

## Difficulty in Diagnosis of Leiomyosarcoma of Infrahepatic Inferior Vena Cava

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### Abstract

**Background:** Leiomyosarcomas of inferior vena cava (IVC) are rare tumors that mostly are proposed as a primary malignancy of the IVC. The optimal treatment is completely resects the malignant lesion with preservation of venous return. According to the treated experience of one patient in our hospital, we present our opinions as below.

**Methods and Results:** A 61-year-old woman underwent successful surgical treatment for a leiomyosarcoma with the method of infrahepatic inferior vena cava (IVC). A large tumor that was demonstrated in the Spiegel lobe liver with IVC tumor thrombus was imagined by tomography and magnetic resonance. The tumor was found from IVC, which was performed by suprahepatic and infrahepatic IVC occlusion with Satinskys clamp in the operation. The patient underwent a combined operation which is en bloc resection of the IVC tumor and lobotomy of the left lateral section of liver. Pathological examination confirmed that is primary leiomyosarcoma of the IVC. The patient had a normal live for nearly one year and no recurrence.

**Conclusion:** It is difficult to distinguish leiomyosarcoma from a hepatic tumor. About two thirds of these patients were confirmed as the diagnosis of leiomyosarcomas only after laparotomy. The misdiagnosis to be considered as tumor arising from segment I of the liver with IVC tumor thrombus was lead to the tumor to predominant intra-luminal growth. Radical surgical en bloc resection is the mainly treatment for IVC leiomyosarcomas. Using suprahepatic IVC and infrahepatic IVC occlusion with Satinsky clamp, surgical management of an infrahepatic IVC leiomyosarcoma is a simple vascular surgical techniques.

**Keywords:** Leiomyosarcoma; Inferior vena cava; Sarcoma; Surgery

### Introduction

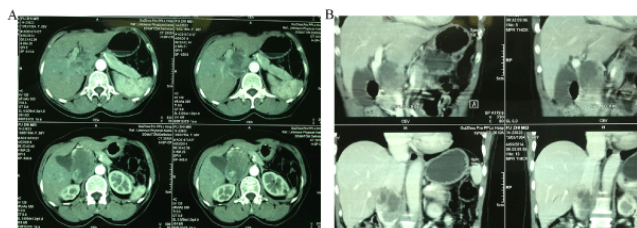
Leiomyosarcomas of the inferior vena cava are rare malignant and slow-growing tumors with a poor prognosis. It was first reported by Perl and Virchow in 1871, in the German literature [1,2]. There were approximately 400 documented cases in the literature, which were originated from the smooth muscle cells of the media and predominantly proposed within the IVC [3-6]. Resection was often presented with a challenge as these tumors may require reconstruction of the inferior vena cava (IVC). These papers reported on a case of surgical resection of an infrahepatic IVC leiomyosarcoma mimicking a hepatic tumor with IVC tumor thrombus.

### Case Report

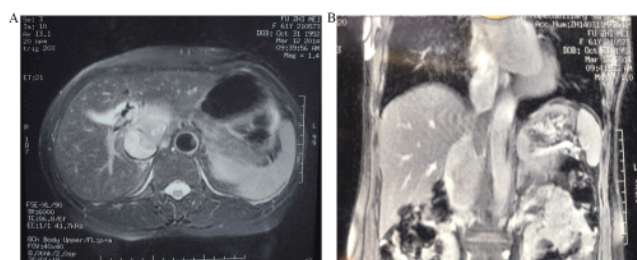
A 61-year-old woman was detected as her right upper abdominal in pain. Laboratory findings included that the total bilirubin of 3.8 mol/L, albumin 39.3 g/L, alanine aminotransferase (ALT) 46 IU/L, aspartate aminotransferase (AST) 98 IU/L, and alkaline phosphatase (ALP) 65 IU/L. She was HBsAg (-), HBV-DNA (-). Her alpha-fetoprotein (AFP), carcinoembryonic antigen(CEA)and carbohydrate antigen 19-9 (CA199) were all within the normal range. The functional status of the liver was assessed as Child A. Enhanced computed tomography of the abdomen revealed a tumor and was detected in the Spiegel of the liver with 7×6×5 cm mass. Areas of hemorrhage and necrosis may be noted within the mass on CT (Figure 1). Magnetic resonance imaging (MRI) of this tumor revealed a contrasting that low intensity on the T1-

weighted image and high intensity on the T2-weighted image and extending from the left caudate lobe to IVC (Figure 2). We diagnosed this tumor is a hepatic tumor in the Spiegel lobe with IVC tumor thrombus.

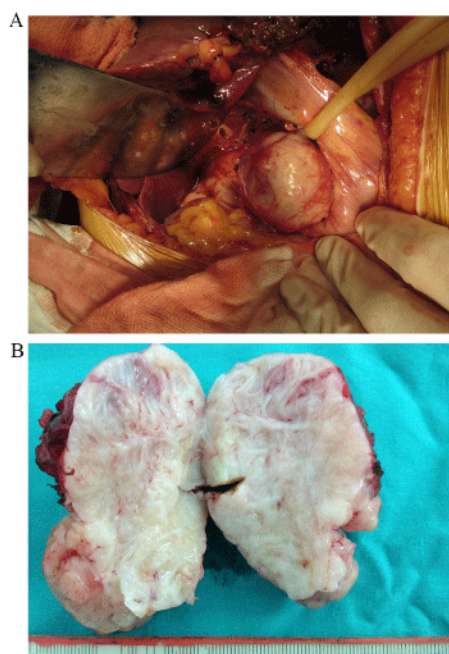
A right subcostal laparotomy with upper midline extension to the xiphoid process was performed. A laparotomy was performed and intraoperative findings revealed a 7 cm×6 cm×5 cm hard- tumor involving the suprahepatic IVC (Figure 3A). This tumor was not located in the Spiegel lobe of the liver but originated in the IVC. The falciform ligament was divided until the anterior surface of the suprahepatic IVC was exposed, and the infrahepatic IVC was dissected and mobilized. The tumor and the right renal vein was reached and exposed. In order to exposing the infrahepatic IVC lengthen, the left lateral section of liver was resected. The tumor was then dissected and removed from proximal to distal. After completed mobilization of tumor with adjacent pancreas, en bloc resection of the IVC tumor was performed under THVE (first Pringle's maneuver, then the infrahepatic IVC occlusion with a Satinsky clamp, and last, suprahepatic IVC occlusion with a Satinsky clamp). The primary IVC wall was repaired longitudinally; and the relaxation was on the contrary. That was maintained for 10 minutes for the stage of enable resection. Intraoperative blood loss was 800 ml. During THVE, the patient's hemodynamic condition was carefully monitored. The right kidney was not removed. The right renal vein was not sacrificed. The tumor specimen was removed (Figure 3B). Pathological examination confirmed primary leiomyosarcoma of the IVC (Figure 4).



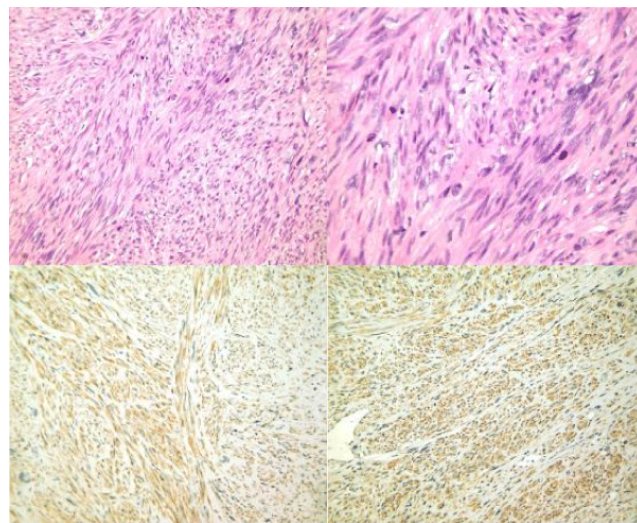
**Figure 1:** Enhanced computed tomography (CT) of the abdomen. The tumor was detected in the Spiegel of the liver on enhanced CT of the abdomen. Areas of hemorrhage and necrosis may be noted within the mass on CT.



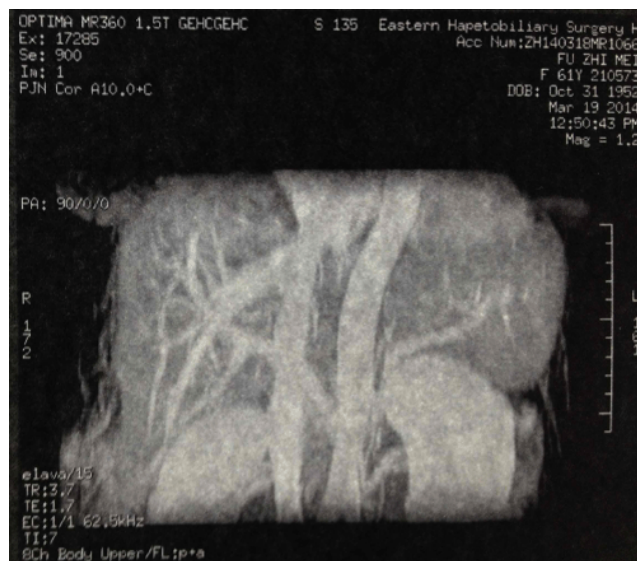
**Figure 2:** Magnetic resonance imaging (MRI) of this tumor revealed high signal intensity on T2-weighted imaging, and with IVCTT.



**Figure 3:** (A) Intraoperative picture of IVC leiomyosarcoma. (B) Resected leiomyosarcoma.



**Figure 4:** Pathological examination confirmed primary leiomyosarcoma of the IVC



**Figure 5:** MRI after 6 days postoperatively.

Tumor profile was gray, and there is complete tumor capsule. Edge of the specimen from the tumor about 0.5 cm, cutting edge no tumor cell growth. The patient's postoperative recovery was uneventful. She did not take anticoagulant drugs. Postoperatively, renal and liver function normalized on 7 days after surgery. MRI after surgery showed IVC is not narrow (Figure 5). She was discharged at 8 days after surgery. There was no clinical sign of leg edema throughout the postoperative course. Currently, the patient has normal life for nearly one year and no recurrence. To prevent recurrence, the patient will be radiotherapy.

## Discussion

Leiomyosarcomas originating in the IVC are rare malignant tumors. It is difficult in diagnosis of leiomyosarcoma. A large tumor was demonstrated in the Spiegel lobe liver with IVC tumor thrombus was imaged by tomography and magnetic resonance. The diagnosis was considered as tumor arising from segment I of the liver with IVC tumor thrombus. But in operation, the tumor was found from IVC, which was performed by suprahepatic and infrahepatic IVC occlusion with Satinsky's clamp in the operation. The patient underwent a combined operation which is en bloc resection of the IVC tumor and lobotomy of the left lateral section of liver. Pathological examination confirmed that is primary leiomyosarcoma of the IVC. The patient had a normal life for nearly one year and no recurrence. So the leiomyosarcoma of IVC is mimicking hepatic tumor with IVC tumor thrombus. In some cases it is difficult to distinguish leiomyosarcoma from a hepatic tumor [7]. Kulaylat et al. reported about two thirds of these patients were confirmed as the diagnosis of leiomyosarcomas only after laparotomy. We misdiagnosis to be considered as tumor arising from segment I of the liver with IVC tumor thrombus was lead to the tumor to predominant intra-luminal growth.

It is difficult to distinguish leiomyosarcoma from a hepatic tumor.

- This tumor commonly shows vague abdominal symptoms. Patients may also present with lower extremity edema. Patients may present with Budd Chiari syndrome when the tumor is involving the supra hepatic IVC [8,9].
- CT and MRI can confirm the existence of this tumor. Making the preoperative diagnosis is usually difficult. Although the modern imaging techniques make rapidly in progress, it is difficult to diagnose a leiomyosarcoma preoperatively. About two thirds of these patients were confirmed as the diagnosis of leiomyosarcomas only after laparotomy [10]. In this case CT revealed the existence of the tumor in the liver and spreaded in IVC. The misdiagnosis to be considered as tumor arising from segment I of the liver with IVC tumor thrombus was lead to the tumor to predominant intra-luminal growth [11].
- At MRI and CT, leiomyosarcomas of the IVC is lack of blood supply, so no enhancement in the arterial phase is the only difference method comparing with IVC tumor thrombus.
- Leiomyosarcomas of the IVC are growing slowly and at the time of diagnosis, they are often very large so as too difficult to make curative resection [4,5,7].

Surgical resection with a tumor-free margin is recommended to promote survival [12,13]. If the circumference of the IVC to be resected is <75%, cavoplasty can be performed. If the amount of IVC to be resected is >75%, complete resection and reconstruction is required [6]. In this case, the IVC tumor was predominant intra-luminal growth. We think the surgical management of partial resections of the IVC invading <30% circumference of the vena cava, reconstruction of the IVC is not required.

The technical challenges in this operation related to the control of infrahepatic IVC that it used suprahepatic IVC and infrahepatic IVC occlusion with Satinsky clamp, without using umbilical method encircled suprahepatic IVC and infrahepatic IVC. A major advantage of this manoeuvre is that it avoids the mobilization of the suprahepatic and infrahepatic IVC, which method is simple. The time of occlusion of the IVC is only 10 minutes. If the IVC bleeding is not to be controlled, forceps/clamps are then placed at the proximal and distal aspects of the segment should be resected.

With radical resection, the 5-year survival rate for these patients are between 30% and 70% [4,6,8,14-17]. No randomized controlled trials are exist to evaluate the use of neoadjuvant or adjuvant therapy for patients with leiomyosarcomas, but many clinicians advocate the use of radiotherapy for patients that found have positive margins [6].

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## Conflict of interest statement

Aijun Li, Teng Zhao, Lei Yin, Xiaoyu Yang, Mengchao Wu have no conflict of interest.

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