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# CASE REPORT: HEPATIC FUSOCELLULAR SARCOMA

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## ABSTRACT

**Primary hepatic sarcoma is a rare malignant neoplasm from mesenchymal cells and it is unusual issue in national and international literature. The majority of the cases are asymptomatic or oligosymptomatic. In the imaging tests, there are no specific features for this type of tumor, the main tumor markers do not increase and there is no association with viral hepatitis. The diagnosis performed by histology and immunohistochemistry. Therefore, this article aims to report a clinical/ surgical case of primary hepatic sarcoma. For this, it was performed a retrospective descriptive study along with the general surgery team of a public hospital in Belo Horizonte in 2014. This is a patient with hepatic fusocellular sarcoma, affecting the left lobe of the liver. After multidisciplinary assessment, it was decided for a hepatectomy in three liver´s segments. The patient was discharged with satisfactory clinical/ surgical outcome remained asymptomatic and no signs of disease recurrence after nine months of follow-up. In conclusion, the surgical resection is the mainstay of treatment of primary liver sarcoma and the main determinant of prognosis and tumor recurrence after initial workup.**

**KEYWORDS:** Sarcoma, liver neoplasms, hepatectomy.

## 1. INTRODUCTION

Sarcomas are rare malignancies source of mesenchymal cells and therefore presents a heterogeneous histological pattern[1,2]. These tumors account for less 1% of all malignant neoplasms of the adult, being more common in children and adolescents whose rate is approximately 10% of the tumors. At aged between children and adolescents, there is evidence of a high rate of mortality and local recurrence and distant metastasis[1,3]. However, primary liver sarcomas represent only 0.1 % to 2 % of liver tumors. Considering all the hepatic sarcomas, most comes from metastasis and only a small portion corresponds to the primary sarcoma of the liver[4]. In adults, 95 % of primary liver tumors correspond to Hepatocellular Carcinoma (HCC) is the fifth most common solid tumor in the world[3].

Most Soft Tissue Sarcomas (STS) arises from the extremities (60 % of cases), most commonly the lower. However, they may also develop in the thorax (30 % of cases) and in the head, neck and other sites (10 %)[2].

Surgical resection R0 with minimum margin of one centimeter corresponds to the treatment of choice, the only method that can be cured. The prognosis is related to histological type, degree of differentiation and tumor involvement[4].

The aim of this study was to report the case of a patient with spindle cell sarcoma and liver conducting a review of the literature on this topic so scarce in scientific circles.

## 2. CASE REPORT

G.V.P., female, 70 years old, Ethnicity Caucasian, sought medical care from general surgery in January 2014, complaining of chronic diarrhea. The consultation, had an abdominal ultrasound performed on 10/18/2013, which showed increased volume of liver due to a massive oval solid mass, heterogeneous, vascularized, located in the transition of IVb and V segments with exophytic component, projecting inferiorly, measuring about 14.8 X 13.4 X 8.4 cm, shown in Figure 1.

The patient also presented a computed tomography of the abdomen (11/13/2013), which showed increased liver size in the left lobe, with homogeneous parenchyma. It was observed a large expansive lesion, well outlined, lobulated contour, hypodense with heterogeneous enhancement, after contrast medium and trend homogenization in the later stages. The lesion, located in segments II and IVb, has measured about 14.3 X 14.5 X 8.5

cm and extending the hepatic limits with lower compression and displacement of the transverse colon, reaching out to touch the greater gastric curvature according to Figure 2.

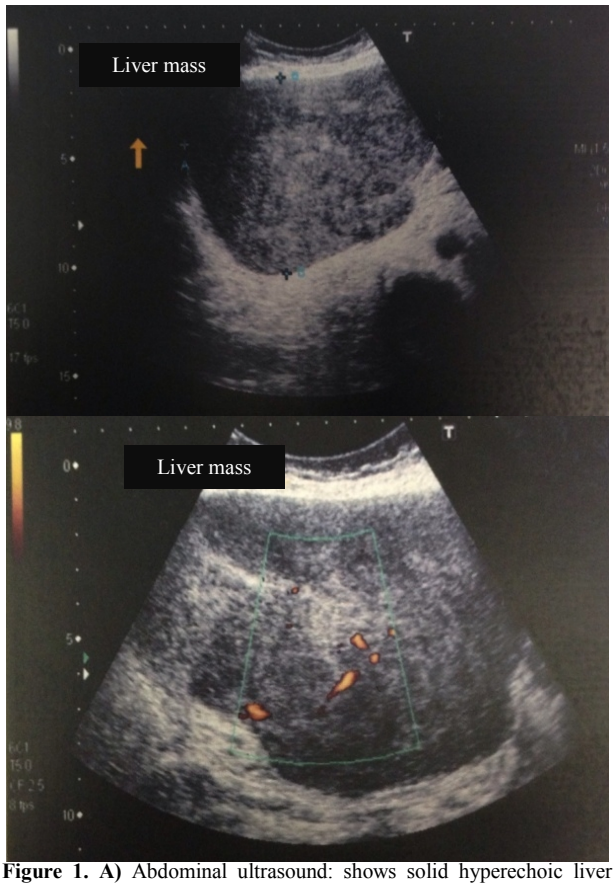


**Figure 1. A)** Abdominal ultrasound: shows solid hyperechoic liver tumor. **B)** Abdominal ultrasound: vascularized nodule on Doppler.

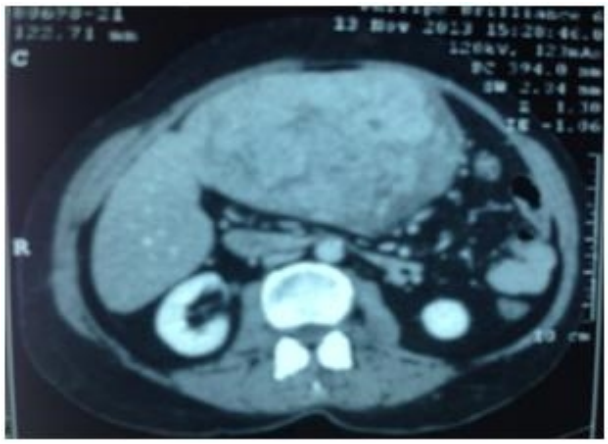

**Figura 2.** Abdominal tomography: presence of hepatic mass contoured little enclosed in right lobe with heterogeneous enhancement of intravenous contrast.

Furthermore, had the following tumor markers 11/27/2013: CA19.9 IU/ mL 11.1; carcinoembryonic antigen (CEA) of 0.68 ng/ mL and alpha-fetoprotein 3.2 ng/ mL. A hypothetical diagnosis of hepatocellular carcinoma (HCC), which was indicated trisegmentectomy, which was held on 01/10/2014.

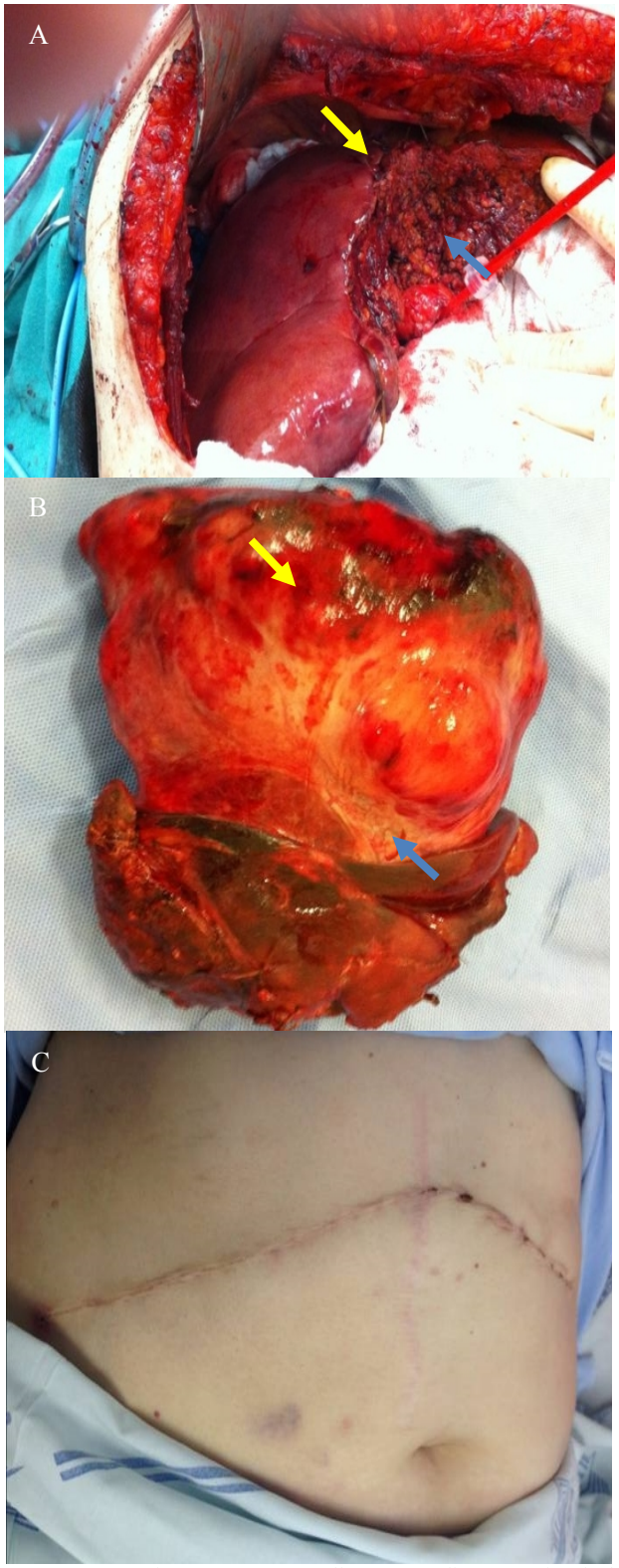


**Figure 3. A)** trisegmentectomy: Yellow arrow: liver resection bed; blue arrow: Pringle maneuver; **B)** anatomical resection part: yellow arrow: tumor; blue arrow: liver bed; **C)** surgical incision scar.

The surgery was uneventful and the patient was transferred to the intensive care unit for postoperative care. The material was sent to the pathological examination showed that constituted neoplasia fusocellular cells with moderate atypia and few mitoses. The cytology of ascetic fluid was negative for atypical cells and immunohistochemistry demonstrated expression to just smooth muscle actin-positive in rare cells for cytokeratin, no necrosis and presence of three five mitotic figures per high-power field, which favored the diagnosis of spindle cell sarcoma of low grade indefinite histogenesis. The patient remained stable and was discharged on the 5th postoperative day (POD) to the infirmary, evolving with deep vein thrombosis (DVT) and pulmonary embolism (PE) in 9 POD (Figure 4). She was treated for this complication and the patient was discharged on postoperative day 21st for outpatient follow.

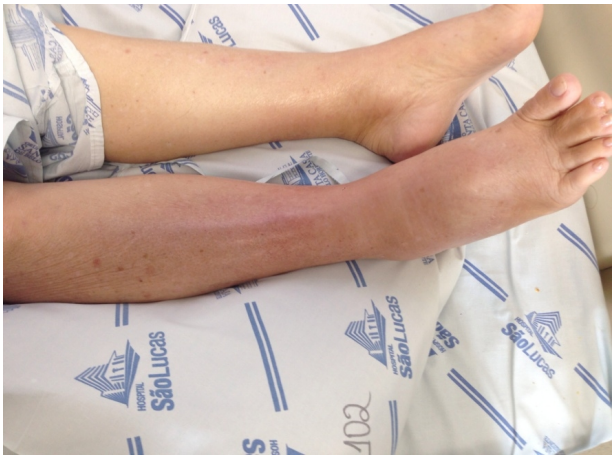


**Figure 4.** Patient with edema and hyperemia of the right leg.

## 3. DISCUSSION

Soft Tissue Sarcomas (STS) are rare malignancies originating from mesenchymal tissues resulting from neoplastic transformation of structures derived from the mesoderm and neuroectoderm[1].

The World Health Organization (WHO) defines more than 50 STS subtypes besides the bones, being fundamental the correct classification of these neoplasms, since they have different biological behaviors and differences in the response to therapeutic modalities[1]. However, there is a difficulty in making a correct diagnosis of mesenchymal tumors in the absence of immunohistochemical markers specific to each histological subgroup. The same material sent to pathologists can generate a diagnostic disparity around 25%[1].

Some of the most common histological types of STS are: angiosarcoma, embryonal sarcoma, leiomyosarcoma, epithelioid hemangioendothelioma, fibrosarcoma, rhabdomyosarcoma, and malignant fibrous histiossarcoma[5]. In this study, the histological subtype was compatible with hepatic leiomyosarcoma.

As in the report described case, the primary hepatic sarcoma is more common in females (2:1) and after the fifth decade of life. In addition, most patients are asymptomatic at diagnosis, which is usually random and routine tests such as abdominal ultrasonography[6]. When symptoms occur, are nonspecific as abdominal discomfort or pain, weight loss, or chills[7,8,9,10].

As opposed to the STS, which are associated with alcoholism, cirrhosis and chronic hepatitis, hepatic sarcomas etiology and risk factors remain unclear in most patients[11,12]. In agreement with the literature, the patient had tumor markers (carcinoembryonic antigen- CEA, alpha-fetoprotein and Ca19.9) negative both preoperatively and postoperatively, which contradict with the main differential diagnosis of primary liver tumor is STS (70% alpha-fetoprotein high diagnosis)[3].

The vast majority of patients with primary hepatic sarcoma presents negative serology for viral hepatitis, as well as in the case report. In imaging studies, no specific features for this type of cancer, however, the most common presentation is the solitary intrahepatic tumor with a diameter of major axis greater than 5 cm, which agrees with the present study[4]. Despite advances in of imaging methods, the histopathology and immunohistochemistry are still the gold standard for diagnosing the disease. The use of percutaneous biopsy is still debatable because a negative sample does not exclude malignancy and there is still risk of injury of vessels and severe biliary tract and tumor dissemination. Owing to this consideration, currently, it is recommended to tumor resection guided by intraoperative ultrasound, and, in cases of resection of contraindication, biopsy should be performed by laparoscopy or conventional surgery[4].

Response to the rarity of primary hepatic sarcoma and the low number of reports in the literature, currently there is no standard therapy. However, studies indicate that the main form of treatment is surgical resection with minimum margin of tumor involvement of free 1cm (R0) and similar to other hepatic malignancies. Currently, the morbidity and mortality of liver resection has been decreasing due to the emergence of new surgical techniques and a broader understanding of the perioperative management. There are few cases of treatment for the tumor enucleation (R1), but with high local recurrence rate. Liver transplantation, used to treat other liver tumors, is still uncertain as treatment in primary hepatic sarcoma with high rates of metastasis. The patient whose case was reported here, for having made a resectable tumor, normal liver function and good surgical status, was chosen to carry out the hepatectomy trisegmentectomy type R0[7,8,9,10].

Still has no standard treatment with chemotherapy and/ or radiotherapy neoadjuvant and/ or adjuvant. Some studies suggest treatment with adjuvant chemotherapy

for poorly differentiated tumors, metastases and tumor recurrence with use of doxorribicin, ciclosfamide of etoposide phosphate and orifosfamide because it seems slow the course of the disease. With regard to radiotherapy, primary hepatic sarcomas seem to be radioresistant[13]. Regarding the described case, the patient was referred to radiotherapy service and clinical oncology and due to the histological results of resection with clear margins and it is a low-grade tumor; it was decided to clinical follow-up, not requiring treatment adjuvant.

The local recurrence, distant metastasis and survival are related to the histological type, degree of differentiation and the presence of surgical excision margin involvement. Most metastases occurs in the first two post-surgical resection years and represents the major site the lung. The median survival for patients who underwent R0 resection was 77% at 5 years. Moreover, patients undergoing resection R1 have mortality rates of around 100% at 3 years[14,15,16]. The patient in question is the 9th month of monitoring and asymptomatic without evidence of tumor recurrence.

## 4. CONCLUSION

The fusocellular sarcoma liver is an uncommon subtype with few cases described in the literature. The experiences of isolated cases in different centers is important to improve surgical techniques, evaluate the benefits of neoadjuvant therapy and/ or adjuvant chemotherapy and/ or radiotherapy and propose more effective treatment regimens. Surgical resection R0 is the mainstay of treatment, the main determinant of tumor recurrence and prognosis.

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