

# Leiomyosarcoma of the Inferior Vena Cava

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**Leiomyosarcomas (LMS) of the inferior vena cava (IVC) are rare and comprise less than 1% of all soft tissue sarcomas. Patients usually present in the fifth or sixth decade of life with back pain and/or increased abdominal girth. The diagnosis is usually made with noninvasive imaging [computed tomography (CT) or magnetic resonance (MRI)] or by invasive methods such as endoscopic ultrasound (EUS) with biopsy. We report the case of a 76 year-old female with a chief complaint of vague intermittent abdominal pain. CT scan revealed a mass possibly arising from the pancreas. She underwent a diagnostic EUS which confirmed that the mass was retroperitoneal in origin compressing the IVC; fine needle aspiration (FNA) showed spindle cells. The patient underwent laparotomy and pathology revealed a leiomyosarcoma of the IVC.**

## CASE PRESENTATION

**A** 76-year-old Caucasian woman with a past medical history of coronary artery disease, diabetes mellitus, gastroesophageal reflux disease and anxiety presented with a two month history of progressively worsening sharp lower quadrant abdominal pain radiating to her right upper quadrant. She denied associated fever, chills, weight loss, night sweats, nausea, vomiting, constipation, diarrhea, melena or hematochezia. Initial laboratory evaluation showed mild kidney injury and normal CEA (1.3 ng/ml) and CA 19-9 (30 U/ml). Computed tomography (CT) scan of her abdomen and pelvis showed a 2.3 cm x 2.4 cm hypoechoic, retro-peritoneal mass between the pancreas and inferior vena cava (IVC). Magnetic resonance imaging (MRI) confirmed a solid mass along the posterior head of the pancreas and anterior to the IVC at the origin of renal veins. There was compression of the IVC however there was no thrombus or invasion.

Endoscopic ultrasound (EUS) confirmed

a peri-duodenal, peri-pancreatic diffusely hypoechoic, heterogeneous retroperitoneal mass, measuring 3 cm x 2.8 cm x 2.4 cm. Fine needle aspiration (FNA) revealed spindle cells. The positron emission tomography (PET) scan showed increased metabolic activity in the proximal duodenum or pancreatic head, without loco-regional or distant spread. At laparotomy, a 5.0 cm x 4.0 cm x 3.0 cm mass extending from anterior aspect of the IVC was noted, and en-bloc resection performed. Pathology revealed spindle cell proliferation with atypical features without necrosis or increased mitotic activity. Immunohistochemistry was positive for vimentin, desmin, actin, myosin and negative for CD117 (ckit) and S-100 with a Ki67 proliferative index of 40 to 60%. Findings were consistent with high grade, poorly differentiated leiomyosarcoma [Stage 2a (T1bN0M0), Grade 3].

Adjuvant radiotherapy was administered, and outpatient follow up revealed recurrence-free survival at 36 months.

## Discussion

Vascular leiomyosarcomas (LMS) are rare soft tissue sarcomas and account for 2% of all leiomyosarcomas<sup>1</sup> and 0.7% of all soft tissue

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sarcomas in adults.<sup>2</sup> Over half of these vascular LMS arise from the IVC. Patients usually present in the fifth or sixth decade of their lives with a male to female ratio of 1:4.<sup>3</sup> Clinical symptoms are vague and include back pain, increasing abdominal girth, weight loss and abdominal pain.<sup>4</sup> The tumor commonly grows extra-luminally (in more than 60%)<sup>5</sup> and hence pedal edema is not a common presentation. Clinical presentation depends on location and luminal versus extraluminal growth. Segment I (infra-renal involvement) occurs in 36% and can present as lower extremity edema, deep vein thrombosis (DVT) and abdominal mass. Segment II involvement (hepatic to renal veins) is the most common (up to 44%) and can

present as abdominal pain, nephrotic syndrome and hypertension. Segment III involvement (right atrium to hepatic veins) is least common and can cause cardiac arrhythmias and Budd-Chiari syndrome. Liver involvement by direct contiguity or metastases is less common but has been noted (up to 20%).<sup>6</sup> These tumors tend to metastasize primarily to lung and liver and occasionally to bone and brain.<sup>7,8</sup>

Tumors originating from segment II respond better to therapy.<sup>3,4</sup> Other prognostic factors include tumors arising from tunica media and absence of palpable abdominal mass. Presence of abdominal pain (which occurs earlier in segment II involvement due to rich innervation) portends



**Figure 1. Retroperitoneal Peri-ampullary Peri-pancreatic Mass**



**Figure 2. Peri-pancreatic mass**

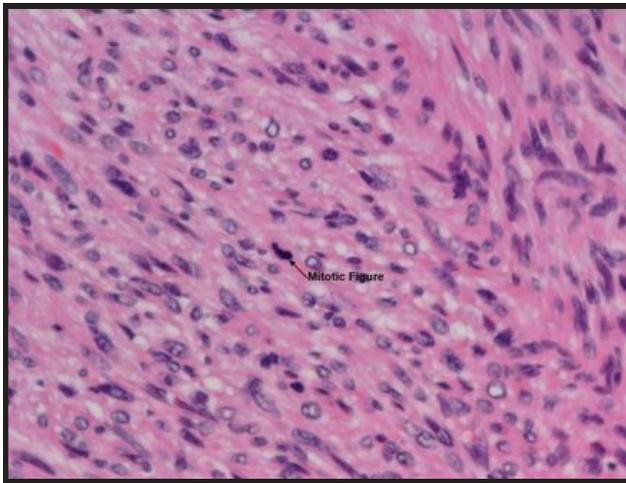


**Figure 3. Relation of mass to IVC**



**Figure 4. Mass undergoing FNA**

## A CASE REPORT



**Figure 5. Mitotic figures on pathology**

better prognosis. Improved survival rates are noted with early detection, surgery and neoadjuvant therapy.

Surgical resection is the treatment of choice. The most important predictor of 5-year survival is complete resection of the lesion.<sup>3,9</sup> Other important prognostic factors include the histopathological grade<sup>10</sup> and location of the tumor.<sup>11</sup> Data regarding prognostic value of tumor size are conflicting.<sup>10,12</sup> Median time to recurrence is 8 to 27 months.<sup>13</sup> Adjuvant radiotherapy is indicated for high grade tumors and margin-positive resections.<sup>9</sup>

Very little literature is available on the role of EUS in the diagnostic work up of this tumor. To the best of our knowledge, there is one case report of EUS-FNA diagnosis of IVC LMS during evaluation of a large right retro-peritoneal tumor.<sup>14</sup> EUS can be very useful in confirming relation of tumor to IVC and providing tissue diagnosis – as was the case in this patient. It's role in diagnosis of incidental finding of IVC LMS during EUS for other indications needs to be ascertained. It is likely that EUS is perhaps most helpful in segment II lesions.

## CONCLUSION

LMS of the IVC is a rare tumor that presents a diagnostic challenge. It should be considered in the differential of retro-peritoneal tumors, as survival improves with early detection and resection. EUS can be helpful in the diagnostic evaluation. ■

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