Agressive inflammatory myofibroblastic tumor of the liver with underlying schistosomiasis: A case report

Vera Lucia Pannain, Juliana Vial Passos, Ariovaldo da Rocha Filho, Cristiane Villela-Nogueira, Adriana Caroli-Bottino

Inflammatory myofibroblastic tumor (IMT) occurs infrequently in the liver. It is controversial whether it represents a low grade mesenchymal neoplasm or a reactive inflammatory lesion. Local recurrence and metastasis are rare and some tumors are associated with infectious agents. We report on a case of a large and partially resected IMT with local recurrence and diaphragm and kidney infiltration detected on routine surveillance two years later. Histologically, the tumor showed spindle cells without atypia, mitosis or necrotic areas in a myxoid and collagenized background with inflammatory cells. In the liver portal tracts, granulomatous lesions with viable eggs of Schistosoma mansoni were identified. Immuno-histochemistry demonstrated spindle cells which were smooth-muscle actin and vimentin positive. In conclusion, this case points out that these histological patterns do not predict the aggressive biological behavior of the lesion. A reason for the recurrence and the infiltration may be incomplete tumor resection. Further investigation is necessary in order to better clarify an infectious cause in some IMTs.

© 2010 Baishideng. All rights reserved.

Key words: Inflammatory myofibroblastic tumor; Liver; Recurrence; Schistosoma mansoni

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a lesion composed of myofibroblastic spindle cells, plasma cells, lymphocytes, and eosinophils. It can occur in soft tissues and visceras[1]. It was previously called plasma cell granuloma, inflammatory myofibrohistiocytic proliferation and inflammatory pseudotumor, but IMT is the designation currently used[1]. IMT is more frequently described in the lung and abdomen of young patients, but it can also be found in the central nervous system, salivary glands, larynx, bladder, breast, spleen, skin and liver[1].

Pack and Backer published the first case occurring in the liver[1]. Since then, reports on IMT in the liver with different progression have been described[1]. Concerning pathogenesis, it is controversial whether IMT is a neo-
plasm or a reactive pseudotumoral lesion. The inflammatory pseudotumor has been associated with trauma, auto-immune disease, and infectious disease. Recently, anaplastic lymphoma kinase (ALK) gene translocations or ALK protein expression in IMT has been reported, mainly in young patients.

Differential diagnosis of malignant disease is sometimes difficult. The clinical presentation is frequently associated with local mass and upper abdominal pain, as well as jaundice, intermittent fever, and weight loss. Surgical resection is the principal treatment, but corticosteroids and nonsteroidal anti-inflammatory therapy are sometimes used.

Here, we report on a case of a large incompletely resected IMT, with local recurrence and kidney and diaphragm infiltration on routine surveillance two years later.

CASE REPORT

A 40-year-old Brazilian woman born in northeast Brazil, presented with abdominal pain, fatigue and weight loss of 2 kg. She denied having fever or abdominal trauma, and she was receiving oxamniquine therapy for hepatic schistosomiasis. On physical exam, she presented with a hard mass that occupied the superior right quadrant of the abdomen and was painful at superficial and deep palpation. It was difficult to distinguish it from the liver that seemed enlarged primarily in its right lobe. Laboratory exams showed a cholestatic pattern, with elevated alkaline phosphatase (1176 U/L) and γ glutamyl transpeptidase (1341 U/L). The aminotransferases were also abnormal (aspartate transaminase = 108 mg/dL and alanine transaminase 92 mg/dL). Alpha-feto-protein, CEA, CA19.9 and CA125 levels were all normal.

At the ultrasound exam, a large isoechoic mass was identified in the 5th, 6th and 8th segments of the liver, and there was no mechanical obstruction of the biliary tract. Abdominal computer tomography (CT) confirmed its localization in the liver and described a close contact with the inferior caval vein. Laparotomy was indicated, and a right lobectomy of the liver was performed, but the tumor was incompletely resected because it was in contact with a large vessel. Two years later, routine CT scan detected local recurrence with diaphragm infiltration and a well defined mass in the right kidney. Again, the tumor was partially resected (including a right nephrectomy).

The gross appearance revealed a right hepatectomy specimen with a large mass measuring 11 cm (Figure 1) and other multinodular masses measuring 31 cm × 23 cm × 10 cm (Figure 2), which altogether weighed 3 kg. Those lesions were well circumscribed, and the cut surface was whorled, firm, and shining, with a myxoid aspect and a whitish or yellowish appearance (Figures 1 and 2). The adjacent liver parenchyma was unremarkable. Histologically, the tumor was characterized by spindle cell proliferation with plasmocytes, lymphocytes, histiocytes, and a few neutrophils, dispersed in a myxoid or dense collagen background (Figure 3). Cellular atypia was not identified, neither were mitotic or necrotic areas, and staining did not show the presence of microorganisms. By immunohistochemical analysis, the spindle cells were uniformly positive for vimentin and smooth muscle actin (SMA), supporting the myofibroblastic nature of these cells (Figure 4). There was no reactivity for CD34, CD23, CD117, EBV, P53 and ALK1. The CD68 was positive in histiocytes. In the liver parenchyma, granulomatous lesions with viable eggs of Schistosoma mansoni were identified in the portal tracts. The right kidney showed a large mass in its upper portion measuring 19 cm × 16 cm × 15 cm with the same macroscopic, microscopic and immunohistochemical findings as those described in the liver tumors.

DISCUSSION

IMT is a lesion with intermediate biological behavior that may recur with local or surrounding infiltration or rarely metastasizes. Its usual radiological aspects are similar to those of a malignant neoplasm. It is described in almost all solid organs, including the liver. Patients with hepatic IMT may complain of abdominal pain, jaundice and obliterative phlebitis, but, in this report, the clinical symptoms were nonspecific. As described in the laboratory setting, we observed a cholestatic laboratory pattern suggestive of an infiltrative liver disease. The aminotransferases were fairly abnormal and the radiological exams suggested a malignant tumor.

The pathogenesis of IMT is uncertain. Some cases
Hepatic inflammatory mass might have enlarged together with admissions such as the vena cava did not allow the complete resection of the tumor. Finally, the histological patterns did not predict the aggressive biological behavior, and further investigation is necessary in order to better clarify an infectious cause in some cases of IMT.

REFERENCES

1. Coffin CM, Fletcher CD. Inflammatory myofibroblastic tumour. In: Fletcher CD, Unni KK, Mertens F, editors. WHO classification of Tumors: Pathology and Genetics tumours of Soft Tissue and Bone. Lyon: IARC Press, 2002: 91-93
2. Pack GT, Baker HW. Total right hepatic lobectomy: report of a case. Ann Surg 1953; 138: 253-258
3. Morotti RA, Legman MD, Kerkar N, Pawel BR, Sanger WG, Coffin CM. Pediatric inflammatory myofibroblastic tumor with late metastasis to the lung: case report and review of the literature. Pediatr Dev Pathol 2005; 8: 224-229
4. Hussong JW, Brown M, Perkins SL, Dehner LP, Coffin CM. Comparison of DNA ploidy, histologic, and immunohistochemical findings with clinical outcome in inflammatory myofibroblastic tumors. Mod Pathol 1999; 12: 279-286
5. Sato Y, Harada K, Nakamura Y. Hepatic inflammatory pseudotumor related to autoimmune pancreatitis. Histopathology 2004; 45: 418-419
6. Bankole-Sanni R, Coulibaly B, Denoulet D, Nandiolo R, Mobiot L, Oulai. Inflammatory pseudo-tumor of the liver in a child: case report. Med Trop (Mars) 1997; 57: 269-272
7. Ji XL, Shen MS, Yin T. Liver inflammatory pseudotumor or parasitic granuloma? World J Gastroenterol 2000; 6: 458-460
8. Gómez-Román JJ, Sánchez-Velasco P, Ocejo-Vinyals G, Hernández-Nieto E, Leyva-Cobán F, Val-Bernal JF. Human herpesvirus-8 genes are expressed in pulmonary inflamma-
Pannain VL et al. Aggressive inflammatory myofibroblastic tumor of the liver

tory myofibroblastic tumor (inflammatory pseudotumor). Am J Surg Pathol 2001; 25: 624-629
9 Rosenbaum L, Fekrazad MH, Rabinowitz I, Vasef MA. Epstein-Barr virus-associated inflammatory pseudotumor of the spleen: report of two cases and review of the literature. J Hematop 2009; 2: 127-131
10 Cook JR, Dehner LP, Collins MH, Ma Z, Morris SW, Coffin CM, Hill DA. Anaplastic lymphoma kinase (ALK) expression in the inflammatory myofibroblastic tumor: a comparative immunohistochemical study. Am J Surg Pathol 2001; 25: 1364-1371
11 Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. Am J Surg Pathol 1995; 19: 859-872
12 Dishop MK, Warner BW, Dehner LP, Kriss VM, Greenwood MF, Geil JD, Moscow JA. Successful treatment of inflammatory myofibroblastic tumor with malignant transformation by surgical resection and chemotherapy. J Pediatr Hematol Oncol 2003; 25: 153-158
13 Tsou YK, Lin CJ, Liu NJ, Lin CC, Lin CH, Lin SM. Inflammatory pseudotumor of the liver: report of eight cases, including three unusual cases, and a literature review. J Gastroenterol Hepatol 2007; 22: 2143-2147
14 Kalil M, Battisti Netto O, Vieira LCA, Cintra LC. Forma pseudotumoral intra-abdominal da esquistossomose. Rev Col Bras Cir 2006; 33: 203-204
15 Andrade ZA. Schistosomal hepatopathy. Mem Inst Oswaldo Cruz 2004; 99: 51-57
16 Someren A. "Inflammatory pseudotumor" of liver with occlusive phlebitis: report of a case in a child and review of the literature. Am J Clin Pathol 1978; 69: 176-181
17 Chan JK, Cheuk W, Shimizu M. Anaplastic lymphoma kinase expression in inflammatory pseudotumors. Am J Surg Pathol 2001; 25: 761-768
18 Tsuzuki T, Magi-Galluzzi C, Epstein JI. ALK-1 expression in inflammatory myofibroblastic tumor of the urinary bladder. Am J Surg Pathol 2004; 28: 1609-1614
19 Coffin CM, Hornick JL, Fletcher CD. Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. Am J Surg Pathol 2007; 31: 509-520

S- Editor Tian L L- Editor O'Neill M E- Editor Lin YP