A giant solitary fibrous tumour of the liver: a case report

A Giant Solitary Fibrous Tumour of the Liver: A Case Report.

ROMAN Jan [1,2], VÁVRA Petr [1,2], VÁVROVÁ Michaela [3], ŽIDLÍK Vladimír [4], PELIKÁN Anton [1,2,5,6]

[1] Faculty of Medicine, University of Ostrava, The Czech Republic
[2] Department of Surgery, University Hospital Ostrava, The Czech Republic
[3] Department of Nuclear Medicine, University Hospital Ostrava, The Czech Republic
[4] Department of Pathology, University Hospital Ostrava, The Czech Republic
[5] Department of Surgery, St. Mary´s Hospital, Newport Isle of Wight, The United Kingdom
[6] Department of Health Care Sciences, Faculty of Humanities, Tomas Bata University in Zlín, The Czech Republic

ABSTRACT

Introduction

Solitary fibrous tumour of the liver is a rare mesenchymal tumour, occurring usually in women and with various symptomatology. The symptoms mostly result from pressure of the tumour mass on surrounding organs. Due to unknown biological behaviour and gradual increase of tumour volume, surgical resection is mostly the preferred treatment option.

Case

A 75 years old woman with a history of endometrial cancer, presenting with an incidental finding of liver mass, initially considered of infectious origin (either echinococcosis or cysticercosis). Further diagnostic did not clarify the aetiology, a surgical revision was rejected at the time. The subsequent follow up was interrupted by the development of symptoms of gastrointestinal and renal obstruction, which led to a complete surgical removal of the tumour, sized 30x25x20 cm. A histopathological examination showed a *CD34* and *STAT6* positivity, leading to a diagnosis of a giant solitary fibrous tumour of the liver. The patient recovered well, without any signs of recurrence.

Conclusions

The solitary fibrous tumour of the liver is an exceedingly rare tumour of indeterminate malignant potential, without any specific radiological findings. A complete resection is the treatment of choice, followed by thorough follow up.

MESH Keywords: liver neoplasms - case reports - biopsy - surgical oncology - general surgery

Corresponding author:
Jan Roman, MD
Department of Surgery, University Hospital Ostrava
17. listopadu 1790/5, 708 52 Ostrava, The Czech Republic
mail@janroman.cz, tel. 00420 737 376 178

INTRODUCTION

Solitary fibrous tumour (SFT) is a rare tumour of mesenchymal origin. The most common synonyms are fibrous mesothelioma, benign fibrous mesothelioma, localized fibroma, or pleural fibroma [1]. It most often occurs on the pleura (where it was first described), or in the mediastinum, thyroid gland, or peritoneum. The primary localization of the tumour in the liver is rare, with the first literature description of such tumour being from 1959 [2]. It occurs predominantly in women (70 % of patients are women), without age predominance. Symptoms are variable and usually depend on the size of the tumour [3]. They may include abdominal pain, bloating, dyspepsia, cholestasis, hydronephrosis, weight loss and even hypoglycaemia. The diagnosis is often difficult, and a surgical resection is diagnostic and therapeutic option of treatment. The biological behaviour of these tumours is mostly benign. The risk of aggressive behaviour increases with the age of patients, large tumour volume, high cellularity, atypical cells, and high mitotic activity.

CASE

The patient M. J. is a 75 years old Caucasian woman, obese (BMI 31.22), with essential hypertension and asymptomatic multiple cholecystolithiasis. She underwent a hysterectomy with bilateral adnexectomy in year 1995 for endometrial cancer followed by adjuvant radiotherapy of the lesser pelvis. No other relevant information was found in family or epidemiological history, aside from smoking 10 cigarettes per day for 20 years in her young years.

During regular gynaecological follow-up due to the endometrial cancer, a solitary lesion in the liver was found in April 2016. The initial diagnostics were performed in a different hospital. Initially, based on the radiological description, the lesion was suspected as alveolar echinococcosis or cysticercosis. To rule out dissemination of the process, staging posteroanterior view of the thorax, computed tomography (CT)*(Figure 1A)* of the abdomen and magnetic resonance imaging (MRI)*(Figure 1B)* of the liver were performed, without any signs of dissemination. Blood test showed a slight eosinophilia, the total serum IgE was 100 kU/l. Alpha-fetoprotein (AFP) levels were normal. The patient was administered *mebendazole*. During the initial diagnostic process, the patient was completely asymptomatic. The patient was then transferred to our clinic for further examination.

Due to relatively low antibody levels, the infectious origin of the liver lesion was often questioned. In June 2017, the size of the lesion was 112x106x105 mm *(Figure 1B)*, enlarged since the original MRI. The patient was discussed at a multidisciplinary team meeting. To complete the staging process and evaluate a possible aetiology, a colonoscopy and gastroscopy were recommended. Only the gastroscopy was performed. No stomach lesion was found. The patient was recommended to a surgical revision with a biopsy and an eventual surgical resection. The patient refused proposed treatment. As a following staging procedure, a fluorodeoxyglucose (^{18}F) positron emission tomography – computed tomography (^{18}F-FDG PET/CT) was performed *(Figure 1C)*. The scan showed inhomogeneous region of slight hypermetabolism. The infectious origin was excluded. A CT-navigated biopsy was performed, histologically showing a stromal tumour of an unknown origin. As the lesion was regarded benign and the patient was not showing any symptoms, the “watch and wait” strategy with regular follow-up was established. Chemotherapy was not indicated.

During the follow-up (consisting of clinical check-ups every six months with blood work including tumour markers), there was a slow gradual increase of the lesion's size. At the end of 2019, the patient became symptomatic, presenting with severe dyspepsia and pains following food ingestion. These symptoms greatly limited peroral intake. The lesion was well defined during clinical examination, extending from the right upper quadrant of the abdomen to the left, crossing the medial line. It was homogenous, slightly pendulating, without signs of fixation to surrounding structures. Skin was regularly coloured, with enlarged superficial veins. A current abdominal CT was performed, showing the tumour, now sized 30x25x20 cm, suspected to originate from the ventral portion of the liver (*Figure 1D*). The tumour mass caused a compression of both ureters resulting in grade 2 to 3 obstructive nephropathy bilaterally (treated by stenting both ureters during the preoperative stage). Due to worsened symptoms, a surgical revision was recommended again. In case of inoperability, a tumour biopsy and cytoreductive surgery was considered. This proposition was accepted by the patient.

After preoperative evaluation and preparation (including stenting renal pelvises bilaterally due to the compression of both ureters), a surgical exploration was performed in general anaesthesia. A long right subcostal incision was performed, extending well to the left side. The abdominal cavity was practically filled by the tumour mass. The tumour itself was well defined, greatly vascularised, strongly bleeding after minimal insults. It was evident it is connected to the liver by a wide, flat plane of liver tissue, which was gradually reduced to only the Glisson's capsule on the surface of the tumour. Both falciform and teres hepatis ligaments were dislocated to the right, revealing the tumour to originate from segments S4b and S3. The extent of the tumour forced an extensive liver mobilisation. Initially, a transection of the pedicle parenchyma was attempted, but led to profuse bleeding and utilisation of several haemostatic manoeuvres. Due to the favourable shape of the tumour pedicle, two curved bowel clamps were used to crush and clamp the pedicle parenchyma. With the two clamps in place, the resection was sharply completed, and the tumour was removed from the abdominal cavity, without further bleeding. The liver stump was then sutured over both clamps using a 3/0 polypropylene suture. Because of multiple cholecystolithiasis and a large size of the gall bladder, a cholecystectomy was performed to prevent postoperative cholecystitis and pancreatitis. A surgical drain was introduced into the abdominal cavity and the abdominal wall was sutured. The duration of the surgery was 177 minutes, blood loss was estimated to 700 ml. The total weight of the tumour was 6500 g and the size was 30x25x20 cm *(Figure 2)*.

The postoperative period was without major complications, the patient was extubated, without the need for vasopressor support. Two grams of *cefazolin* were administered every 8 hours, supported by administration of hepatoprotective medication. The laboratory results on the second postoperative day showed no unexpected pathology. The patient was dismissed from the hospital on the 5th postoperative day.

Histologic findings showed a **solitary fibrous tumour (SFT) of unknown biological behaviour**. Microscopically, it was hypercellular, consisting of cells with elongated hyperchromatic nuclei, without atypical signs (*Figures 5 and 6*). The tissue was partially necrotic. There were no signs of increased mitotic activity or invasion of surrounding structures. Immunohistochemical analysis showed strong positivity of *CD34, STAT6, Bcl2, CD99, caldesmon* and focally *calponin*. Other markers (*actin, desmin, S100, CD117, DOG1, CD10, EMA, CKAE1/3* and estrogen) were negative. Proliferation activity in Ki67 was 3%. The

resection line was free of tumorous cells, resulting in a R0 resection of the tumour, gall bladder histology only showed chronic inflammation.

The first follow-up visit took place one month after discharge from the hospital. The patient had recovered well; however, she was unable to increase the amount of food intake by a larger margin, without any reflection on body weight, which remained approximately the same (75 kg, with accounting for tumour weight). This condition was treated with *metoclopramide*. Laboratory parameters did not include any significant pathology including any elevation in tumour markers. No other problems were found during the check-up. The plan for further follow-up includes a clinical check-up with a clinical examination of the abdomen, blood work including tumour markers and abdominal ultrasonography to exclude possible malignant changes.

DISCUSSION

Currently, a total of 85 cases of solitary fibrous tumour of the liver have been described, with variable presentation and treatment methods [4]. Approximately 80 % of patients are asymptomatic in the time of diagnosis, more advanced tumours may present with abdominal pain, malaise, palpable abdominal mass, bloating, dyspepsia, nausea, cholestasis, hydronephrosis, weight loss and many other symptoms usually resulting from the pressure on surrounding organs [1]. In our case, the most problematic was the slowly progressing restriction of solid food intake, resulting in an involuntary weight loss of approximately 10 kilograms from 2017 to 2019.

Radiological diagnosis is difficult as the findings are non-specific. It is also not possible to reliably differentiate between benign and malignant lesions (like hepatocellular carcinoma, sarcoma, haemangioma or inflammatory pseudotumor) due to overlapping features [5]. On ultrasound, SFT typically appears hypoechoic, but may contain heterogenous areas due to myxoid degeneration [6]. A computed tomography scan is more precise, showing a well-defined encapsulated mass with heterogenous enhancement. Calcifications and mass effect are common. Magnetic resonance imaging shows lesions of low to intermediate intensity in T1-weighing and a mix of hyperintense and hypointense areas in T2-sequence. These finding are highly suggestive of SFT, however they are not diagnostic. ^{18}F-FDG PET/CT scans are usually not helpful due to commonly large sizes of these tumours and their low metabolic activity. Due to difficult evaluation of radiological signs, a surgery is often necessary to obtain a sample of tumorous tissue for histologic evaluation.

Various morphological patterns include tumours with storiform growth, with a predominance of epithelioid cells, or forms in which cellular pleomorphisms, increased cellularity, or both predominate. Tumours of this morphological appearance are often described as "atypical" SFTs, but the criteria for malignant SFTs have not yet been clearly established. Tumours with atypical morphological features can therefore be considered "border-line" instead of completely benign lesions [7]. Many possible origins and variable histologic features were responsible for a difficult definitive identification of this tumour type in the past. Currently, immunohistochemical features like CD34 and NAB2-STAT6 positivity allow more precise identification.

Treatment usually consists of surgical resection. A specific approach to surgical intervention is guided by the size and precise location of the tumour. A non-anatomical resection or

anatomical liver resections are reported the most often. A conservative treatment of SFT is not considered adequate as there is a risk of malignant growth of the tumour [4]. Another limitation of conservative approach is a slow growth of the tumour, possibly increasing the difficulty of future surgical interventions. Another option in treatment of SFT is transarterial chemoembolization in case of unresectable tumour. *El-Khouli et al. (2008)* reported good results, but without any decrease in the tumour's volume [8]. There is a lack of available data to further evaluate this treatment method. Administration of radiotherapy or chemotherapy in case of malignant solitary fibrous tumour were not found effective.

CONCLUSIONS

The solitary fibrous tumour of the liver is a rare, often incidental finding. It is considered benign, but malignant growth was also reported. A gradual growth mostly results in pressure on other organs. A surgical resection is the treatment of choice. Transarterial embolization is another treatment possibility. Due to indeterminate malignant potential a regular follow-up is necessary, including tumour markers and imaging methods.

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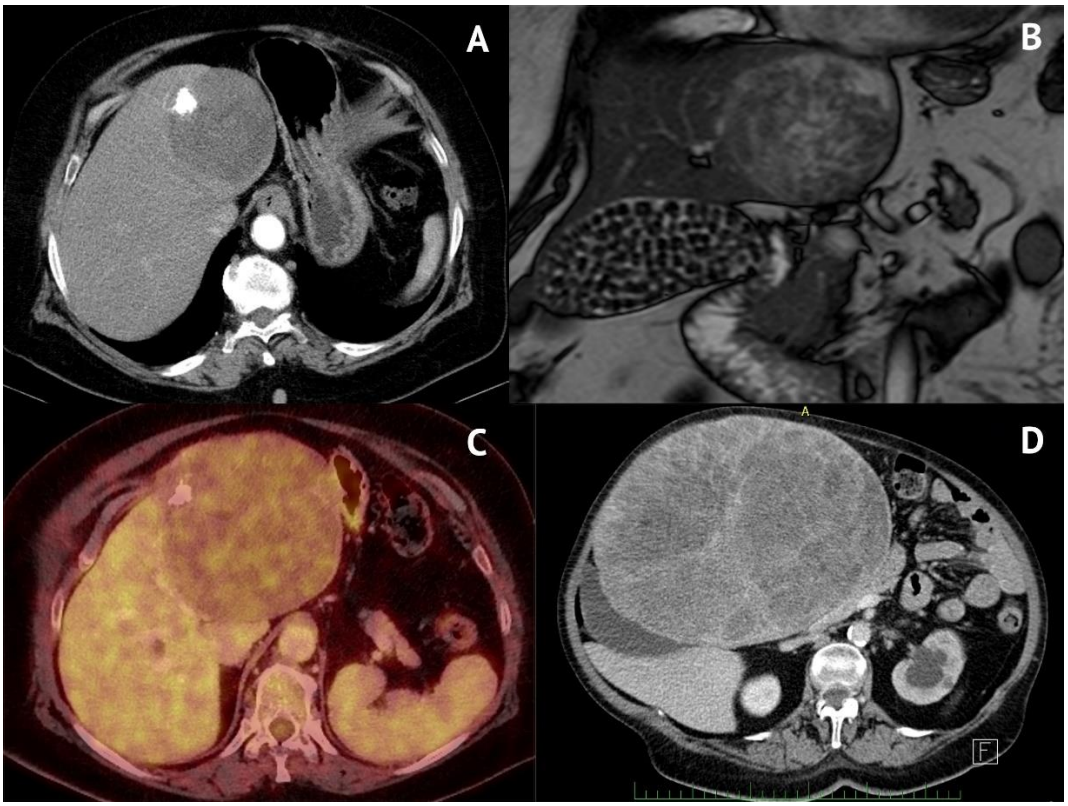

Figure 1 *– Showing the gradual tumour growth during the follow-up.* ***A*** *– an initial axial CT scan, arterial phase, capsulated mass with calcification, March 2016. Largest diameter 9 cm.* ***B*** *– an MRI native scan, T2 weighed, coronal plane, heterogenous mass, multiple cholecystolithiasis, August 2016. Largest diameter 10 cm.* ***C*** *– a ^{18}F-FDG PET/CT scan, transversal plane, mild deposit of ^{18}F-FDG, August 2017. Largest diameter 13 cm. By courtesy of Buriánková, MD, FNOL Olomouc.* ***D*** *– a preoperative axial CT scan, portal phase, transversal plane, December 2019. Largest diameter 30 cm.*

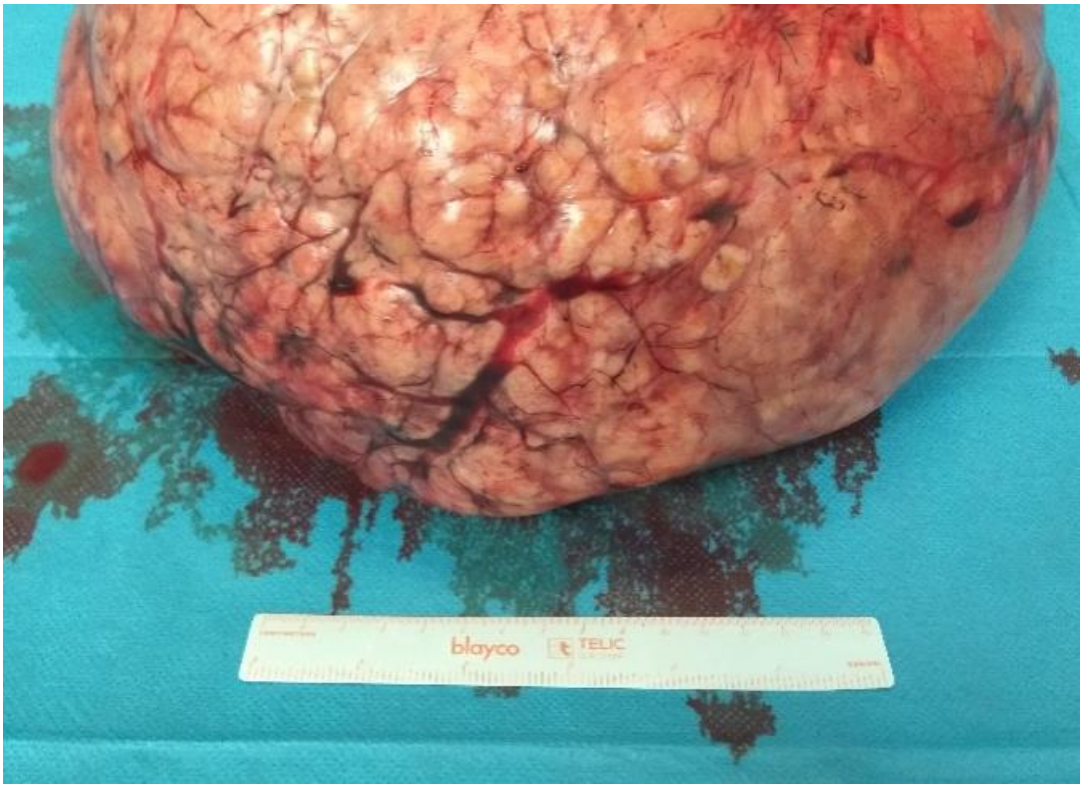

Figure 2 *- Macroscopic detail of the tumour mass, the total size was 30x25x20 cm*

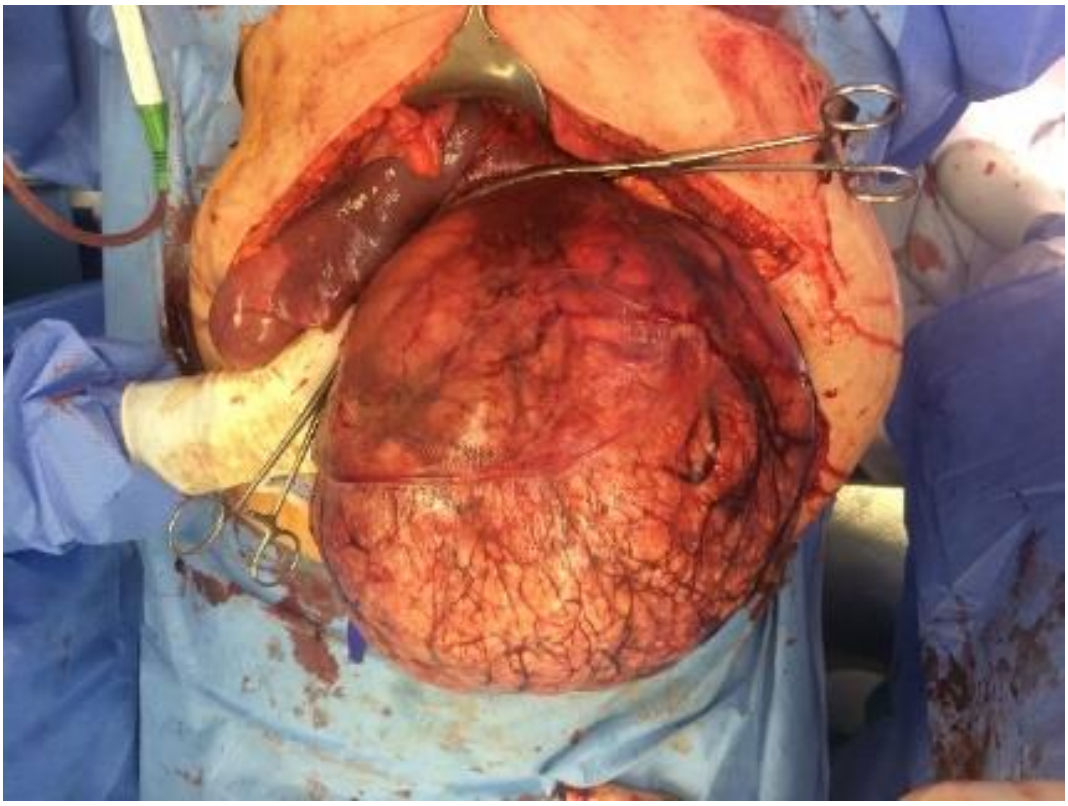

Figure 3 *- Perioperative image showing clamping of the tumour pedicle using two curved bowel clamps before resection.*

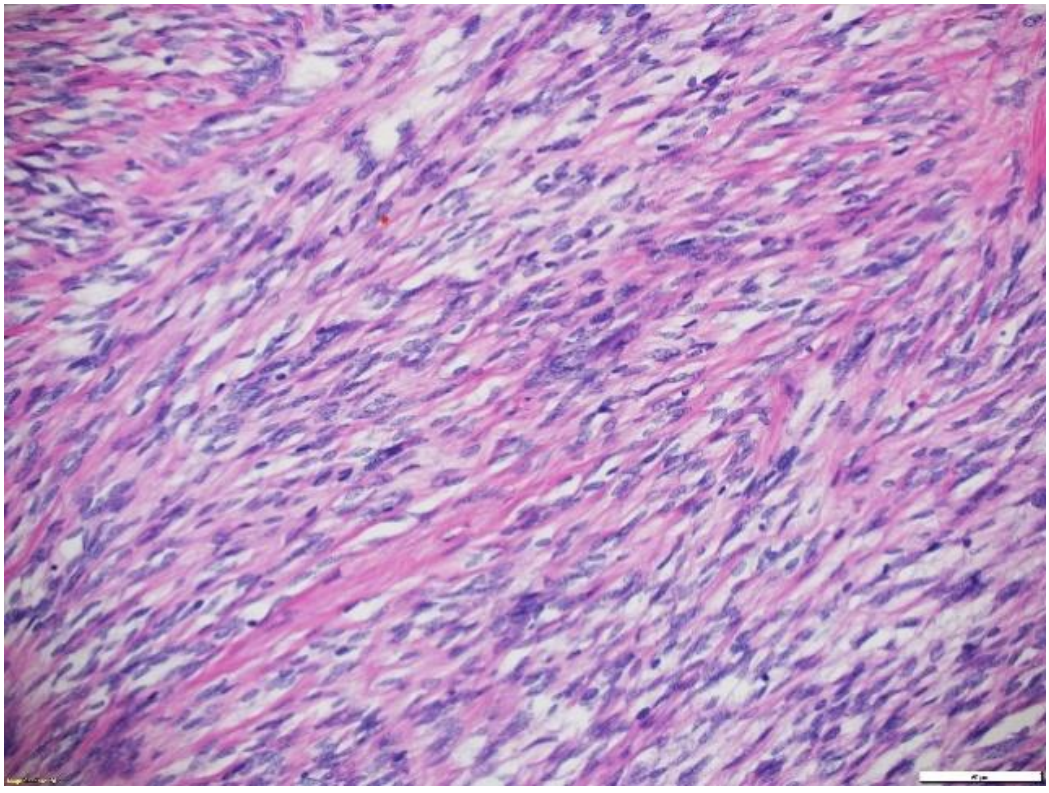

Figure 4 *- Hypercellular tissue of the SFT, cells with elongated hyperchromatic nuclei. 200x magnification, HE stain, Olympus BX63*

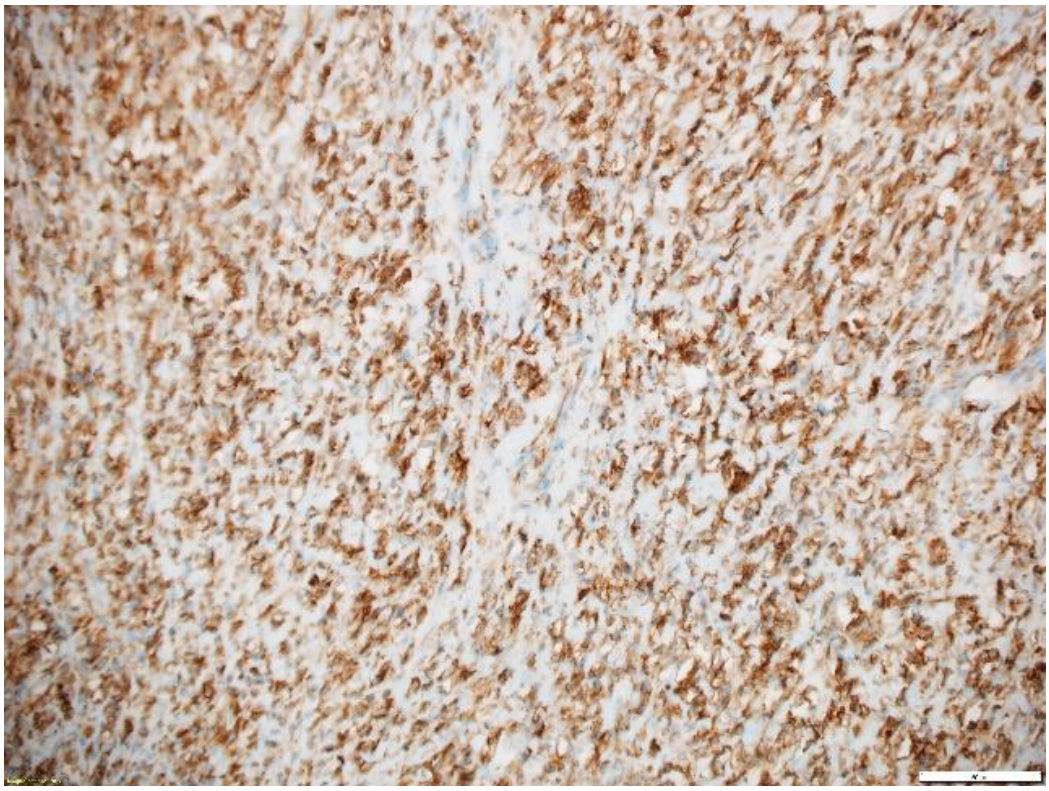

Figure 5 *- Immunohistochemistry - CD34 staining. 200x magnification, Olympus BX63*