Leiomyosarcoma of the Inferior Vena Cava

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CLINICAL SUMMARY

A 70-year-old with a history of bilateral breast cancer status post-mastectomies presented with nonspecific abdominal pain and right-sided back pain. She was treated symptomatically with oral pain medication; however, a routine cancer surveillance CT revealed a large, expansile mass involving the inferior vena cava (IVC).

Interventional radiology was consulted for a core biopsy of the mass, which was performed through a trans-jugular venous approach in a hybrid 4D-CT angiography suite. The procedure was performed safely and efficiently while obtaining 18-gauge core samples for surgical pathology.

IMAGING FINDINGS

An initial contrast-enhanced diagnostic CT scan of the abdomen and pelvis demonstrated a $4.9 \times 2.9 \times 5.3$ cm enhancing soft-tissue mass within the IVC (Figure 1). A conventional inferior vena cavagram showed occlusion of the IVC by the mass (Figure 2). A CT scan was obtained in the hybrid 4D-CT angiography suite (Canon Medical Systems USA, Inc.), with the biopsy needle located within the mass just prior to biopsy (Figure 3).Post-biopsy CT image with trans-jugular sheath in place revealed no evidence of hemorrhage or related complications (Figure 4).

DIAGNOSIS

Leiomyosarcoma of the IVC

DISCUSSION

First described by Pearl in 1871, leiomyosarcoma (LMS) of the IVC is the most common primary tumor of the IVC, although it is exceedingly rare, with only about 600 cases reported worldwide.^{1,2}

The condition has a 3:1 female predominance; for this reason, hormonal influence has been suggested, but not proven, as a risk factor.³

As with the patient described in this report, most cases of LMS are discovered in the sixth decade and often present with abdominal and or back pain.¹

Leiomyosarcoma of the IVC is classified into three categories:²

- Segment I: Infra-renal, presenting as lower extremity edema, right upper quadrant back, and flank, pain
- Segment II: Inter-renal and supra-renal, up to but not including the hepatic veins; presenting as abdominal and back pain, and sometimes renovascular hypertension.
- Segment III: Supra-hepatic, up into the right atrium; presenting with abdominal distention, nausea, vomiting, hepatomegaly, ascites, and jaundice.

While imaging findings of an enhancing intraluminal soft-tissue mass are fairly characteristic in the context of the typical patient demographic and presentation, obtaining tissue for pathology confirmation is essential. Consequently, the question is often raised: What is the safest and most accurate way to perform the biopsy?

The conventional percutaneous approach, via ultrasound or CT guidance, may be limited by the lack of a safe needle trajectory. At other times, more operator-dependent techniques such as adjunctive hydrodissection are necessary. In both cases, smaller devices are used prevent major bleeding while obtaining biopsy samples. With these techniques, tract seeding of the tumor has been described.⁴

A less commonly utilized procedure used to perform soft-tissue tumor biopsies is the transvenous approach. First reported in animal studies by Charles Dotter, MD, in 1962, the procedure was performed by Robins and Bookstein in humans in 1972.

The approach has been described in multiple studies in which a transjugular liver biopsy set is used to obtain adequate tissue for sampling. This has proven to be safe and



FIGURE 1. (A) Axial, (B) coronal, and (C) sagittal CT images demonstrating the large expansile, enhancing soft-tissue mass within the IVC.



FIGURE 2. Inferior vena cavagram showing occlusion of the IVC by the soft tissue mass (arrow).

effective when performed with a combination of fluoroscopy and intravascular ultrasound (IVUS) and/or CT.

Interventional radiologists will typically choose the imaging modality with which they have the most experience and which has kept their complication rates within the guidelines of the Society of Interventional Radiology.^{4,5}



FIGURE 3. Axial nonenhanced CT obtained in the hybrid 4D-CT suite with biopsy needle located within the mass, just prior to biopsy (arrow).



FIGURE 4. Post-biopsy CT image with trans-jugular sheath in place with no evidence of hemorrhage or related complications.

While some advocate cone beam for guidance, the authors believe a 4D-CT unit is the far superior technology for this and many other IR procedures.

With relative ease, we were able to place the tip of our biopsy needle within the most solid portion of the tumor while retaining the ability to perform a diagnostic CT scan to evaluate for appropriate position and immediate post-biopsy complications.

CONCLUSION

The transvenous approach to biopsy of soft-tissue masses is a safe and effective means of obtaining diagnostic tissue samples, especially when used in conjunction with hybrid 4D-CT imaging.

REFERENCES

1. Sulpice L, Rayar M, Levi Sandri G-B, et al. Leiomyosarcoma of the inferior vena cava. *J Visc Surg.* 2016;153(3):161-165.

2. Xu J, Velayati A, Berger BJ, Liu M, Cheedella NKS, Gotlieb V. Leiomyosarcoma of the inferior vena cava in an hiv-positive adult patient: a case report and review of the literature. *Am J Case Rep.* 2017;18:1160-1165. 3. Laskin WB, Fanburg-Smith JC, Burke AP, Kraszewska E, Fetsch JF, Miettinen M. Leiomyosarcoma of the inferior vena cava: clinicopathologic study of 40 cases. *Am J Surg Pathol.* 2010;34(6):873-881.

4. Sherk WM, Khaja MS, Majdalany BS, et al. Transvenous biopsy in the diagnosis of intravascular or perivascular neoplasm: a single-center retrospective analysis of 36 patients. *J Vasc Interv Radiol.* 2019;30(1):54-60. 5. Khalilzadeh O, Baerlocher MO, Shyn PB, et al. Proposal of a new adverse event classification by the society of interventional radiology standards of practice committee. *J Vasc Interv Radiol.* 2017;28(10):1432-1437.e3.