Delayed liver metastasis of a meningeal solitary fibrous tumor

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Abstract

Solitary fibrous tumors (SFT's) are rare soft tissue neoplasms of mesenchymal origin, most commonly reported in the thoracic cavity. They exhibit an aggressive and infiltrative nature and have a tendency to recur either locally or distantly, the latter typically being a late event. Primary therapy consists of complete excision and prognosis is poor in case of recurrence. In this manuscript, we discuss the imaging features and treatment options for a patient presenting with delayed liver metastasis ten years after treatment for a meningeal SFT. (Acta gastroenterol. belg., **2011**, 74, **567-569**).

Key words : solitary fibrous tumor, haemangiopericytoma, delayed liver metastasis.

Introduction

Solitary fibrous tumors (SFT's) are rare soft tissue neoplasm of mesenchymal origin, with features of pericytic, fibroblastic, and myofibroblastic differentiation. The cellular variant of SFT is virtually indistinguishable from Stout's description of haemangiopericytoma (HPC), which is a rare soft tissue neoplasm of pericyte origin (1). Based on recent insights, SFT and HPC are seen as a spectrum of a single entity (1,2). SFT is reported in almost every site of the body, most commonly occurring in the thoracic cavity. In this manuscript we present a case of delayed liver metastasis of a primary meningeal SFT. Imaging features and treatment are discussed.

Case report

A 37-year old man presented with vague abdominal complaints and right upper quadrant pain. His medical history revealed a resection of a solitary fibrous tumor of the supratentorial meninges 13 years ago. Three years later a local meningeal recurrence was treated with surgical resection and postoperative radiation (total dose 55.8 Gy). Clinical examination on admission 10 years later, showed signs of hepatomegaly. Blood analysis showed elevated levels of alkaline phosphates (792 U/L, normal upper level 270 U/L) and gamma-GT (284 U/L, normal upper level 53 U/L). Abdominal ultrasound confirmed an enlarged liver with the presence of multiple, large-sized, nodular lesions. The working hypothesis included hepatic metastasis of an unknown primary versus delayed metastasis of a solitary fibrous tumor. Additional tests for blood tumor markers (CEA, NSE, CA 19-9, CA 125) showed normal values. Abdominal CT imaging depicted the presence of 4 large-sized hepatic lesions (largest diameter for all lesions ranging between 7 and 18 cm) with peripheral enhancement and central foci of necrosis, and a large right-sided adrenal nodule (diameter 10 cm) with similar appearance (Fig. 1). There were no enlarged lymph nodes. CT of the brain and chest was normal. Subsequent ultrasound-guided biopsy of one of the hepatic lesions was compatible with metastasis of a solitary fibrous tumor, histologically identical to the previously resected intracranial lesions. The primary tumor consisted of a monotonous spindle cell proliferation with variable cellularity and a characteristic branching vascular pattern (Fig. 3). A similar picture was seen in the liver metastasis (Fig. 4a). There was partial CD34 expression (Fig. 4b) as expected

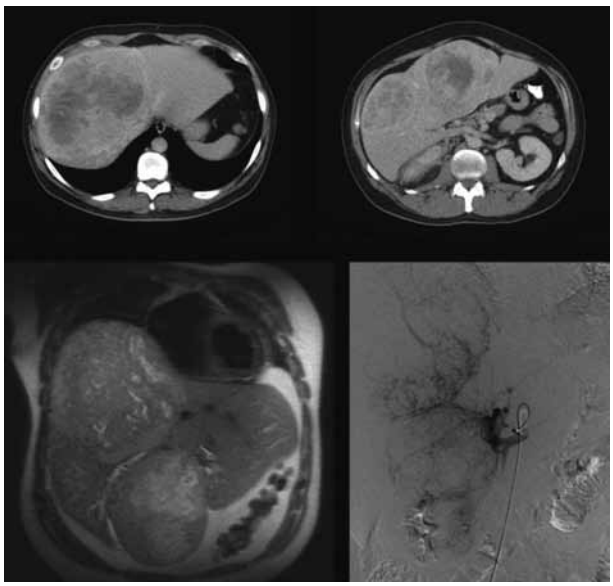

Fig. 1. — Multifocal, voluminous metastases of the liver and right adrenal gland on axial contrast-enhanced computed tomography (top row), coronal T2-weighted MRI (lower left) and conventional angiography (lower right). Note the hypervascularity of the lesions and foci of necrosis.

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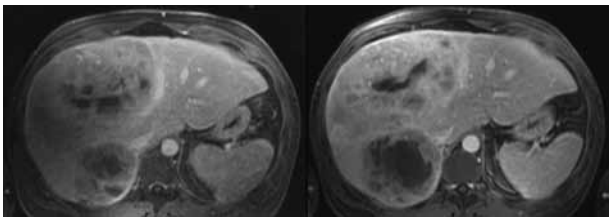

Fig. 2. — MRI before (left) and after (right) intrahepatic chemo-embolization therapy. No significant change in volume nor necrosis was observed.

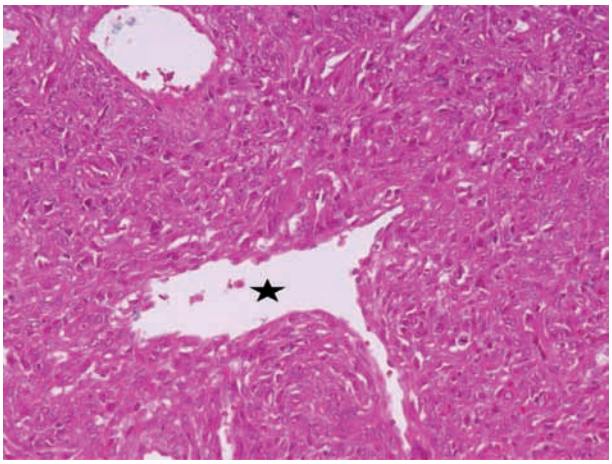

Fig. 3. — The primary tumor : monotonous spindle cell proliferation with variable cellularity and a characteristic branching vascular pattern.

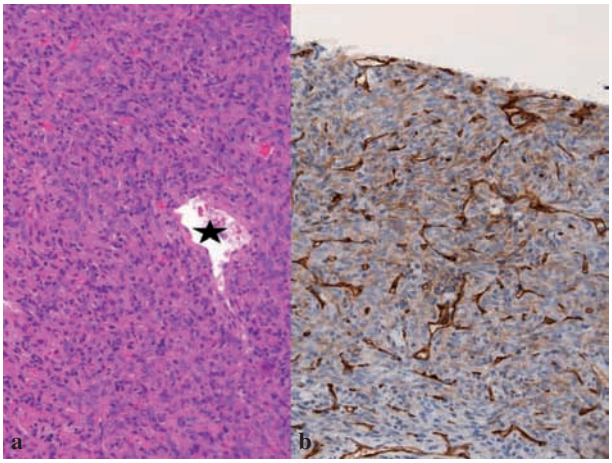

Fig. 4. — The liver metastasis : a. A similar picture : monotonous spindle cell proliferation with variable cellularity and a characteristic branching vascular pattern. b. Partial CD34 expression as expected for a solitary fibrous tumor.

for a solitary fibrous tumor. Based on the histological features the tumor was benign, but it is well known that the behaviour of SFT is unpredictable, ie. there is no strict correlation between histology and clinical behaviour. The patient was treated with two cycles of intrahepatic chemo-embolization, using a mixture of microparticles (size range 250-900 μm) and Adriamycine (50 mg/cycle). Posttreatment magnetic resonance imaging showed no response and the patient was referred for further surgical exploration (Fig. 2). A right hepatic trisectionectomy (extended hepatectomy) and wedge resection of the left lobe was performed together with a righ-sided adrenalectomy. On follow-up 8 months after surgery, the patient presented with rapidly progressive disease – including a new intracranial tumoral lesion and multifocal pulmonary and skeletal metastatic disease. Palliative care was started.

Discussion

Intracranial SFT's are aggressive and infiltrative tumors with a tendency to recur either locally or regionally within the cranial or spinal meninges (3). Pathological criteria of malignancy include large tumor, size (more than 50 mm), infiltrative margins, high cellularity, nuclear pleomorphism, areas of tissue necrosis and increased mitotic index (more than 4 mitosis in 10 HPF) (4). A wide range of recurrence rates is reported, going from 38.7% to 90%, and in general recurrence is a late event (5,6,7). Age, sex, size, and location of the primary tumor are not associated with recurrence (5). SFT may also metastasize to distant organs, including bone, lung, kidney, pancreas, adrenal gland, and liver and rarely breast, thyroid, or lymph nodes, with a reported metastatic rate ranging between 12% and 57% (5,8). The development of these extraneural metastases also seems to be a late event with an average of 8 years after initial therapy (up to 16 years) (3). The probability of developing metastasis increases with time : Guthrie et al. reported a 5- and 10-year probability of developing metastases of 13% and 33% respectively, Jeong et al. reported a 5- and 10-year probability of 4.4% and 24.9% (5,6). Therefore long-term follow-up is mandatory. In contrast to epithelial tumors, SFT's typically develop voluminous, hypervascular and well circumscribed liver metastases with central areas of necrosis, as illustrated in Figure 1. These liver lesions should be differentiated from other hypervascular lesions such as hepatocellular carcinoma, adenoma or sarcoma. The treatment of choice is complete surgical resection (6,9). Spitz *et al.* reported in a study of 36 patients (treated between 1975 and 1995) a 5-year actuarial survival rate of 79% for cases (n = 26) surgically treated with curative intent, versus 50% for patients (n = 8) undergoing noncurative surgical treatment (9). Espat *et al*. reported a five year overall survival rate of 86% (n = 25) (10). Both studies had a median follow-up of less than 60 months, which is too short given the higher probability of developing distant metastasis after 5 year of follow-up. Some authors advocate a beneficial effect of prophylactic postoperative radiotherapy with a decrease in the local recurrence rate and an extension of the disease-free interval and overall survival (5,6,11). There is no clearly effective therapy in patients who develop locally recurrent or metastatic

disease. Re-resection and/or metastasectomy should be considered if technically feasible, whether or not in combination with radiotherapy (6,9). Stereotactic radiosurgery in recurrent, unresectable central nervous system SFT appears to be a potentially promising therapy (11). In the past, systemic chemotherapy has not shown significant clinical or radiologic responses, i.e. no effect on progression-free survival and overall survival (9,12). New agents, in particular anti-angiogenic drugs, seem promising in the treatment of patients with advanced SFT/HPC, based on the rich vascular characteristics of this tumor type (2). The use of various systemic agents such as interferon alfa, combination therapy with temozolomide and bevacizumab, sorafenib and sunitinib have been reported, but apart from disease stabilization early in therapy, all patients developed progressive disease (12,13,14,15,16).

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