CLINICAL CASE

A not so solitary fibrous tumor of the liver

Une tumeur fibreuse solitaire du foie... pas si solitaire !

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Summary Solitary fibrous tumor (SFT) is a rare neoplasm. Liver parenchyma is a rare location of SFT and, in this case, it usually follows a benign course. We report here the case of a 54-year-old man who presented a large SFT tumor of the right hepatic lobe. The tumor was surgically resected. Local recurrence occurred 6 years later as a 15 cm diameter liver tumor. Histological examination of the resected lesion showed features of an aggressive form of SFT. Two years later, the patient presented with complaints of neck pain and ensuing examinations revealed a tumor of the cranial base. A new surgical resection was performed and histological examination confirmed a metastasis of the SFT. Few weeks later, the patient presented an irreducible psoitis due to an iliac bone metastasis. He died within 1 month.

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Résumé La tumeur fibreuse solitaire est une néoplasie rare. Sa localisation hépatique est peu fréquente et, dans ce cas, est le plus souvent bénigne. Nous rapportons le cas d’un homme de 54 ans ayant présenté une volumineuse tumeur fibreuse solitaire du foie droit. Après un traitement chirurgical initial la tumeur récidivait six ans plus tard sous forme d’une lésion de 15 cm. L’analyse histologique montrait des caractéristiques en faveur d’une forme agressive. Deux ans plus tard, le patient a consulté pour des cervicalgies. Les examens réalisés ont révélé une tumeur de la base du crâne. Une nouvelle résection chirurgicale a été réalisée et l’histologie était en faveur d’une métastase de la tumeur fibreuse solitaire du foie. Quelques semaines plus tard, le patient a présenté un psoitis irréductible en lien avec une métastase osseuse iliaque. Il est décédé un mois plus tard.

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Introduction

Solitary fibrous tumor (SFT) is a rare neoplasm. Liver parenchyma is a rare location of SFT and, in this case, it usually follows a benign course. We report here the case of a patient who presented a SFT of the liver with local recurrence 6 years after the surgical resection, and a second recurrence 2 years later in the form of cervical then iliac bone metastases.

Case report

A 54-year-old-man consulted in 1999 for pain in the right hypochondrium and weight loss. The patient’s medical history comprised a hepatitis A virus infection 35 years earlier and an appendectomy. Clinical examination was normal. Laboratory test showed only an isolated elevation of gamma-glutamyl transpeptidase (438 IU/L) and an elevation of erythrocyte sedimentation rate (35—65 mm). An abdominal CT scan was performed and showed a large (16 cm) hypodense tumor of the right hepatic lobe. Only the peripheral portion and a few septa were enhanced on the portal phase (Fig. 1). Serological examinations for hydatid disease and tumor-related markers CA 19-9 and ACE were negative. No tumoral lesion was found either by colonoscopy or upper-gastrointestinal endoscopy. A right hepatectomy was carried out despite the lack of a diagnostic orientation. On the cut section, the tumor measured 17 cm in diameter and was cystic with hemorrhagic contents. Pathological examination showed a moderately cellular tumor of the right hepatic lobe. Only the peripheral portion and a few septa were enhanced on the portal phase (Fig. 1). Serological examinations for hydatid disease and tumor-related markers CA 19-9 and ACE were negative. No tumoral lesion was found either by colonoscopy or upper-gastrointestinal endoscopy. A right hepatectomy was carried out despite the lack of a diagnostic orientation. On the cut section, the tumor measured 17 cm in diameter and was cystic with hemorrhagic contents. Pathological examination showed a moderately cellular tumor. In the mucoid stroma, cells were polymorphic: sometimes small and round, sometimes spindle-shaped, sometimes voluminous with aci-dophilic cytoplasm, or with polymorphous nuclei. There were few mitoses (less than 5 for 10 high-power-fields) and no necrosis. Immunohistochemical studies revealed tumoral cells positive for vimentin, desmin, actin, and CD34 but negative for cytokeratin, S-100 protein, HMB45 and CD31.

The conclusion was a polymorphous sarcoma with mucoid stroma, grade 2 according to FNCLCC grading. The resection limits were free of tumor.

The patient then received regular follow-up including CT scan of the thorax, abdomen and pelvis every 6 months. In September 2004, there was no tumor in the liver. In October 2005, a new CT scan revealed a recurrence in segment IV of the liver. It appeared as a hypodense tumor, 10 cm in diameter. Compared to the first tumor, enhancement was still moderate and heterogeneous but a large vessel was well depicted within the tumor (Fig. 2). A surgical resection of the lesion was performed without incident. On the cut surface, the 15 cm tumor was heterogeneous, yellowish in colour with necrotic and hemorrhagic areas. Microscopy found a highly cellular tumor composed of oval or round cells (Fig. 3) with moderately atypical nuclei. Mitotic cells were numerous (20 mitoses in 10 high-power-fields) and sometimes atypical. Cytoplasmas were clear and some of them contained hyaline eosinophilic globules. There were also necrotic areas. The vascular pattern resembled hemangiopericytoma, with abundant branching vessels. Immunohistochemistry was positive for CD34 and Bcl2, but negative for desmin, S-100 protein, actin and CD117. The tumor was sent to an expert pathologist and identified as a SFT of the liver with features suggesting a malignant potential: recurrent tumor, large size, high mitotic rate, cellular pleomorphism with atypia, and necrosis (grade 3 according to FNCLCC grading).

Again, the patient received regular follow-up comprising CT scan every 6 to 12 months. A PET scan performed in June 2007 and a CT scan of the thorax, abdomen and pelvis in December 2007 showed no tumor recurrence. In January 2008, the patient consulted for neck pain, which had been evolving for 4 weeks and was resistant to second-level analgesics. A brain MRI displayed a large tumor of the cranial base (Fig. 4). The strong enhancement was clearly
Figure 3  Histological appearance of liver recurrence (hematoxylin and eosin stain): highly cellular tumor composed of oval or round cells with cytoplasmic hyaline grains (thin arrows) and hemangiopericytoma-like vascular pattern (thick arrows). Panel 3a: ×100 magnification, panel 3b: ×200 magnification.

different from the poor enhancement observed in the previous hepatic tumors. However, histological examination of a needle core biopsy found significant similarities with the hepatic tumor resected in 2005. This led to the conclusion of a probable metastasis of the SFT of the liver. Because the lesion was unique, embolization then surgical resection was carried out in March 2008. Histological examination of the tumor confirmed the diagnosis of metastasis of the SFT of the liver.

Few weeks after surgery, the patient presented with complaints of right leg pain. Irreducible psoriasis was diagnosed. An abdominal CT scan showed a third recurrence in the form of a large retroperitoneal tumor, which was poorly enhanced like the hepatic tumors, and comprised large vessels evoking the first recurrence (Fig. 5). The tumor invaded the iliac bone. There was neither hepatic recurrence of the SFT, nor cervical recurrence on the brain CT scan. No treatment was initiated as the patient’s health rapidly deteriorated. The patient died 1 month after the discovery of the pelvic mass.

Discussion

SFT is a rare neoplasm belonging to the group of soft tissue tumors. In the recent modified WHO classification of soft

Figure 4  MRI of the head, T1 weighted sequence after gadolinium injection. The second recurrence of the solitary fibrosis tumor appears as a large, strongly enhanced tumor of the cranial base.

Figure 5  Enhanced CT scan at portal phase. The third recurrence is a large retroperitoneal tumor, poorly enhanced, but with large vessels within the tumor (arrow). Tumor is causing significant compression of the vena cava.
tissue tumors, SFT has been classified into the intermediate (rarely metastasizing) category [1]. It usually affects the pleura, but can also be found in the peritoneum, the pericardium, and non-serosal sites such as lung parenchyma, the upper respiratory tract, the orbit, the thyroid, the parotid gland, or the thymus [2]. Liver parenchyma is a rare location of SFT and, in this case, it usually follows a benign course. Very few case reports are to be found in the literature that describe local recurrence of SFT of the liver, and even less so for metastases [3–7]. Our case report is remarkable by the very progressive form of SFT of the liver complicated with a first local recurrence 6 years after the surgical resection, and a second recurrence 2 years later in the form of cervical then iliac bone metastases.

Clinical and radiographic characteristics being insufficient, the diagnosis of SFT requires a histological analysis of a tumor sample [4]. Pathologic features of SFT of the liver are very close to those described for other locations. Typical lesions associate alternating hypercellular and hypocellular areas and a hemangiopericytoma-like vascular pattern. However, these patterns are not specific [3,8] and immunohistochemical study is very useful to eliminate other soft tissue tumors. Tumoral cells in SFT are positive for CD34 (90 to 95%) [9–11], CD99 (70%) and Bcl2. In the present case, the initial tumor and the local recurrence were positive for CD34, and Bcl2 staining was performed only on the recurrent tumor with positive result. Finally, a retrospective histological review by an expert pathologist found SFT of the liver for the first tumor. Discriminating SFT from hemangiopericytoma may be very difficult as these two soft tissue tumors have overlapping features including CD34 immunoreactivity. Actually, histogenesis of these neoplasms remains still debated and uncertain [12]. Finally, WHO classification had recently grouped together these two lesions under the same chapter [1].

The recurrent tumor presented the following histological features: large size, high mitotic rate, cellular pleomorphism with atypia, and necrosis, suggesting a SFT with an increased malignant potential [11,13]. The lack of series and the low number of cases reported in the literature limit the identification of prognostic factors for SFT of the liver. In addition to histological features, hypoglycemia also seems to be associated with a bad prognosis [6–8]. In these cases, careful follow-up for tumor recurrence after surgical resection seems mandatory, but its modalities (frequency of follow-up, type of radiological examination) are not well established. To our knowledge, characteristics of SFT of the liver on PET scan have been depicted in only two cases of malignant tumor [6–14]: SFT of the liver showed low metabolic activity [14] or hypometabolism consistent with necrosis within the liver lesion [6], but no hypermetabolic activity. It seems thus that PET scan is not adequate for screening of SFT recurrence after surgical resection. In our case, the PET scan performed 6 months before the diagnosis of cervical metastasis showed no suspect hypermetabolism. However, we could not assess if the cervical tumor was already present at that time with no fixation, or if it was absent with a rapid progression thereafter.

Our patient received regular follow-up comprising CT scan of the thorax, abdomen and pelvis every 6 to 12 months. Retrospectively, this attitude seems inadequate because the cervical metastasis was diagnosed at the symptomatic stage. A whole-body CT scan would have been more appropriate to detect the metastasis at an earlier stage. Finally, the highly rapid progression of the pelvic tumor of our patient (no tumor on December 2007 and a large tumor 4 months later) suggests that close follow-up is required in cases of tumor with increased malignant potential. ESMO recommendations for the follow-up after surgical treatment of soft tissue sarcomas are the following [15]: intermediate-high grade patients may be followed for local relapse and metastases every 3–4 months in the first 2–3 years, then twice a year up to the fifth year, and once a year thereafter; whereas low-grade sarcoma patients may be followed for local relapse every 4–6 months in the first 3–5 years, then yearly. The best method for follow-up remains not well established. According to our case report and also literature data, a proposition could be a follow-up with whole body CT scan for high-risk patients, and with conventional hepatic ultrasound for low-risk patients.

Adjuvant chemotherapy or radiotherapy have not been proposed to our patient after the surgical resections. In fact, there are no recommendations and very few data in the literature about adjuvant therapy after surgical resection of SFT [12]. ESMO recommendations for soft tissue sarcomas are the following: adjuvant radiation therapy is recommended for intermediate-high grade, deep and greater than 5 cm diameter soft tissue sarcomas in limited disease; adjuvant chemotherapy can be proposed but its use remains debatable and trials are conflicting [15]. Finally, indication of adjuvant therapy after surgical resection of a SFT of the liver has to be debate by an interdisciplinary staff of a reference oncology center.

Conflict of interest statement
No.

References


