Spontaneous regression of inflammatory pseudotumor of the liver : a case report

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Inflammatory pseudotumor of the liver is a rare benign mass that is often misdiagnosed as hepatocellular carcinoma or hepatic abcess. It is a benign entity of unknown origin : etiology and pathogenesis remain unclear, but it is speculated that this tumour is a reactive inflammatory condition (1).

The lesion is characterized by encapsulated proliferating connective tissue admixed with lymphocytes, macrophages and granular leukocytes (2). Clinical presentation and morphological appearance may vary (3,4). Different kind of treatment have been proposed. In the vast majority of the cases, a surgical excision has been performed and in other cases a conservative treatment (antibiotics or steroids) has been given (1,5). However, the need to treat this lesion has not been fully demonstrated and until now, only few cases of spontaneous regression have been recently described (6-9).

The purpose of this report is to present a new case of inflammatory pseudotumor of the liver regressing spontaneously.

Case report

In October and November 2000, a 19 year-old man presented 3 times on a 15 day-period to the emergency department for a pain localized in the right flank, without irradiation. The pain was relentless, with some paroxysms. One single episode of fever (37.8 °C) occurred 10 days after the beginning of symptoms.

Past history was unremarkable. Laboratory results showed increasing inflammatory response and mild leucocytosis. Urine and blood cultures remained negative. Acute appendicitis was suspected. An abdominal ultrasound showed an ascending appendix without inflammatory sign. It is not known whether the liver was investigated. Computed tomography (CT) of the pelvis also disclosed a thin-walled appendix, without peri-appendicular fat infiltration but mesenteric lymphadenopathies. Two weeks later, he was seen at the outpatient clinic with a suggested diagnosis of acute appendicitis (lower abdominal pain and leucocytosis). At physical examination, tenderness was elicited in the right and left lower quadrants without defence or rebound tenderness. Mc Burney and Murphy's points were painless on palpation. Laboratory results showed the regression of the inflammatory syndrome. The liver enzymes were consistently normal. An ileoscopy with random biopsies and a small bowel meal showed no anomaly. The pain resolved slowly and spontaneously. An ultrasound examination of the upper abdomen disclosed a hypoechoic, non homogeneous mass, 55 mm high × 51 mm wide in the right hepatic lobe. CT showed a hypodense tumor located antero-laterally at the periphery of the right part of the liver. Its diameters were estimated 42 mm wide and 38 mm high (fig. 1a). A peripheral rim enhancement was observed at contrast CT. The biliary tree was normal and there was no sign of portal vein obstruction. An intra-dermoreaction to tuberculin remained negative. Serum alpha-fetoprotein was in the normal range. All serologic tests remained negative : ANCA, ANA, antimitochondrial antibodies, as well as serology for Brucella, Yersinia, Treponema, Rickettsia (conori, mooseri, burnetti), Coxiella, Bartonella henselae, Toxoplasma, hepatitis B and C viruses, HIV, Epstein-Barr virus and cytomegalovirus.

A biopsy of the mass, performed under CT guidance, showed an important fibrosis infiltrated by lymphocytes and plasmocytes (fig. 2). Inside, some residual biliary ducts and clusters of hepatic parenchyma showed cholestasis. Peripherally, one focus of necrosis was observed, without any polymorphonuclear inside but surrounded by fibrous tissue with scattered histiocytes, lymphocytes and numerous small vessels but no giant cells. Staining with Ziehl, Giemsa and periodic acid-Schiff reagents did not show any microorganism.

The patient was seen 1, 4 and 7 months later. He was symptom-free, and clinical examination and laboratory results were normal. CT of the abdomen at one month showed some regression of the mass, whose diameter was 25 mm. At 4 months, the mass was reduced to a small (10 × 15 mm) hypodense image, probably fibrous

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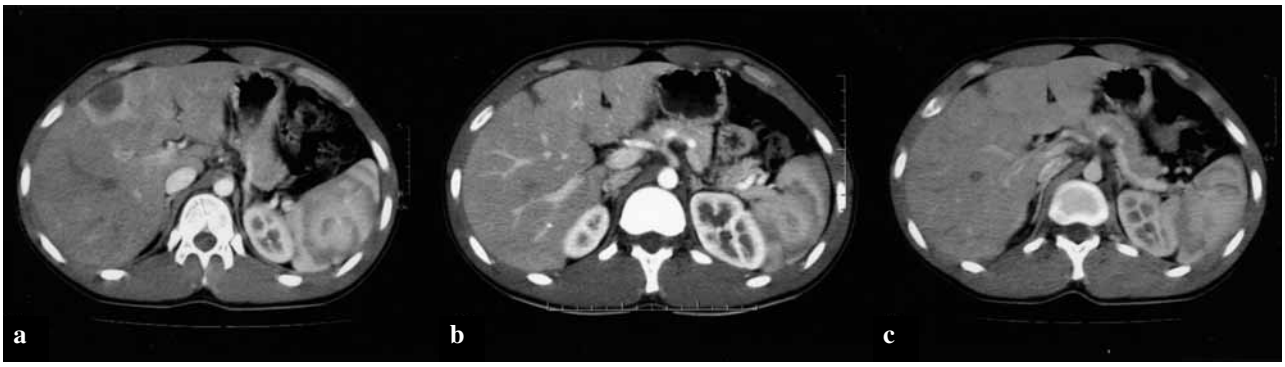

Fig. 1. — Computed tomodensitometry evolution of the liver inflammatory pseudotumor at the time of diagnosis (a), 4 months (b) and 7 months (c) later, respectively.

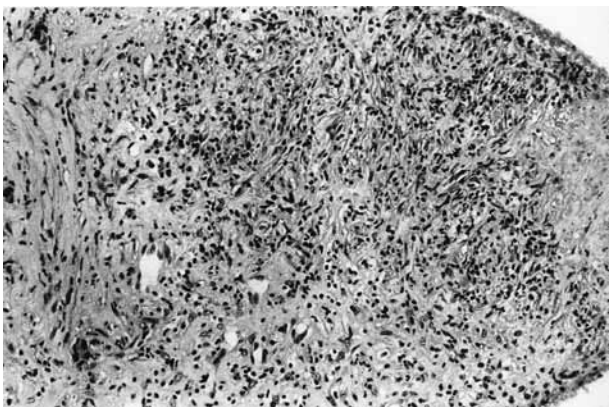

Fig. 2. — Liver histology of the lesion showing fibrosis infiltrated by lymphocytes and plasmocytes (H.E.S. ; original magnification, ×125).

tissue making a notch on the surface of the liver (fig. 1b). At 7 months, the mass had disappeared (fig. 1c).

Discussion

We describe the case of a 19 year-old man with a rather large liver tumor discovered in a context of a right flank pain but otherwise in excellent general condition corresponding to an inflammatory pseudotumor. Liver pathology showed, in the middle of an important fibrosis, a necrotizing granulomatous lesion. Intra-dermoreaction to tuberculin and all the serological tests for autoimmune and potential infectious diseases remained negative. Without any treatment, the pain resolved completely and the liver pseudotumor regressed completely.

Inflammatory pseudotumor is a rare benign condition which may occur in many organs including the spinal cord meninges (10) and testicular tunic (11). Liver involvement was first described in 1953 by Pack and Baker (12).

Basically, histopathologic patterns reveal an encapsulated fibro-vascular tissue that contains inflammatory cells and that may exhibit changing cellular composition (13). This benign tumor has been described as having the propensity to invade blood vessels and bile ducts resulting in occlusive phlebitis and biliary obstruction (2,14).

Clinical signs, laboratory results and imaging features of inflammatory pseudotumor of the liver are nonspecific. The most common presenting symptoms are fever, weight loss and upper abdominal pain. Laboratory data usually show systemic signs of inflammation associated or not with abnormal liver function tests but without elevated serum alpha-fetoprotein activity (6) although high values of this tumor marker have been reported in one case (4).

Liver ultrasound demonstrates a variable pattern of echoic lesion. It has been documented to be hypo- or hyper-echoic with or without mosaic pattern (2,5) and with ill defined or well circumscribed margins (15,16). On CT, the lesion may be homogeneous or heterogeneous and hypo-, iso-, or hyper-dense after administration of contrast material (3,17,18). The tumor usually shows peripheral rim enhancement on delayed post-contrast CT (17). The lesion may be hyper- or hypo-vascular angiographically (1,3).

Beside the difficulties linked to its diagnosis, controversies do exist in the literature regarding the treatment which should be given (1,5). In most cases, a surgical excision has been performed and in many other cases, a conservative treatment (antibiotics or steroids) has been administered. The spontaneous regression observed in our case and in several other cases (6-9) outline the possibility of therapeutic abstention. However, it must be emphasized that three patients with liver pseudotumor have died and that potentially in relation with this tumor outlining the need for careful follow-up in case of therapeutic abstention.

In conclusion, the spontaneous regression observed in our case of pseudotumor of the liver favours the possibility of absence of treatment in such a case. It must however be outlined that an histological diagnosis and a careful follow-up must be done if this therapeutic option is chosen.

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