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Misdiagnosis of Leiomyosarcoma of Infrahepatic Inferior Vena Cava as A Liver Tumor in Caudate Lobe with Inferior Vena Cava Tumor Thrombus

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Inferior vena cava; Sarcoma; Surgery

1. Abstract

1.1. Background

Leiomyosarcoma of the inferior vena cava (IVC) is a rare malignant tumor. Surgical resection is currently the only potential curative treatment. According to the treated experience of one patient in our hospital, we present our opinions as below.

1.2. Methods and Results

We present a case of 61-year-old woman with leiomyosarcomas in the IVC. The tumor appearance on pre-operative imaging simulated a hepatic tumor with IVC tumor thrombus. In operation, the tumor was found from IVC. The patient was underwent successful surgical treatment for a leiomyosarcoma. Pathological examination confirmed that it was a primary leiomyosarcoma of the IVC. The patient had a normal live for two years and without recurrence.

1.3. Conclusion

We encountered a rare case of a leiomyosarcoma of IVC mimicking hepatic tumor with IVC tumor thrombus and misdiagnosis. Radical surgical resection is the primay treatment for IVC leiomyosarcomas. Surgical resection followed by radiotherapy can enhance long-term survival.

2. Introduction

Leiomyosarcomas of the inferior vena cava (IVC) are rare malignant and slow-growing tumors with a poor prognosis [1-6]. It was first reported by Perl and Virchow in 1871 [7,8] and was found in the German literature. Approximately 400 cases have been described in the literature [6,8,9]. Leiomyosarcomas are originated from the mesenchymal cells and predominantly proposed within the IVC [10-12]. Resection was often presented with a challenge as these tumors may require reconstruction of the IVC. Now we report a case of surgical resection of an infrahepatic IVC leiomyosarcoma mimicking a hepatic tumor with IVC tumor thrombus on CT/MRI.

3. Case Report

A 61-year-old woman was examined for pain in her right upper abdominal in pain in April 2014. She was admission to Eastern Hepatobiliary Surgery Hospital with a probable diagnosis of hepatocellular carcinoma (HCC) with inferior vena cava tumor thrombus (IVCTT). She had not any drug history, hepatitis, and family history. 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. 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 (CT) of the abdomen revealed a liver tumor in the left caudate lobe with a 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 2,3). We diagnosed this tumor as a hepatic tumor in the left caudate lobe with IVCTT. Therefore, we decided surgical treatment of HCC associated with IVCTT. A right subcostal laparotomy with upper midline extension to the xiphoid process was performed. A laparotomy was performed and intraoperative findings this tumor was 7cm×6cm×5cm hard-mass, not locating in the left caudate lobe, and originating

in the infrahepatic IVC (Figure 4). 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 much more exposing the infrahepatic IVC, the left lateral section of liver was resected under Pringle's maneuver about 12 minutes. After completed mobilization of tumor, en bloc resection of the IVC tumor was performed under total hepatic vascular exclusion (THVE). The tumor specimen was removed (Figure 5). The IVC wall was reconstructed with end-to-end direct anastomosis using polypropylene. The patient obtained negative margins (R0 resections). During THVE 10 minutes, the patient's hemodynamic condition was carefully monitored. Intraoperative blood loss was 800 ml. Pathological examination confirmed primary leiomyosarcoma of the IVC. The patient's postoperative recovery was uneventful. She did not take anticoagulant drugs. The renal and liver function normalized on 1, 3, and 7 days after surgery. MRI after surgery showed IVC is not narrow (Figure 6). She was discharged at 8 days after surgery. There was no clinical sign of leg edema throughout the postoperative course. To prevent recurrence, the patient had received adjuvant radiotherapy after operation one month. Following up 2 years, the patient is alive with survival of 2 years no recurrence (Figure 7. This picture can be concealed). Later, contact was lost.

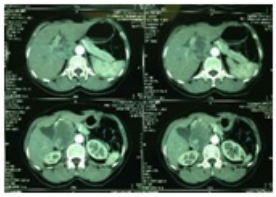

Figure 1. Enhanced computed tomography (CT) of the abdomen revealed a liver tumor in the left caudate lobe with a mass.

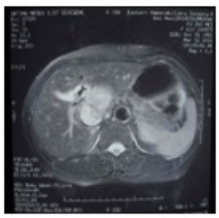

Figure 2. Magnetic resonance imaging (MRI) of this tumor revealed high signal intensity on T2-weighted imaging, and with IVCTT.

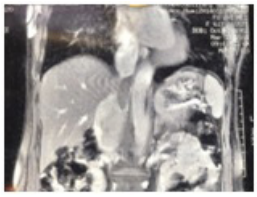

Figure 3. Magnetic resonance imaging (MRI) of this tumor revealed high signal intensity on T2-weighted imaging, and with IVCTT.

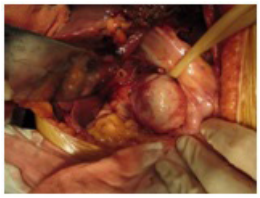

Figure 4. Intraoperative picture of IVC leiomyosarcoma.

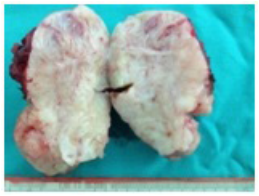

Figure 5. Resected leiomyosarcoma.

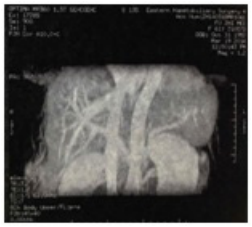

Figure 6. MRI after 6 days postoperatively.

Figure 7. Two years later, the patient came back, no recurrence.

4. Discussion

1. We have analyzed the reasons for misdiagnosis. Leiomyosarcomas of the IVC are rare malignant. These tumors usually go unnoticed unless it has metastasized to distant tissues [10,11,13]. Clinical signs and symptoms are very vague and atypical. Usually it is often misdiagnosed as an abscess cavity in the liver or primary hepatic malignancy [6]. 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 and obstructing the IVC. CT and MRI can confirm the existence of this tumor. Making the preoperative diagnosis is usually difficult [13]. About two thirds of these patients were confirmed as the diagnosis of leiomyosarcomas only after laparotomy [9-13]. In our case, the tumor appearance on pre-operative imaging mimicking hepatic tumor with IVCTT. Radiological investigations are the key to proper diagnosis. We need identify leiomyosarcomas with HCC and intrahepatic cholangiocarcinoma (ICC).

2. Surgical resection with a tumor-free margin is recommended to

promote survival [1-6]. Depending upon the exact location, further treatment options vary. The optimal treatment is completely resect the malignant lesion with preservation of venous return. Resection was often presented with a challenge as these tumors may require reconstruction of the IVC. The need of vascular reconstruction is not always mandatory. It is important to consider the length and circumference of the IVC to be removed. If the circumference of the IVC to be resected is less than75 %, cavoplasty can be performed. If the amount of IVC to be resected is greater than 75 %, complete resection and reconstruction is required [12]. We present the case of a patient with the tumor growing into the cavity of IVC or intraluminal growth pattern, looking like a cauliflower. There was a small pedicle connected with IVC and tumor. The defect in the IVC after resection was not large. So the IVC wall anastomosing was performed, without replacement.

3. With radical resection, the 5-year survival rate are between 30% and 70% [6,10,12]. Now no randomized controlled trials are exist to evaluate the use of therapy for patients with leiomyosarcomas [1-6]. No consensus regarding adjuvant treatment exists. We present a rare case of leiomyosarcoma of IVC using adjuvant radiotherapy after surgery. This case report will help to such rare tumors in diagnosis and treatment to prolong patient survival.

5. Conclusion

In conclusion, leiomyosarcomas originating in the IVC are rare malignant tumors. We experienced a rare case of a leiomyosarcoma of IVC mimicking hepatic tumor with IVC tumor thrombus. Further research are needed to understand the disease and to guide its management.

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