Solitary Fibrous Liver Tumor: Is Surgical Approach the Best Option?

Paula Novais¹, Carlos Robles-Medranda¹, Vera Lucia Pannain², Daniel Barbosa³, Beatriz Bicca², Homero Fogaça¹

1) Gastroenterology Division; 2) Department of Pathology; 3) Surgery Division, Clementino Fraga Filho Hospital, Federal University of Rio de Janeiro, Brazil

Abstract

Solitary fibrous tumor of the liver is a rare tumor, where the evolution, malignant potential, and invasive growth have not been well defined. Although most cases are benign, there is no strict correlation between histological findings and biological behavior, and follow-up surveillance is necessary. We present the case of a large solitary hepatic fibrous tumor and its clinical outcome after a 4-year follow-up. Additional therapeutic options are also reviewed. The surgical resection is a plausible therapy in large solitary fibrous tumors of the liver, although liver transplantation may be discussed when the tumor is considered unresectable. However, such aggressive approaches are questionable in asymptomatic patients due to the natural history of this tumor, which is not well known, and the risk of complications.

Keywords

Solitary fibrous tumor – liver tumor – liver transplantation – CT scan – MRI

Introduction

Solitary fibrous tumor (SFT) is a mesenchymal neoplasia, which frequently occurs in pleura. It has also been described in other sites including the liver (mainly in the right lobe), orbits, superior respiratory tract, abdomen, breast and soft tissue [1].

The hepatic location is extremely rare, with only approximately 30 cases described in the literature [2-4]. The symptoms are commonly related to the abdominal mass, when the tumor reaches a large size. The diagnosis can be preliminarily deduced from the clinical history as well as from imaging examination. However, it can only be confirmed by histopathological and immunohistochemical studies. The recommended treatment is surgical resection [5], mainly because of the intrinsic potential of malignancy that may be misdiagnosed in needle-sample biopsies [6].

We report the case of a large hepatic SFT that initially appeared to be an unresectable mass, and we describe the clinical course with a 4-year follow-up. We also discuss the therapeutic options in cases where the possibility of malignancy is present.

Case report

A 34-year-old female, with no past medical history, was initially admitted to our hospital in May 2001 with abdominal pain and an increase of abdominal circumference. There was no previous history of alcohol, drugs or medication intake. Upon physical examination, the liver was 11 cm below the lower edge of the ribs. There was no ascites, edema, hepatic failure signs, or cirrhosis stigmata. Laboratory tests including alpha fetoprotein were within the normal range. Serum viral markers were all negative and no esophageal varices were found upon an upper endoscopy study.

Abdominal ultrasound with Doppler fluxometry revealed a large heterogeneous hypoechoic mass of 20 x 16 cm limited to the right hepatic lobe; the procedure was considered technically difficult. The surgical team at that moment found the tumor to be unresectable. A right hepatic arterial ligature was performed to decrease the tumor size. The biopsy of the tumor showed lesions associated with SFT: a few neoplastic spindle cells
Fig 1. CT scan of solitary fibrous tumor. A: CT scan without contrast. B: Contrast enhancement in arterial phase, large heterogeneous circumscribed mass in the right hepatic lobe and left medial segment.

Fig 2. Histological examination: A. Collagen bundles with few spindle cells (H-E x 200); B. CD34 immunostaining showing diffuse reactivity.

Fig 3. MRI in T1 with hypointense areas, suggesting cystic degeneration and necrosis in the SFT.

with uniform and elongated nuclei without atypia and separated by abundant thick collagen bands with capillaries (Fig. 2A).

The clinical course and the tumor size did not change in the following 11 months, after which another tumor biopsy was performed and showed the same findings as before. The immunohistochemistry study revealed CD34 (Fig. 2B) and vimentin antibody positivity in the neoplastic cells, confirming SFT.

There were no other therapeutic options and, due to the large tumor size with malignant potential, the patient was listed for liver transplantation. Two years later, a CT-scan and magnetic resonance imaging (MRI) did not show any change in the tumor size, although small multiple hypodense areas were seen in the MRI, suggesting cystic degeneration and necrosis (Fig. 3).

In the following year, as the patient was clinically stable, without alteration in her liver function, and as the risk of malignant transformation is not well defined in the literature, another attempt of surgical resection was made prior to liver transplantation. The remnant liver volume at CT-scan was estimated to be 32%, and finally a right hepatic lobectomy including segments IV a, b; V; VI; VII and VII was performed successfully after vascular exclusion of the liver. The postoperative course was complicated by an infected hematoma, which was controlled by antibiotic therapy and intensive care support for 21 days.

The patient was discharged 40 days after surgery in excellent clinical condition.

The tumor weight was 3,700 g and size 25 x 23 x 13cm (Fig. 4). The tumor was encapsulated and had a gray-white surface with whorled and myxoid areas. Microscopic findings showed acellular areas with hyaline collagen bands and a few spindle cells with fusiform nuclei and minimal cytoplasm. No cellular atypia and no mitotic activity were found. The cells were immunoreactive for vimentin and CD34, and negative for cytokeratin, S-100 protein, and CD-117 antibodies.

Currently, two years after surgery, the patient is asymptomatic and has normal liver function and no signs of local or distant recurrence.
Solitary fibrous liver tumor

Discussion

The hepatic SFT is extremely uncommon [8], with a varied presentation. The patient may be asymptomatic or present with abdominal pain, increased abdominal volume, hypoglycemia [9], or alterations of liver tests and compression of biliary ducts leading to cholestasis [7]. In the differential diagnosis it is necessary to exclude the possibility of hepatocellular carcinoma, sarcoma, leiomyoma, and inflammatory myofibroblastic tumor. The radiological findings are unspecific, and cannot distinguish between benign and malignant tumors [1, 7]. The lesion is usually a solitary mass, highly vascular, well circumscribed, encapsulated, and showing heterogeneous enhancement (due to the varied density from the collagen component) in CT and MRI images [7]. Histopathological and immunohistochemical studies are necessary for correct diagnosis. The CD34 antibody is a marker for SFT diagnosis, although it is not specific [10] and can also be positive in angiosarcomas and gastrointestinal stromal tumors [7]. However, the clinical course and more specific immunostainings as the anti-CD31 in cases of angiosarcomas, and the C-kit stain CD117 for GIST are useful and offer a definitive diagnosis.

The main concern in such cases is that SFT can develop a malignant transformation [11]. It is possible that malignant and benign areas are present in the same lesion. The hypercellularity, nuclear atypia, necrosis and high mitotic activity are considered criteria for malignancy [1, 7]. Larger tumors (> 10 cm) are correlated with lower disease-free metastasis survival [12]. Necrotic areas should alert the physician to the possibility of atypical features or malignant transformation [7, 12], and biopsy may not identify a malignant component in the sample taken.

Surgical resection is the recommended treatment [6], although high morbidity and mortality have been described in large lesions which require extensive resections [5]. There are no data about liver transplantation in patients with SFT. However, unresectable rare non-hepatocellular or bile duct tumors that arise within the hepatic parenchyma are a formal indication of liver transplantation [13]. Therefore, this approach was proposed to our patient, and could be considered as a therapeutic option. In these cases, when patients are in excellent condition, living donor liver transplantation could be indicated.

In our case, it was initially found that the lesion was unresectable, so ligature of the right hepatic artery was attempted in order to reduce the tumor size. The ligature of the hepatic artery, used in other liver tumors, arises as an option to generate hypotrophy of the tumor, allowing a better resection [14]. However, this approach did not succeed and probably it is not a good option for SFT. As the patient remained stable for 4 years with a normal hepatic function and without additional tumor growth, a new resection was attempted to avoid liver transplantation. The resection was successful and the lesion was completely removed.

There was a good surgical response and no malignancy was found on pathologic examination. However, a life threatening surgical complication occurred as a result of such extensive surgery. The question remains whether such aggressive treatments are worthwhile in asymptomatic patients with excellent liver condition where the real risk of malignancy is unknown. Due to the rarity of this tumor, the prognosis has not been well defined [15].

Conclusions

Surgical resection is a plausible therapeutic option in large solitary fibrous tumors of the liver and transplantation can be considered when the tumor is non-resectable. However, such aggressive approaches are questionable in asymptomatic patients due to the natural history of this tumor, which is not well known, and the risk of other complications.

References