Int J Clin Exp Med 2014;7(8):2365-2368
www.ijcem.com /ISSN:1940-5901/IJCEM0000905

Case Report

Inferior vena cava leiomyosarcoma confirmed by catheter suction biopsy during digital subtraction angiography

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Received May 27, 2014; Accepted July 9, 2014; Epub August 15, 2014; Published August 30, 2014

Abstract: Leiomyosarcoma of vascular origin is a rare disease and most cases arise in the inferior vena cava. Inferior vena cava leiomyosarcoma (IVCLMS) usually presents in females in their sixth decade of life. The clinical symptoms are often non-specific and the diagnosis is often delayed. Current imaging techniques can accurately differentiate inferior vena cava neoplasms from other non-neoplastic lesions. However, definitive diagnosis of IVCLMS needs histologic evidence. We report a case of IVCLMS in a 61-year old Chinese woman. This is the first IVCLMS case confirmed by catheter suction biopsy during digital subtraction angiography.

Keywords: Leiomyosarcoma, inferior vena cava, catheter suction, digital subtraction angiography

Introduction

Inferior vena cava leiomyosarcoma (IVCLMS) is a rare, slow growing smooth muscle sarcoma arising from the media of the inferior vena cava (IVC) [1]. Since the first report by Perl *et al.* in 1871, more than 400 cases of IVCLMS have been reported [2, 3]. IVCLMS occurs usually in adults with a female predominance. The mean age is around 60 years [1]. Clinical presentations vary, depending on tumor size and the levels of IVC involved. Wide surgical excision with negative margins is the standard treatment and the only hope for prolonged survival [4]. However, IVCLMS is often diagnosed at advanced stage and negative surgical margins often can not be achieved [5]. The long term outcome after surgery is dismal, and the five-year and ten-year disease-free survival are 31.4% and 7.4%, respectively [2].

Clinical symptoms are non-specific and often precede the diagnosis for years. Accurate radiologic diagnosis of IVCLMS can be achieved by combination of various imaging techniques, such as computed tomography (CT), magnetic resonance imaging (MRI), cavography and ultrasound (US) [6]. Conversely, definitive diagnosis of IVCLMS still needs histological examination and tissue procurement is usually done by laparotomy or percutaneous needle aspiration/biopsy [7].

Digital Subtraction Angiography (DSA) is one fluoroscopy technique developed in 1970s and used in interventional radiology. A series of images of the vessels of interest are taken before and after injecting contrast medium. The signals of surrounding soft tissue and bone are digitally eliminated in a real time fashion. DSA provides accurate images of blood vessels and is especially useful in identifying and diagnosing vascular abnormalities [8].

Here we report one case of IVCLMS in a 61-year-old Chinese woman. The diagnostic tumor sample was procured by DSA catheter suction, which has not been reported in English literature.

Case report

The patient was a 61-year-old female, presented with intermittent, vague abdominal pain of the right upper quadrant for more than 10

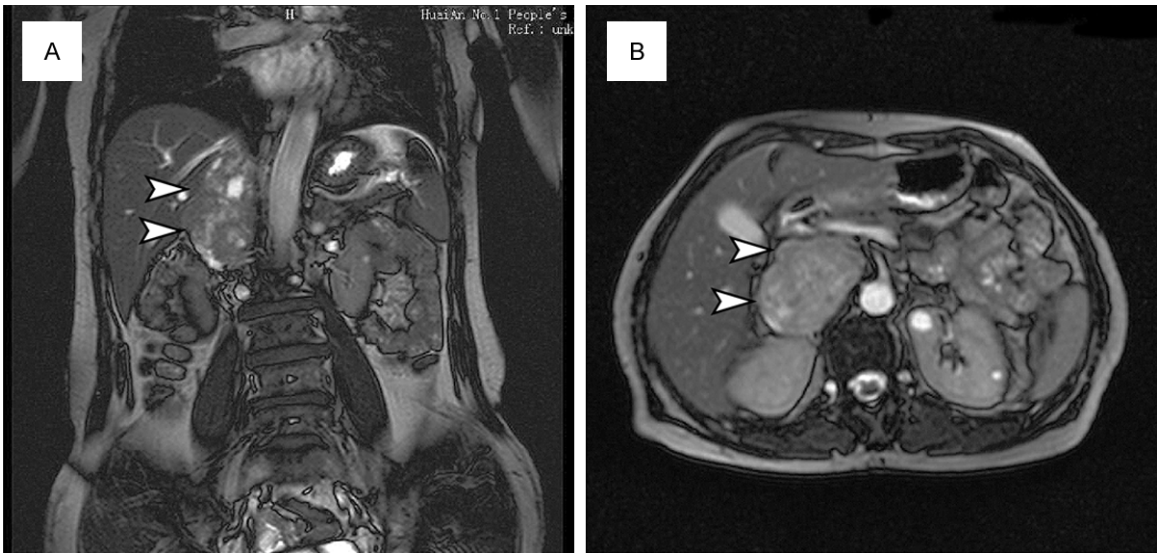

Figure 1. The MRI features of the IVC tumor with mixed T1WI and T2WI signals. The coronal view (A) and transverse view (B) shows an 8 x 7 x 6.5 cm mass (white arrowheads) within the IVC lumen. The hepatic segment IVC is dilated significantly (A). The lower end of the mass involves the main stem of right renal vein (B).

years. The pain episodes usually lasted several days with asymptomatic intervals of several months. She presented to an outside hospital in December 2012 and MRI of abdomen rendered a preliminary diagnosis of "IVC thrombosis". The patient was transferred to our hospital for further care. The outside MRI images were reviewed and showed widened hepatic segment of IVC by an 8 x 7 x 6.5 cm intraluminal lesion. The lesion extended from the opening of the hepatic vein to the right renal vein (**Figure 1A**, **1B**). Color Doppler ultrasound (Phillip - Iu22) performed at our hospital revealed an 8.6 x 5.6 cm mass with low-level echoes located in the epigastrium and on the right anterior aspect of the spine. The mass was within IVC lumen causing blood backflow and showed abundant blood flow signals with clear demarcation. The overall radiologic features were suggestive of an IVC smooth muscle neoplasm.

To further evaluate the lesion and degree of obstruction, DSA was performed with a GE Innova-3100 Cardiovascular Digital Cath & Angio machine. The catheters were inserted through right femoral vein by modified Seldinger technique. DSA showed a large filling defect within IVC, extending from the stem of right renal vein up to the hepatic segment. To procure the tissue, a 6F guiding catheter (Cordis, Warren, New jersey, USA) was extended to the mass and suction pressure was exerted by a 20-ml syringe. The procedure was uneventful and our patient did not develop any complications, such as pulmonary embolism or bleeding.

Grossly, the specimen was composed of small fragments of tan-white soft tissue. Sections showed a cellular spindle cell neoplasm with interlacing bundles. The nuclei showed polymorphism and hyperchromasia (**Figure 2A**, **2B**). Mitotic figures were 5-10 per 10 high power fields (hpfs). The neoplastic cells were positive for desmin (**Figure 2C**), and actin (not shown). The neoplastic cells were negative for S100, and CD117 (not shown). Ki67 revealed a proliferative index of approximately 60%-70% (**Figure 2D**). The overall pathologic findings were consistent with IVCLMS. The patient voluntarily gave up further treatment.

Discussion

IVCLMS is a rare vascular smooth muscle malignancy. The non-specific symptoms include nausea, vomiting, abdominal pain, hepatomegaly, portal hypertension, lower extremity edema, and deep vein thrombosis [1]. Traditionally, using hepatic vein and renal veins as landmarks, IVCLMS is divided into three subtypes (upper, middle and lower). Upper IVCLMS is above the origin of the hepatic vein; middle IVCLMS is between the hepatic vein and renal veins; lower IVCLMS is below the renal veins.

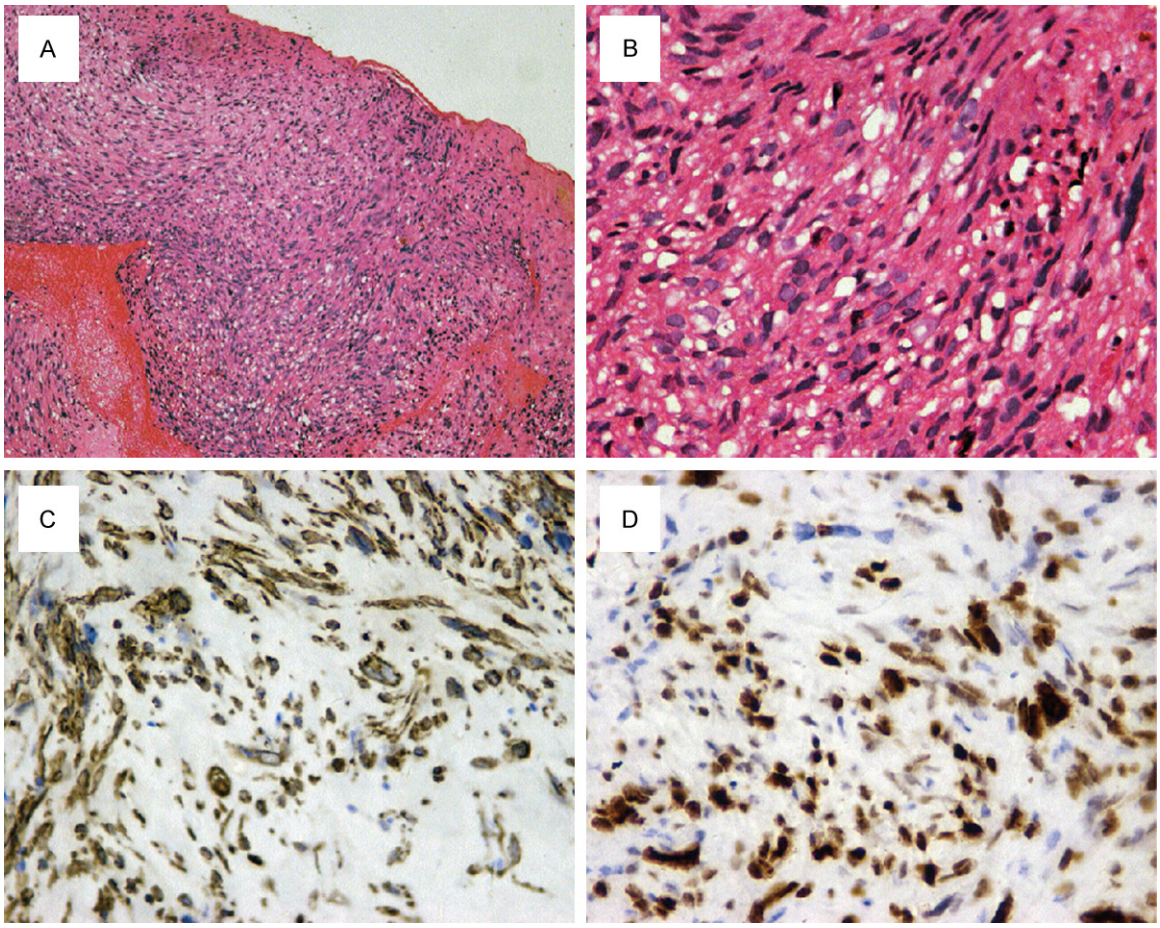

Figure 2. Histologic features of the IVC leiomyosarcoma. (A) Hematoxylin and Eosin stain shows a cellular spindle cell neoplasm with interlacing bundles. (B) The neoplastic cells have significant nuclear pleomorphism and hyperchromasia. The neoplastic cells are positive for Desmin (C). (D) Ki-67 reveals a proliferative index of approximately 60%-70%.

Middle IVCLMS comprises 33% of all cases and is associated with relatively better prognosis. The upper and lower IVCLMS comprises 1% and 8% of all cases, respectively. Approximately 34% of IVCLMS involved more than one segments of IVC and the remainder did not specify the tumor origins [3]. The bulk of the tumor of our case is between hepatic vein and renal veins, consistent with a middle IVCLMS.

Due to its rarity, if radiologists are not aware of the imaging features of IVCLMS, it could be misdiagnosed as other more common IVC lesions, including thrombi, cancer thrombi, benign leiomyoma, and other retroperitoneal malignant neoplasms [6]. Our case had been misdiagnosed as "IVC thrombosis" at an outside hospital. The following clinical and radiologic features help differentiate IVCLMS from other IVC lesions. IVC thrombus presents as a filling defect with discernible boundaries from vessel walls, and shows no enhancement. Tumor thrombi are most commonly seen with hepatocellular carcinoma and renal cell carcinoma and patients usually have prior history. The benign leiomyoma involving IVC usually originates from intrauterine or pelvic veins, and forms cordlike solid lesion, extending along the vessels to IVC and right atrium [9]. Other retroperitoneal malignant tumors usually compress, instead of expand, IVC.

Modern imaging modalities of CT and MRI, combining with US and angiography, are capable of diagnosing vascular neoplasms with great specificity. CT provides intra-tumor details, including necrosis, calcification, and hemorrhage. Enhanced CT provides further details of tissue heterogeneity, as well as adhesion or replacement of surrounding tissue. MRI is con-

sidered the best imaging method for vascular neoplasm, which is characterized by the equal or low signal on T1 weighted image (T1WI) and equal or high mixed signal on T2 weighted image (T2WI). Irregularity of tissue enhancement can be shown on the enhanced scanning. Magnetic Resonance Angiography (MRA) provides whole views of the tumor, and evaluates the extent of vascular infiltration and the relationship between the tumor and IVC branches.

DSA is one type of fluoroscopies used in interventional radiology and has certain advantages in diagnosing vascular neoplasms. Besides showing the level of obstruction and bypass circulation, catheters of different calibers are available to examine the status of IVC branches. In addition, tissue procurement by suctioning is theoretically feasible. Even though catheter suction biopsy carries a risk of tissue dislocation and embolism, our limited experience observed no such complications. Although no report about suction biopsy of IVCLMS is found in the literature, a successful suction biopsy by using a 10F steerable embolectomy catheter has been performed in one case of pulmonary artery leiomyosarcoma with no complications observed [10]. In comparison, the previously reported IVCLMS cases are usually biopsied by percutaneous needle aspiration/biopsy or laparotomy before surgery. Our case is the first IVCLMS procured successfully by catheter suctioning during DSA.

IVCLMS show similar histologic features as those of leiomyosarcomas from other locations. Immunohistochemistry is necessary to exclude other more rare spindle cell malignancies. There is no consensus on how to grade IVCLMS. Hilliard *et al*. proposed to a grading method based on mitotic figures (MF): high grade (≥ 10 MF/10 hpfs); intermediate grade (5-9 MF/10 hpfs); and low grade (1-4 MF/10 hpfs) [3]. According to this proposal, our case is an intermediate grade IVCLMS.

Our limited experience suggests catheter suctioning is a relatively safe, quick and less invasive method to procure vascular tumors during DSA. Obviously, more cases are needed to consolidate this impression. It is critical to closely monitor the patients for bleeding and embolic complications during and after the procedure.

Disclosure of conflict of interest

None.

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