Inflammatory Pseudo Tumor-Like Follicular Dendritic Cell Sarcoma of the Liver with Hepatic Lymphoma History: A Case Report and Literature Review

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Case Report

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Abstract

Background: Inflammatory Pseudo Tumor (IPT)-like Follicular Dendritic Cell (FDC) Sarcoma of the liver is a very rare disease and occurs mostly in the lymph nodes. Till now, only 19 cases were reported in the English literature. However, the coexistence of IPT-like Sarcoma and Non-Hodgkin's Lymphoma(NHL) in the same patient has never been reported.

Case presentation: We reported a case of a 47-year-old male patient who had a history of Hepatitis B virus (HBV) infection and presented with right upper abdominal discomfort. Nineteen years ago, he underwent liver resection of segment VII for Hepatic Non-Hodgkin Lymphoma (B-cell lymphoma). Serum alpha fetoprotein (AFP) level was normal. However, Imaging studies revealed a well-circumscribed, solid mass in the right hepatic lobe and was diagnosed as (Hepatocellular Carcinoma) HCC or Hepatic Lymphoma preoperatively. Right hemi-hepatectomy was performed on this patient. Histological report showed features of a mixture of chronic inflammatory cells and variable amounts of spindle cells. Also, Immuno-Histo-Chemical (IHC) studies demonstrated that all the tumor cells showed strong nuclear in situ labeling for EBV-encoded small RNAs (EBER) and strongly positive stainings with CD21 and CD35. The patient is still disease free after a follow-up of 40 months.

Conclusions: To our knowledge, this is the first report demonstrating Hepatic IPT-like FDC Sarcoma in a patient with Primary Hepatic Non-Hodgkin Lymphoma history. In regards to treatment, complete surgical resection should be performed and would acquire excellent long-term outcomes.

Background

Follicular Dendritic Cell (FDC) Sarcoma is a very rare disease with more than half of the cases occurring in lymph nodes. Extra-nodal Follicular Dendritic Cell Tumors mainly arise from intra-abdominal organs such as liver and spleen, which display an aggressive clinical course[1]. Histologically, there are two types of FDC tumor: conventional and inflammatory pseudo-tumor(IPT)-like variant[2].IPT-like FDC sarcoma contains a few spindle cells with prominent lymphocytes and plasma cell infiltration with some neoplastic cells resembling Hodgkin cells[3]. IPT-like FDC Sarcoma of the liver has been rarely reported in recent years, even though it has been receiving increased attention since its first description in 1996 by Selves and his colleagues[4]. Clinico-pathological characteristics of IPT-like FDC sarcoma of the liver have not been fully understood [1, 5]. So far, less than 20 cases of IPT-like FDC sarcomas of the liver have been published in the literature[3, 4, 6–17]. Due to these morphologic features, IPT-like FDC tumors are commonly misdiagnosed as inflammatory lesions and occur almost exclusively in the liver or spleen with a slight female predominance. Recent study indicated that these tumors had a risk of recurrence and metastases[1, 5]. The origin of tumor cells, causes of the disease and the ambiguity of diagnosis are still unknown. IPT-like FDC sarcomas of the liver, which is different from other Extra-Hepatic Follicular Dendritic Cell Tumors, have always been shown to be associated with the Epstein–Barr Virus (EBV). EBV infection is considered as one of the most important etiologies of this tumor. Almost 100% of the IPT-like FDC tumors have always been proven to be associated with the EBV infection[18]. Although IPT-like FDC Sarcoma of the liver is

extremely uncommon, most published cases showed typical histological characteristics, consistent with EBV infection and has a good prognosis after surgical resection of the tumor[1, 19].

The distinction of IPT-like FDC sarcoma from other tumors is very challenging and such tumors are commonly misdiagnosed as reactive lesions. The regression of the tumor mass size, either spontaneously or after treatment with anti-inflammatory agents may increase the likelihood diagnosis of IPTs[20, 21]. The tumor had no enhancement in all three phases after the injection of contrast materials in contrast-enhanced ultrasonography[22]. Central septations, calcification, necrosis or hemorrhage may be present[13]. Some atypical IPT-like FDC sarcomas of the liver may present with arterial enhancement and are hard to differentiate from Hepato-Cellular Carcinomas (HCCs). The definite diagnosis of IPT-like FDC sarcoma of the liver should rely on histopathology. The tumor is a mixture of lymphocytes, storiform and fascicular arrangement of spindle cells which are positive for at least one of the markers for FDCs including CD21, CD23, CD35, Fascin, Clusterin, CXCL13 and epidermal growth factor receptor (EGFR)[23].

Primary Malignant Lymphoma of the liver is also rare. In addition, the occurrence of IPT-like FDC Sarcoma and Non-Hodgkin's Lymphoma (NHL) in the same patient has never been submitted. In this report, we present a case of IPT-like FDC of the liver with Hepatic Non-Hodgkin Lymphoma (NHL) history of which both were successfully resected.

Case Presentation

We evaluated a 47-year-old man with right upper quadrant abdominal pain and no other associated symptoms. He had history of Hepatitis B Virus (HBV) infection and no Hepatitis C infection. Abdominal Magnetic Resonance Imaging (MRI) scan from the First Hospital of Yi Chang revealed a 19x15x13cm tumor in the right lobe of the liver. The right portal and hepatic veins were not visualized, Inferior Vena Cava (IVC) was compressed (Fig. 1A-D). In 1999, this patient underwent hepatic segmentectomy in our hospital due to NHL (B-cell lymphoma) of the liver. He had no fever, anemia, weight loss or constitutional symptoms. Abdominal Ultrasound disclosed a 17.5x14.2x13.6 cm mass in the right lobe of the liver (not available). Although he had history of HBV infection, alpha fetoprotein (AFP), carcinoembryonic antigen (CEA) were all within normal range and other laboratory tests were unremarkable. HCC or Hepatic Sarcoma was suspected. There was no abnormality in any other area of the abdomen. The chest radiograph was within standard limits with a normal size heart. The Indo-Cyanine Green Retention rate at 15 minutes (ICG-R15) was 4.2%. The patient agreed to perform right hepatectomy.The abdomen was carefully inspected during operation and no other lesions were noted within the mesentery, small bowel or spleen. Frozen sections of suspicious lymph nodes were tested negative for tumor.The right lobe of the liver was notably atrophied, likely secondary to tumor thrombosis of the right portal vein, which was showed on MRI imaging. The patient tolerated the surgery well, recovered smoothly and he was discharged on postoperative day seven (day 7).

Surgical specimen was collected for histological examination, processed with standard paraffin technique and stained with routine hematoxylin-eosin (HE) staining procedures. Immuno-Histo-Chemical (IHC) and in situ hybridization analyses were performed on a 4-μm thick section. The patient did not receive any

adjuvant therapy, is currently well and alive. He is asymptomatic with no evidence of recurrence for more than 40 months post operation.

Histological Findings

The excised mass measured 20x18x15cm, which was well delimited from the surrounding parenchyma. The surgical margins were free of tumor, with areas of necrosis and hemorrhage in the center of the tumor (Fig. 2A). Microscopically, the hepatic tumor comprised of spindle-like cells forming a vaguely storiform pattern with blunt cellular border and eosinophilic cytoplasm (Fig. 2B, hematoxylin-eosin stain, x200) and diffuse sheets with the infiltration of small lymphocytes and plasma. The nuclei were oval to elongated with small or inconspicuous nucleoli. The cell boundary was indistinct, mitoses of the tumor cells were negligible.

IHC studies

The submitted tissue was fixed in formalin, embedded in paraffin, and cut into 4 μm slice for further analysis. For IHC studies, antigen retrieval by microwaving in sodium citrate buffer was performed. As a result, spindle-like cells were found to express CD21 (Fig. 2C) and CD35 (Fig. 2D) but not CD23, SMA, Desmin, ALK1 and S-100. Also, no chromosomal abnormality was observed. EBV-encoded nuclear RNA in situ hybridization was performed, showing positive signals on the spindle cells (Fig. 2E) but not on the surrounding lymphocytes. The positive stain rate of Ki67 in spindle cells was about 25% (Fig. 2F). Based on these findings, a Follicular Dendritic Cell Tumor of the Liver was diagnosed. The post-operative period was unremarkable and the patient received regular follow-up after discharge. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. This study was approved to publish the case details by the Ethics Committee of Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan, China.

Discussion And Conclusions

FDC tumors are very rare neoplasms that mainly arise in lymph nodes while one-third of the cases develop in extranodal sites, which have a heterogeneous histology with storiform and fascicular arrangement of plump spindle cells[24]. The FDC tumors gained an increased attention since the first case of a tumor with follicular dendritic cell differentiation was reported by Monda *et al.* in 1986[25]. So far, there are less than 100 cases of follicular dendritic cell tumors which have been reported in English literature[12, 25]. The differential diagnosis include: Sarcoma, Hodgkin's Disease, Leiomyosarcoma, Gastro-Intestinal Stromal Tumor (GIST) and Inflammatory Pseudo-Tumor[8, 26, 27]. According to Immuno-Histo-Chemical analyses, FDC Sarcoma shows spindle tumor cells arranging in fascicular or storiform patterns, which are immuno-reactive for one or more FDC markers: CD21 (C3d receptor, positive in 93% of cases) and CD35 (C3b receptor, positive in 89% of cases). Other quite specific markers used are: R4/23 (63%), Ki-67 (5%-50%), EMA (41%), Vimentin (61%), HLA-DR (57%), CD45 (21%) and S-100 protein (31%). The tumor cells typically lack expression of CD1a, CD 68, and Desmin[12, 14].

Primary Hepatic Sarcomas are very rare, representing less than 0.1% of all Primary Hepatic Tumors[7]. IPT-like FDC Sarcoma of the liver is an extremely uncommon tumor and only 20 cases have been reported till now in English literature, including the present patient[3, 4, 6–17] (summarized in Table 1). In 1996, Shek and colleagues reported the first case of Hepatic FDC Sarcoma[6]. The most common main complaint of the patients was upper abdominal pain and weight loss. Malaise, anemia and fever were also part of the initial presentation in some number of patients. In five cases, patients were completely asymptomatic and the mass was found incidentally using Contrast Tomography (CT) scan and abdominal ultrasound. The age of all the patients at initial presentation ranged from 19 to 82 years (mean 47.8 years), the mean tumor diameter was 11.9 cm (3–20 cm), and the mean reported survival was more than 30 months (follow-up ranging from 6 to 108 months). The histology is similar to that of the conventional FDC tumor and it is generally considered to be a distinctive variant which is characteristically restricted to the abdomen and seems to be a separate clinico-pathologic entity. Comparing to the conventional FDC Sarcoma, it has a marked female predominance (female to male ratio is 4:1), whereas conventional FDC Sarcomas are not more prevalent in only one sex[1]. IPT-like FDC Sarcomas have prominent inflammatory component, which makes it challenging to differentiate them from inflammatory pseudo-tumors. IPT-like FDC Sarcomas are strongly associated with the presence of EBV (85% in our review), which is rare for conventional FDC Sarcomas. Both IPT-like FDC Sarcomas and Conventional FDC Sarcomas generally show an indolent clinical behavior. Nevertheless, Conventional FDC Sarcomas of the liver can be more aggressive than IPT-like FDC ones, may recur or metastasize and even lead to death.

Table 1
Characteristics of patients with hepatic FDC sarcoma of the liver.

Case	Sex	Age (year)	Main complaint	Diameter (cm)	EBV	Treatment	Recurrence/ Survival	Published year
1	F	68	Malaise, weight loss, anemia	11	+	SR	No/>30 m	1996[4]
2	F	35	Epigastric discomfort, fever,weight loss	20	+	SR	Yes/>30 m	1996[6]
3	M	37	Malaise, weight loss, anemia	15	+	SR	No/>24 m	1998[7]
4	F	19	Right upper quadrant pain, weight loss, palpable mass	12	+	SR	No/40 m	2001[3]
5	F	56	Gastrointestinal discomfort	15	+	SR	Yes/NA	2001[3]
6	F	40	Epigastric pain, weight loss	12.5	+	SR	No/108 m	2001[3]
7	F	49	Incidental at ultrasound	4.2	+	SR	No/9 m	2001[3]
8	F	31	Abdominal distention, weight loss	15	+	SR	No/60 m	2001[3]
9	F	57	Epigastric pain, weight loss	9.5	+	SR	No/36 m	2001[8]
10	F	51	Epigastric pain, weight loss	12	+	SR	No/12 m	2001[8]
11	M	82	Incidental on a CT abdomen	15	-	SR	No/18 m	2005[10]
12	F	30	Incidental at ultrasound	5.5	+	SR	No/12 m	2006[9]
13	F	57	Abdominal pain, vomiting, dizziness, liver dysfunction	13	+	SR	No/24 m	2008[14]
14	F	78	Incidental at ultrasound	3	+	TACE	27 m	2010[11]
15	F	59	Asymptomatic	6	+	SR	NA	2010[17]

NA, not available; SR, surgical resection; TACE, Transcatheter Arterial Chemoembolization.

Case	Sex	Age (year)	Main complaint	Diameter (cm)	EBV	Treatment	Recurrence/ Survival	Published year
16	F	53	Right upper quadrant pain, fever, anemia, jaundice	11.5	-	SR	No/6 m	2011[12]
17	M	56	Right upper quadrant abdominal pain	11	NA	SR	No/12 m	2011[16]
18	F	31	Palpable abdominal mass	20	+	SR	NA	2016[13]
19	M	19	Painless swellings around several joints	6	-	SR	NA	2016[15]

NA, not available; SR, surgical resection; TACE, Transcatheter Arterial Chemoembolization.

The initial diagnosis of FDCs is based on clinical examination, imaging and pathologic assessment. The role of imaging is mainly in describing the extent of the mass and staging. When a mass is suspected in the liver, the confirmatory diagnosis of IPT-like FDC Sarcoma is very difficult without pathological findings. It is noteworthy that the diagnosis of IPT-like FDC Sarcoma should be based on the recognition of FDCs from microscopic findings. Actually, the distinction of Hepatic IPT-like FDC Sarcoma from other liver tumors is usually impossible without immuno-histo-chemistry, due to features overlapping with other hepatic malignancies. Honestly, the patient was initial diagnosed as hepatocellular carcinoma (HCC) or Hepatic Non-Hodgkin Lymphoma according to his HBV infection's history, MRI appearance and history of his surgery. Shek *et al.*[6] reported a case of primary IPT-FDC tumor of the liver that was initially misdiagnosed as an inflammatory pseudo-tumor. The tumor recurred 30 months after complete resection[6]. The definite diagnosis of IPT-like Sarcoma of the liver relies on histopathology. Furthermore, CD21 and CD35 have been widely used as the preferred FDC markers which were expressed in almost IPT-like FDCs. Moreover, EBV infection is identified as a key role in the genesis of IPT-like FDC Sarcoma. Almost all IPT-like FDC Sarcomas exhibited positive EBER by in situ hybridization (17/20 in Table 1)[10]. LMP-1 gene, the major oncogene of EBV was identified in several cases of IPT-like FDC Sarcomas[1, 4]. Positive staining of the FDC markers include CD21, CD35 and EBER, while negative expression in CD23, SMA, Desmin, ALK1 and S-100 are in the present case. However, the pathogenic mechanism of EBV in IPT-like FDC Sarcoma remains unclear and further investigation is required.

Primary NHL of the liver has rarely been reported and there are no typical laboratory or image diagnostic findings. Therefore, pathological analysis is the standard diagnostic method[28]. The occurrence of Hepatic NHL and Hepatic IPT-like FDC Sarcoma in the same person has never been previously reported. The patient we reported in this article received hepatic segmentectomy in our hospital due to Non-Hodgkin

Lymphoma (B-cell lymphoma) of the liver in 1999 (no data available due to longevity). Immuno-histochemical studies demonstrated that all the tumor cells were strongly positive stainings with CD20 and CD45. A diagnosis of hepatic NHL was rendered (Supplemental Figs. 1 and 2). Several studies indicated that chronic Hepatitis C Virus (HCV) infection may be associated with the pathogenesis of NHL. However, the present case has HBV infection and no HCV infection. The detailed mechanism of HCV-mediated lymphomagenesis remains unclear[29]. Our search of the literature found no such cases. This is the first report to demonstrate hepatic IPT-like FDC Sarcoma in a patient with a history of Primary Hepatic NHL.

Surgical resection is the treatment of choice for Primary Hepatic IPT-like FDC Sarcomas and Hepatic NHL whenever possible. The efficacy of chemotherapy and radiotherapy is unclear. Daniel *et al*.[30]reported that even if complete resection has been achieved for Hepatic NHL, postoperative chemotherapy is mandatory. The patient did not receive chemotherapy after the first hepatectomy. Tsunemine *et al*.[11]suggested that Trans-Arterial Chemo-Embolization (TACE) was useful for the management of Hepatic FDC Sarcoma and the patient was still alive 27 months after the diagnosis of Hepatic FDC Sarcoma, which was favorably controlled by repeated TACE. Shinagare *et al*.[16]reported a case whereby the patient was not determined to be a surgical candidate because of large tumor mass and small residual liver volume. The patient received four cycles of standard-dose of chemotherapy comprising of Cyclophosphamide, Doxorubicin, Vincristine and Prednisone (CHOP) therapy and portal vein embolization in an attempt to cause hypertrophy of the residual liver which made him a surgical candidate. Seven months later, the patient underwent a successful resection of the 12-cm nodular mass[16]. In our review, nineteen of the patients (95%) with Hepatic IPT-FDC Sarcoma have undergone partial hepatectomy.

In conclusion, we report a unique case of hepatic IPT-like FDC Sarcoma with Hepatic Non-Hodgkin Lymphoma history. IPT-like FDC sarcoma is receiving growing attention and the diagnosis may be correct with the aid of immuno-histochemical analysis being CD 21 and CD35 the most reliable FDC markers. IPT-like FDC Sarcoma should be considered in differential diagnosis when confronted with a liver tumor in a patient with Primary Hepatic Lymphoma or HBV infection history. Complete resection remains the preferred method for the management of Primary Hepatic IPT-like FDCs. In analyzing the reported cases, we brought forth differential diagnosis and provided evidence for further exploration of the pathogenesis of the tumor. Continued accumulation of characteristics of IPT-like FDC Sarcomas of the liver would help in future patient care.

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Figures

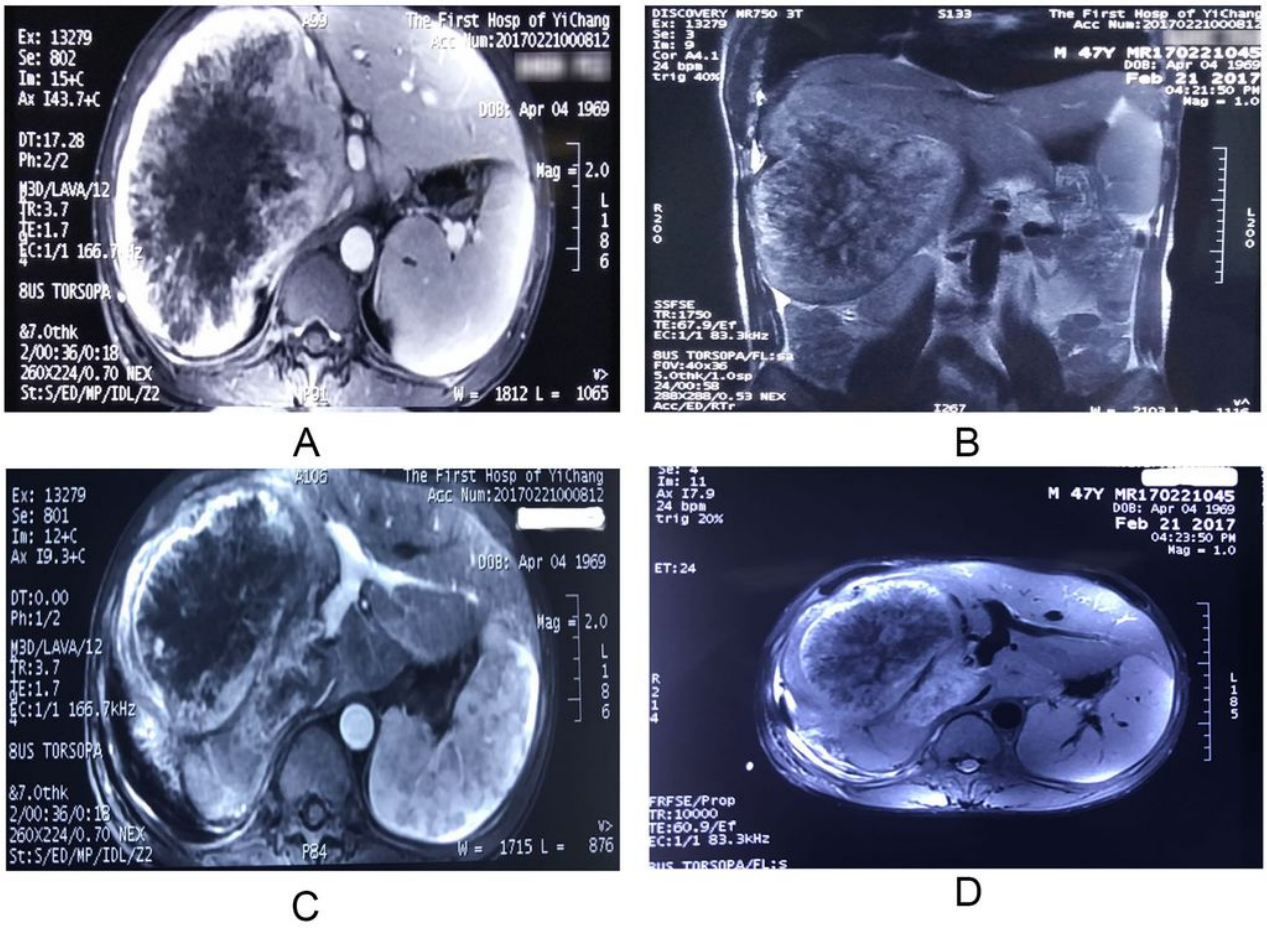

Figure 1

A-B: MRI of the up-abdomen showed a 19x15x13cm lesion occupied most of the right liver. No evidence of other abdominal lesions. A MRI image in the axial plane (A,C-D) and in the coronal plane (B).

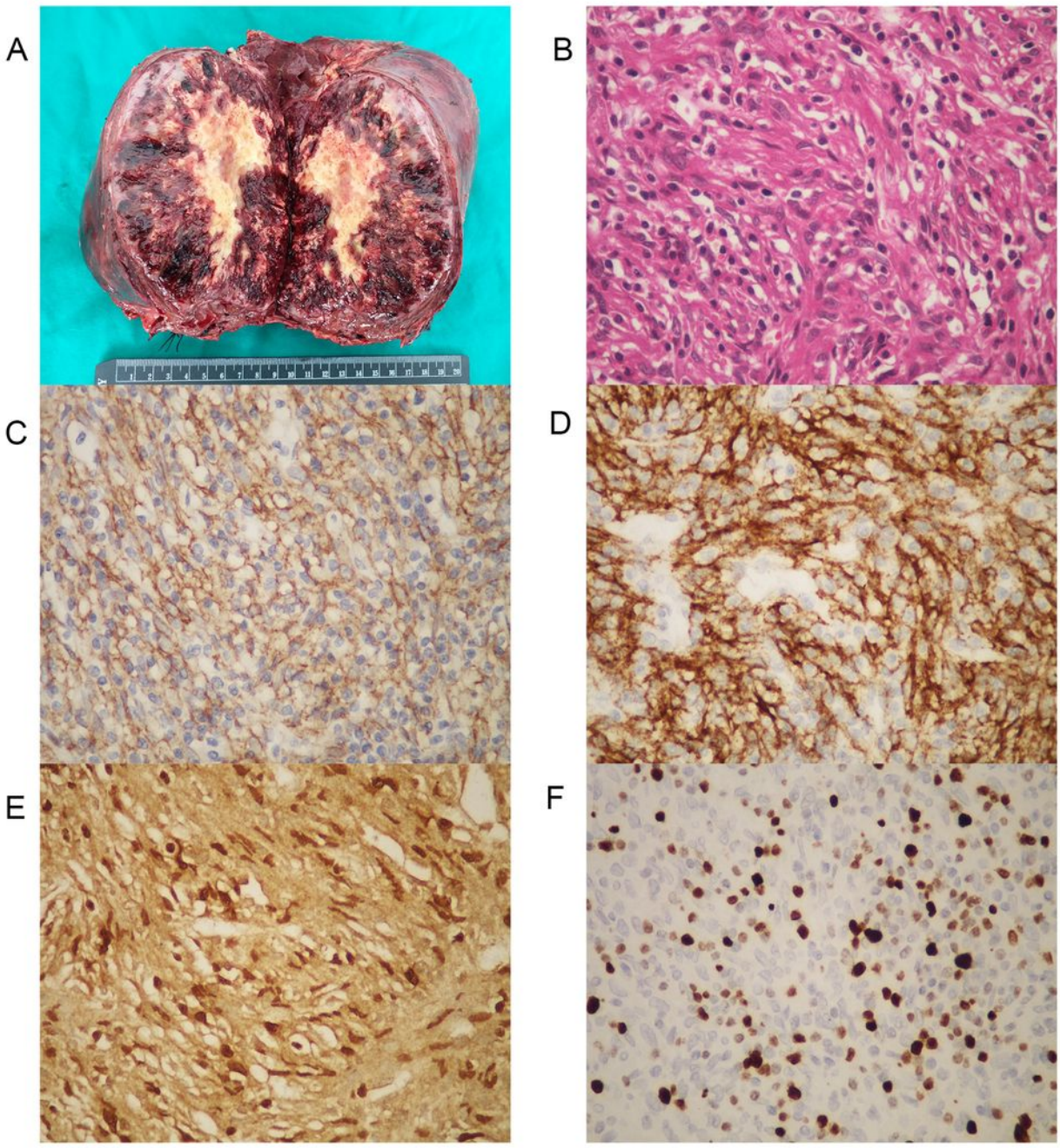

Figure 2

Macroscopic and microscopic findings of the excised tumor. A: The tumor measured 20×18×15 cm and was solid. B: Microscopically, the hepatic tumor comprised spindle-like cells forming a vaguely storiform pattern and diffuse sheets with the infiltration of small lymphocytes and plasma cells (HE). C-D: Immunohistochemical staining for CD21 and CD23, a marker for follicular dendritic cells, showed that spindle-like cells were positive for CD21(C) and CD35 (D). E: Positive stains of EBV-encoded nuclear RNAs

on the most of spindle cells by in situ hybridization. F: One fourth of the tumor cells were positive for Ki67. (Original magnification X400).

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