindle fibroblasts, myofibroblasts, foamy histiocytes, and polymorphonuclear cells, a group of features characteristic of the IMT (Figure 2) and distinctive from other more commonly seen diseases like abscesses or abscessed neoplasms (Figure 2). Patient required six days at Intensive Care Unit and was discharged eight days later. Symptoms were resolved and no recurrence or complications became during a 3-year follow-up period.

Discussion

The IMT represents a diagnostic dilemma because it closely resembles abscesses and malignant tumours, particularly when lesions are multiple or located in extrahepatic organs. Moreover, there are no specific biochemical markers for IMT and some cases are diagnosed after surgery or autopsy.[1-5] Needle aspiration biopsy can be useful for diagnosis,[6] but in our case open right trisegmentectomy was also required. Characteristically, the incidence is higher in the infancy and young adulthood. Usual symptoms include intermittent high fever and weight loss.[7] Main symptoms of this patient were those for systemic inflammatory response (hypotension, fever, tachycardia and tachypnea) and abdominal pain, common findings in patients with liver abscess. As in other cases, abscess of the liver or fibrolamellar carcinoma of the liver was initially suspected and then diagnosed to be IMT.[3,8,9]

The etiology of IMT remains unknown, but several etiologic agents have been proposed such as parasites and bacteria.[8] Cholangitis, primary biliary cirrhosis, primary sclerosing cholangitis and liver cysts have been reported to be associated with IMT.[10] Hepatic IMT is commonly misdiagnosed as pyogenic liver abscess, which is required to be ruled out in order to avoid unnecessary surgical treatment because abscesses usually resolve with antimicrobial therapy or percutaneous tube drainage.[11] Surgical resection for IMT is preferred,[12] although spontaneous regression have been reported.[13] There are few reported cases of IMT in patients with biliary atresia,[14] but to our best knowledge, the association with esophageal atresia has never been described. IMT diagnosis should be considered in patients with systemic inflammatory response and an atypical solid mass in the liver. Biopsy and/or needle aspiration may be performed. If no diagnosis is achieved, surgical treatment and histopathologic analysis is required.

Conflict of interest. *None.*

Acknowledgments. *The authors thank Dr Aldo Torre, Dr Norberto C Chávez-Tapia, Dr Sandra García-Osogobio, Dr Karen Arispe-Angulo and Dr Isabel Vasquez-Manzano for the critical review of this document.*

References

1. Singh P, Singla M, Gupta N, Muzaffar J, Babu R, Sakhuja P, Aggarwal A, Kar P. Inflammatory pseudotumour of the liver: a diagnostic dilemma. Trop Gastroenterol 2008;29:40-43.
2. Chen HW, Lai EC, Huang XJ, Chen FN, Lu RL, Pan AZ, Lau WY. Inflammatory myofibroblastic tumours of the spleen and liver. Asian J Surg 2008;31:25-28.
3. Tsou YK, Lin CJ, Liu NJ, Lin CC, Lin CH, Lin SM. Inflammatory pseudotumour of the liver: report of eight cases, including three unusual cases, and a literature review. J Gastroenterol Hepatol 2007;22:2143-2147.
4. Salgado-Sánchez E, Flores-Flores J, Pérez-Toriz MU, Pérez-Cruz R, Salgado-Sánchez J. Myofibroblast tumour. Rev Gastroenterol Mex 2003;68:219-221.
5. Sürer E, Bozova S, Gökhan GA, Gurkan A, Elpek GO. Inflammatory myofibroblastic tumour of the liver: a case report. Turk J Gastroenterol 2009;20:129-134.
6. Yoshida T, Nishimori I, Kumon M, Kohsaki T, Taniuchi K, Ohtsuki Y, Onishi S. Inflammatory pseudotumour of the liver: report of a case diagnosed by needle biopsy. Hepatol Res 2003;27:83-86.
7. Kim SR, Hayashi Y, Kudo M, Matsuoka T, Imoto S, Sasaki K, Shintani S, Song KB, Park SY, Kim JH, Ando K, Koterazawa T, Kim KI, Ninomiya T. Inflammatory pseudotumour of the liver in a patient with chronic hepatitis C: difficulty in differentiating it from hepatocellular carcinoma. Pathol Int 1999;49:726-730.
8. Park KS, Jang BK, Chung WJ, Cho KB, Hwang JS, Kang YN, Kim MJ, Kwon JH. Inflammatory pseudotumour of liver: a clinical review of 15 cases. Korean J Hepatol 2006;12:429-438.

9. Yan FH, Zhou KR, Jiang YP, Shi WB. Inflammatory pseudotumour of the liver: 13 cases of MRI findings. World J Gastroenterol 2001;7:422-424.
10. Hoosein MM, Tapuria N, Standish RA, Koti RS, Webster GJ, Millar AD, Davidson BR. Inflammatory pseudotumour of the liver: the residuum of a biliary cystadenoma? Eur J Gastroenterol Hepatol 2007;19:333-336.
11. Khan R, Hamid S, Abid S, Jafri W, Abbas Z, Islam M, Shah H, Beg S. Predictive factors for early aspiration in liver abscess. World J Gastroenterol 2008;14:2089-2093.
12. Tang L, Lai EC, Cong WM, Li AJ, Fu SY, Pan ZY, Zhou WP, Lau WY, Wu MC. Inflammatory myofibroblastic tumour of the liver: a cohort study. World J Surg 2010;34:309-313.
13. Yamaguchi J, Sakamoto Y, Sano T, Shimada K, Kosuge T. Spontaneous regression of inflammatory pseudotumour of the liver: report of three cases. Surg Today 2007;37:525-529.
14. Liu YW, Concejero AM, Chen CL, Cheng Y-F, Eng H-L, Huang T-L, Chen T-Y, Wang C-C, Wang S-H, Lin C-C, Yong C-C, Yang C-H, Jordan AP, Jawan B. Hepatic pseudotumour in long-standing biliary atresia patients undergoing liver transplantation. Liver Transpl 2007;13:1545-1551.