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Case Report

Transvenous biopsy of inferior vena cava leiomyosarcoma: two case reports ☆,☆☆

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ABSTRACT

Leiomyosarcomas of the inferior vena cava (IVC) are uncommon malignancies. There is limited research detailing optimal diagnostic and clinical management. Here, we present 2 unique cases of IVC leiomyosarcoma including one in which the mass was partially ruptured through the vessel at initial presentation. We detail radiologic findings, 2 different transvenous approaches for biopsy of these masses, and subsequent oncological management.

Introduction

Leiomyosarcomas of the inferior vena cava (IVC) are extremely rare malignancies, with only a few hundred cases reported in the literature [1–5]. They carry a poor prognosis. Due to their location, IVC leiomyosarcomas present insidiously with non-specific abdominal pain and are often discovered as advanced disease [1]. While the masses are identified in routine imaging, they require biopsy for definitive diagnosis. Typically, biopsy is performed percutaneously. However due to their rarity, much remains unknown about optimal diagnostic management of the disease outside of evidence from a few case reports in the literature [1–5].

Here we present 2 cases of IVC leiomyosarcoma in which transvenous biopsy was considered safer than percutaneous biopsy. The first is a case where workup revealed a mass that was partially ruptured through the IVC into the right adrenal gland. The second is a case in which preserved flow in the IVC adjacent to the tumor was considered high risk for biopsy-

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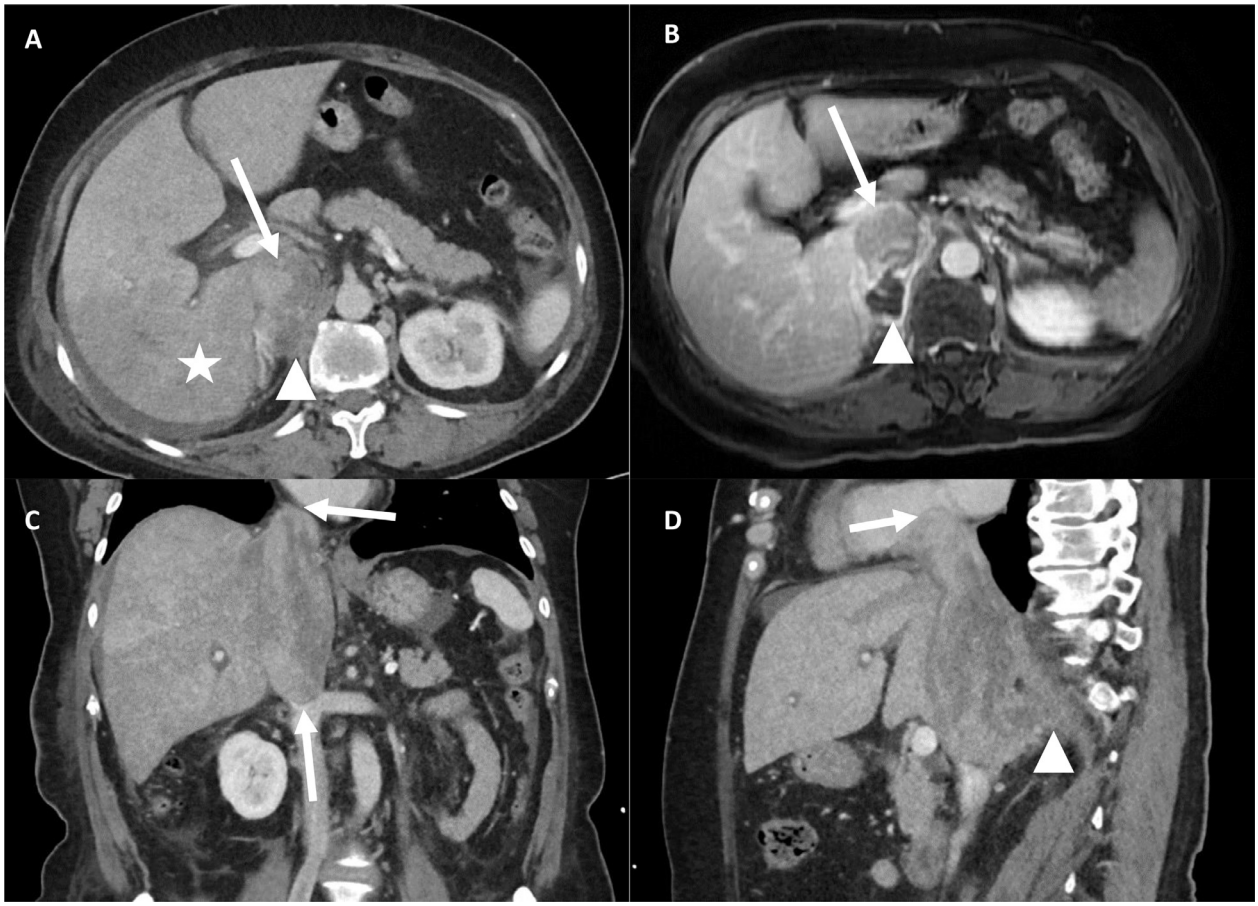

Fig. 1 – IVC mass; Case #1. (A) Axial CT showing the IVC mass (arrow) with extension into the right adrenal gland (arrowhead) and hepatic congestion (star). (B) Axial T1-weighted post-contrast MRI showing the IVC mass (arrow) with extension into the right adrenal gland (arrowhead). (C) Coronal CT showing extent of mass from the level of the renal veins to the lower right atrium (arrows). (D) Sagittal CT showing the mass invading the lower right atrium (arrow) and right adrenal gland (arrowhead).

associated hemorrhage. In both cases a transvenous biopsy approach was used to sample the tissue, with the first case employing both fluoroscopic and cone-beam CT image guidance. We detail this approach and subsequent oncological management of the patients.

Case 1

A 63-year-old woman with no major medical history presented to our hospital with generalized abdominal tenderness, fatigue, chest pain, and shortness of breath. CT of the abdomen and pelvis with contrast revealed a 5.0 × 5.4 × 12.9 cm mass in the proximal inferior vena cava extending from the level of the renal veins to the right atrium (Fig. 1). Bland thrombus was observed inferior to the mass and venous congestion was observed throughout the right hepatic lobe, secondary to right hepatic vein outflow obstruction from the mass.

The patient was admitted and referred to interventional radiology for tissue sampling. Review of the imaging found that the mass had partially ruptured through the IVC into the right adrenal gland (Fig. 1). Thus, transvenous biopsy was considered the most appropriate method to limit the risk of seeding or further rupture from a percutaneous approach.

Percutaneous access was achieved from the right common femoral vein. Angiography revealed a filling defect from the mass just superior to the renal inflows, with prominent retroperitoneal collaterals (Fig. 2). A 7 French trans jugular liver biopsy cannula was manually modified to approximate the relatively straight trajectory through the mass and advanced through a sheath into the proximal IVC (TLAB cannula; Argon Medical Devices, Plano TX). The cannula was positioned 10mm inferior to the mass and the sheath retracted to expose the cannula to enable the best angulation (Fig. 3).

After placement of the canula under fluoroscopy, cone beam CT was used to locate the tumor relative to the placement of the transvenous biopsy needle (Fig. 3). It showed the needle centered just inferior to the mass – suitable for biopsy. Subsequent biopsy was performed under fluoroscopic guidance. Of note, intravenous ultrasound (IVUS) was initially considered instead of CT. However, the tumor significantly decreased the diameter of the IVC, making positioning of an IVUS probe in a way that would allow visualization of the biopsy needle difficult.

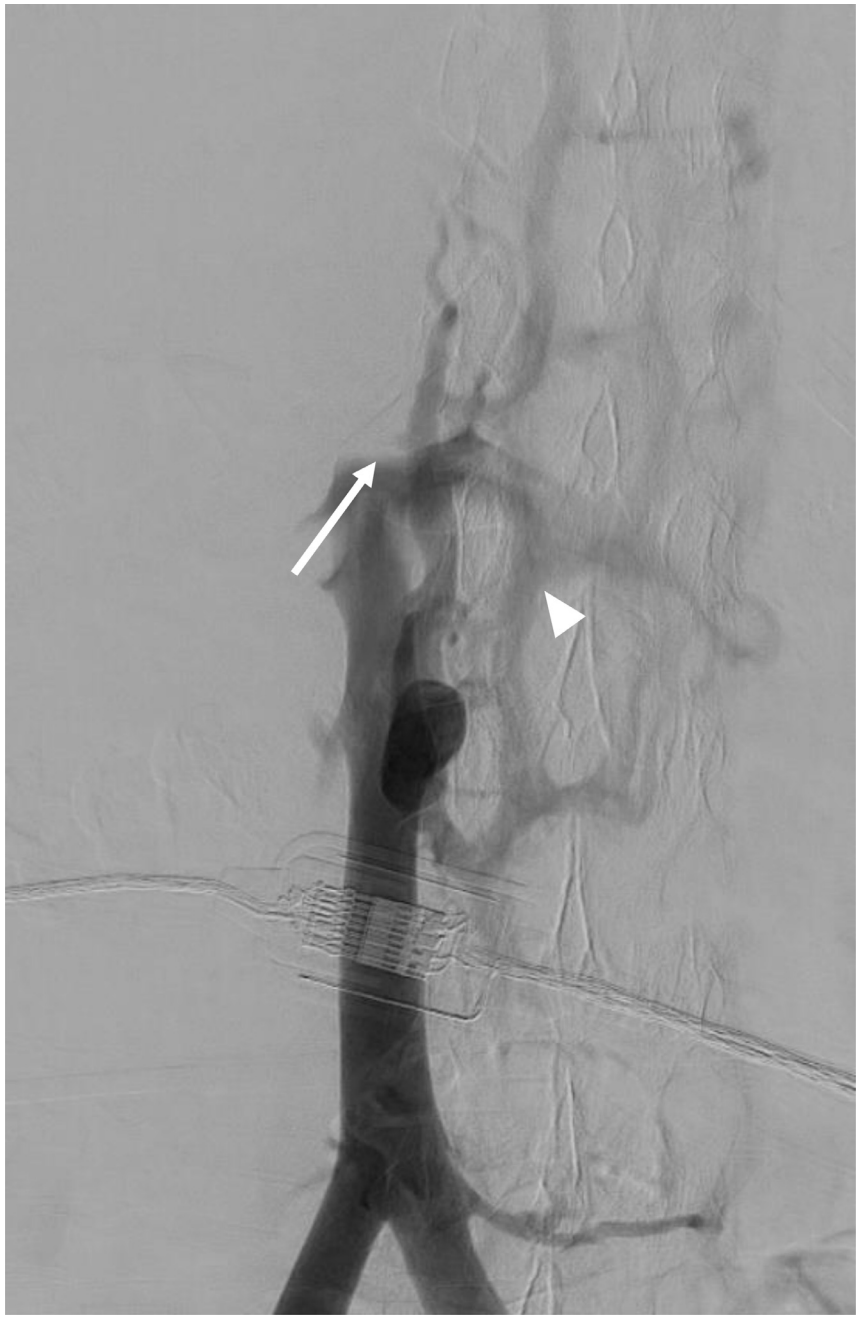

Fig. 2 – Case #1; Digital subtraction venography showing abrupt filling defect in the IVC due to the mass (arrow). Prominent retroperitoneal collateral veins are seen (arrowhead).

Pathology revealed spindle cell tumor composed of highly pleomorphic hyperchromatic large cells in a fibrous background with patchy necrosis (Fig. 4). Frequent mitoses were noted (approximately 15 mitoses per 10 hpf). Immunohistochemical staining demonstrated tumor cells to be diffusely strong positive for caldesmon and desmin, diffusely weak positive for smooth muscle actin (SMA) and CD34, and negative for pan-cytokeratin and S100 (Fig 4). These findings were compatible with the diagnosis of leiomyosarcoma.

The patient was referred to oncology and started on neoadjuvant chemotherapy with Doxorubicin and Ifosfamide. Despite aggressive treatment, the patient developed a progressively worsening Budd Chiari-like syndrome secondary to hepatic vein outflow obstruction from the tumor. She died of complications approximately 6 months after the diagnosis.

Case 2

The second patient was a 61-year-old female who presented with a 3-day history of epigastric pain and shortness of

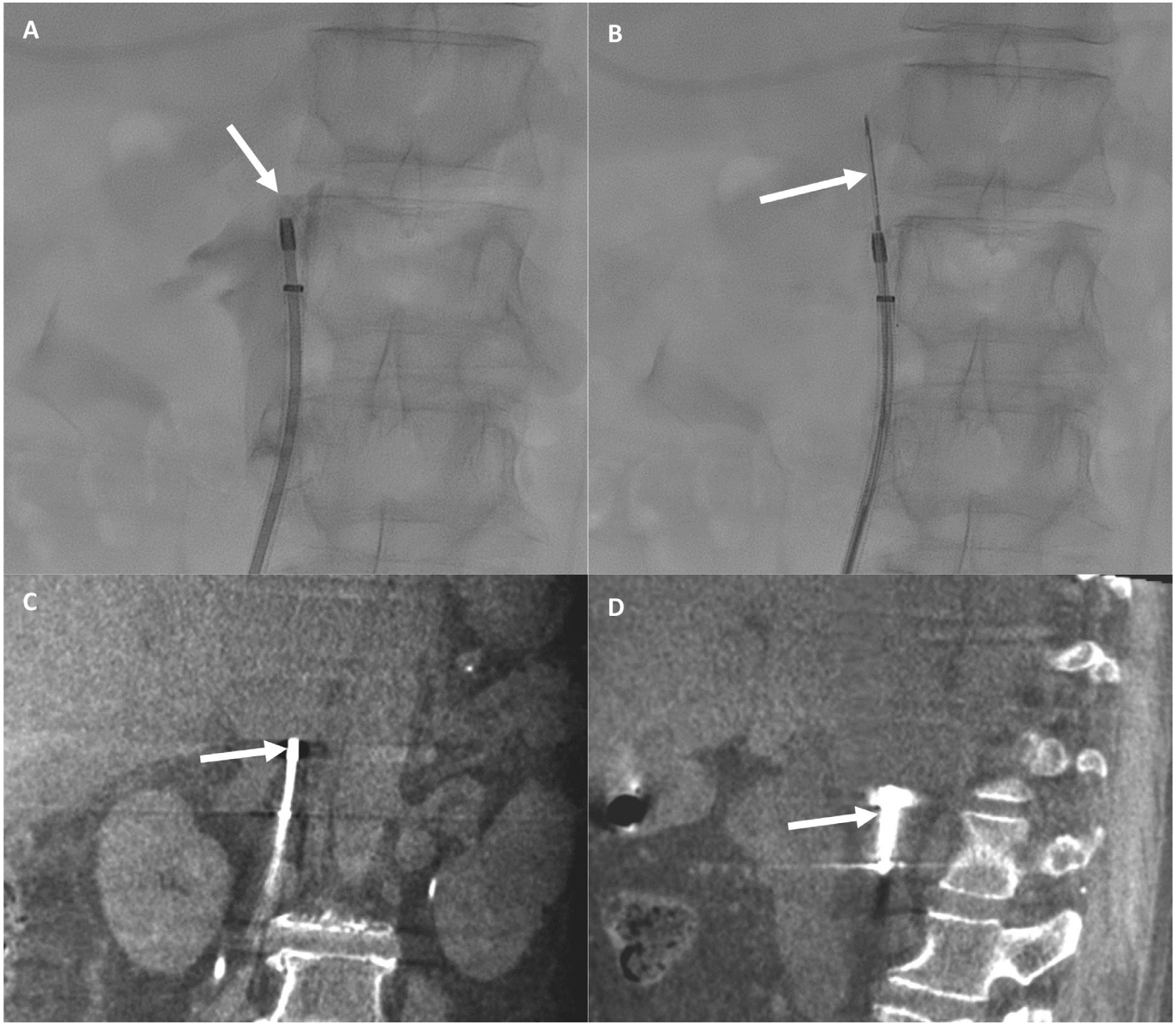

Fig. 3 – Transvenous biopsy; Case #1. (A) Position of the biopsy cannula inferior to the mass, with venogram showing the filling defect due to the mass (arrow). (B) Biopsy needle (arrow) deployed into the mass. (C) Intraprocedural coronal cone-beam CT confirming position of the biopsy cannula inferior to the mass (arrow) prior to needle deployment. (D) Intraprocedural sagittal cone-beam CT confirming position of the biopsy cannula inferior to the mass (arrow) prior to needle deployment.

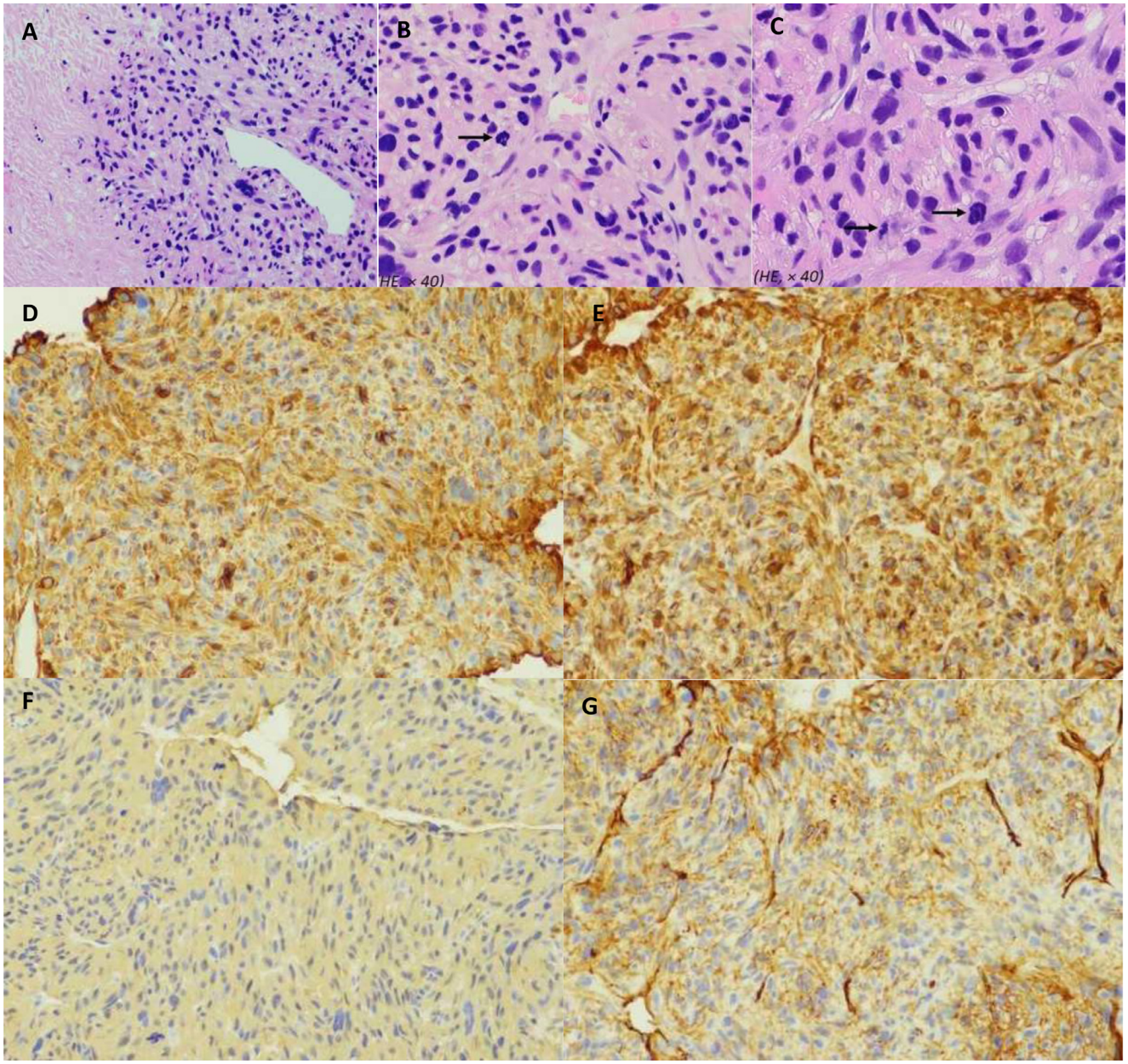

Fig. 4 – (A-C) Case #1; Histological findings of the tumor (hematoxylin and eosin staining)- pleomorphic hyperchromatic large cells and fascicles of atypical spindle cells. Frequent mitoses (arrows) are seen, and patchy necrosis (asterisks). (D-G) Immunohistochemical staining (x 20) showed diffusely strong positive results for (D) Caldesmon, and (E) Desmin and diffusely weak positive results for (F) Smooth Muscle Actin and (G) CD34.

breath. Initial CT revealed large thrombus or mass extending from the intrahepatic IVC down to the level of the renal veins, prompting further workup. A subsequent MRI revealed a 4.6 × 5.2 × 10.9 cm IVC mass with involvement of the proximal right hepatic vein (Fig. 5) .

The patient was admitted and referred to interventional radiology for tissue sampling. Importantly, there was suspected preserved flow through the IVC juxtaposed to the target biopsy site which was higher risk for hemorrhagic complications if the biopsy was pursued percutaneously; hence transvenous biopsy was performed (Fig. 5). Percutaneous access was achieved from the right femoral vein and multiple samples of the suspected tumor thrombus were obtained using 1.8 mm biopsy forceps (Radial Jaw 4 Gastro Pediatric Biopsy Forceps, Boston Scientific).

Pathology revealed spindled cell tumor. Immunohistochemical staining showed tumor cells to be positive for SMA, caldesmon and desmin but negative for CD34, pan-cytokeratin and S100. These findings were compatible with the diagnosis of leiomyosarcoma.

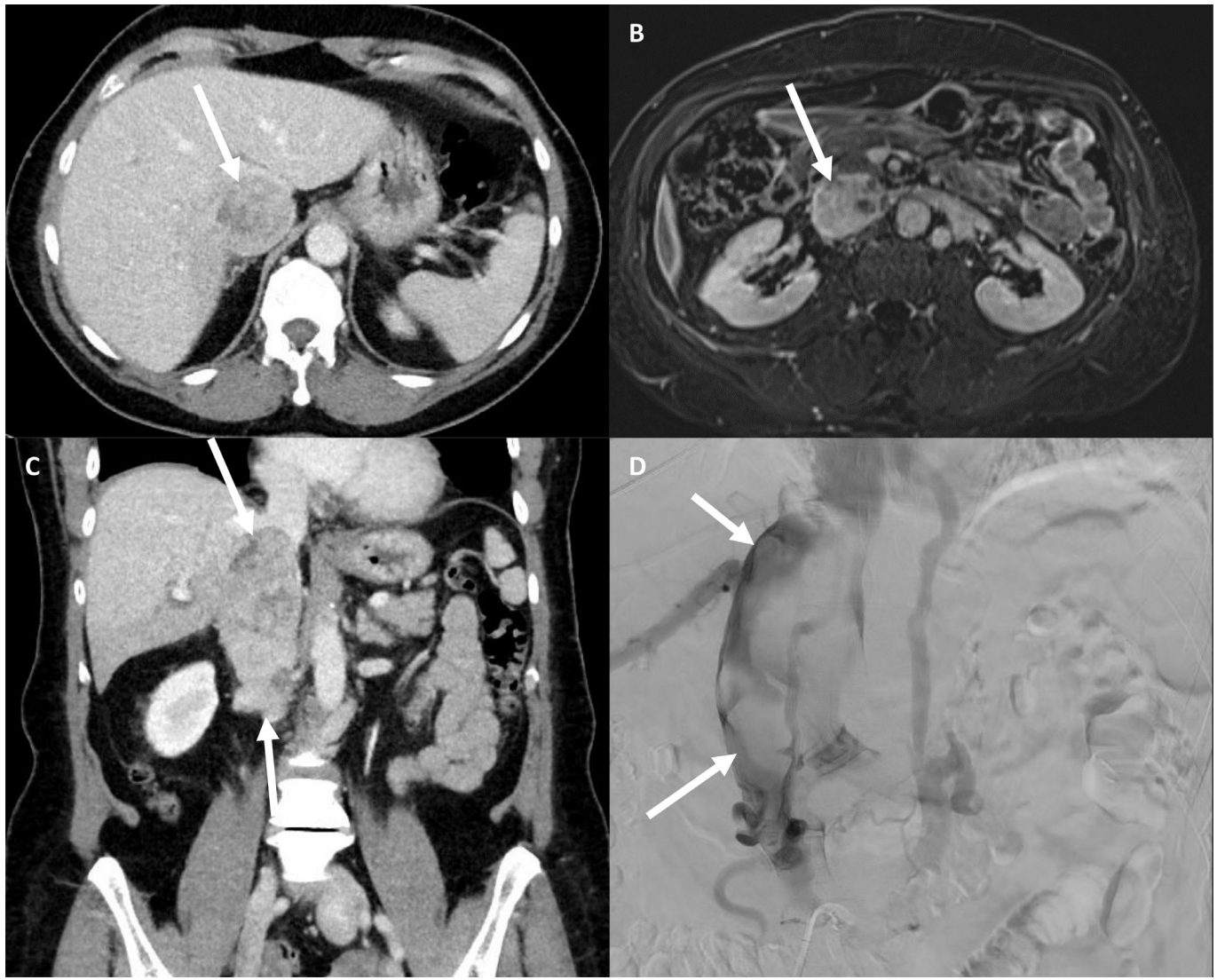

Fig. 5 – IVC mass; Case #2. (A) Axial CT and (B) Axial T1-weighted post-contrast MRI showing the IVC mass (arrow) (C) Coronal CT showing extent of mass (arrows) (D) Digital subtraction venography showing the IVC mass with collateral vessels and preserved flow within a small channel in IVC (arrows).

The patient was discharged and referred to oncology for determination of best neoadjuvant chemotherapy regime. Her clinical course continues to be followed.

Discussion

These cases illustrate two approaches for transvenous biopsy of locally advanced IVC leiomyosarcomas. In the first case, a transjugular liver biopsy set was used for biopsy under fluoroscopy and cone-beam CT guidance. In the second case, forceps were used to biopsy with fluoroscopy guidance, complementing previous cases using forceps and suction biopsy for similar transvenous biopsies of IVC masses [6,7]. These cases join a growing body of evidence supporting transvenous biopsy of IVC masses as an alternative to percutaneous biopsy [8].

Transvenous biopsy remains understudied, despite its use for sampling perivascular renal, hepatic, pancreatic, and pelvic masses [6,9–11]. While it carries a risk of hematologic seeding if the mass is unruptured, it decreases the risk of peritoneal seeding relative to percutaneous biopsy. Indeed, the first case illustrates the unique diagnostic scenario in which the mass was already partially ruptured through the IVC. Thus, a transvenous biopsy approach was taken to avoid the risk of possible peritoneal seeding or further IVC rupture.

These cases are congruent work suggesting that endovascular biopsies are safe and effective alternatives to percutaneous biopsy and may carry a lower risk of hemorrhage [6]. This lower risk of hemorrhage was taken into consideration in the second case in which the risk of hematologic seeding of transvenous biopsy had to be carefully balanced with what was considered a greater risk of peritoneal hemorrhage from percutaneous biopsy. Much remains unknown about the relative risks of these complications; and future decisions would benefit from large, controlled studies.

Both cases illustrate the importance of multidisciplinary collaboration and communication when managing patients with these rare leiomyosarcomas. In particular, the first patient had very locally advanced disease that spanned from the renal veins up to the right atrium. While surgical resection is often curative and often combined with radiation as a cornerstone of management, both resection and radiation were difficult in this case due to the proximity of the tumor to the heart and surrounding structures. A multidisciplinary tumor board discussion determined that the best course of action was to start systemic therapy in a neoadjuvant fashion with the intent to obtain tumor control, with eventual plan for neoadjuvant radiation and subsequent surgery if the response was robust. The second patient's mass tumor was considered more amenable to possible resection, and a multidisciplinary tumor board recommended a similar course of neoadjuvant chemotherapy followed by possible radiation and likely surgical resection.

For the first patient, doxorubicin and ifosfamide were chosen as the neoadjuvant regime because rapid tumor response was needed to prevent further liver damage from hepatic venous obstruction due to the mass. A doxorubicin and ifosfamide regimen carries a survival advantage and highest response rate for all soft-tissue sarcomas including leiomyosarcoma [12]. Other neoadjuvant chemotherapy options include dacarbazine, gemcitabine, and trabectedin, but all these agents have a lower chance for tumor response and are partially metabolized by the liver which was of concern given the patient's elevated transaminases. The exact neoadjuvant regime for the second patient is actively being determined.

Conclusion

In summary, these cases illustrate two transvenous biopsy approaches of a rare primary inferior vena cava malignancy. Detailed imaging review and careful consideration about the risks and benefits of transvenous versus percutaneous biopsy approaches is paramount. Transvenous biopsy and subsequent pathology revealed IVC leiomyosarcomas and enabled prompt initiation of oncologic treatment.

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Patient consent

Written informed consent was obtained from the patient. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethics approval

No ethics approval was necessary for the treatment or investigation of this patient. All patient information and images have been de-identified.

Author contributions

CJM: manuscript writer/editor. DB: manuscript writer/editor and Interventional Radiology Resident. VK: manuscript writer/editor and Radiology Resident. PK: manuscript writer/editor and Hematology/Oncology Fellow. IL: manuscript writer/editor and Interventional Radiology Attending. FK: manuscript writer/editor and Interventional Radiology Attending.

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