The Rare Case of a Leiomyosarcoma Originating From the Right Gonadal Vein

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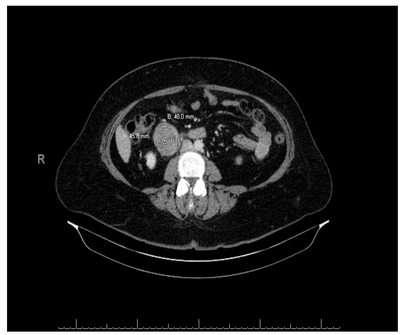
INTRODUCTION

-The most common site for leiomyosarcomas (LMS) to originate is within the endometrium of the uterine wall. LMS may also originate from blood vessels and, the most common site is the inferior vena cava and secondly the renal veins. There had been only ten cases, as of 2015, reported in literature that documented LMS with a gonadal vein origin and even fewer reporting involvement with the right gonadal vein.

CASE PRESENTATION

-Our patient is a 61-year-old female who had previously undergone a right-sided nephrectomy for renal cell carcinoma. During routine surveillance, she was noted to have a new abdominal mass to the right of the inferior vena cava and inferior to the right renal vein. This was not seen on imaging 5 months prior. Computed tomography of the mass described it as circular in appearance with dimensions of 62mm x 48mm x 46mm. Biopsy of the mass confirmed the diagnosis of a leiomyosarcoma. The decision was then made to proceed with surgical excision. There were no intraoperative findings of sarcomatosis or involvement of the inferior vena cava, however significant involvement of the right gonadal vein was noted requiring en bloc resection with the mass.



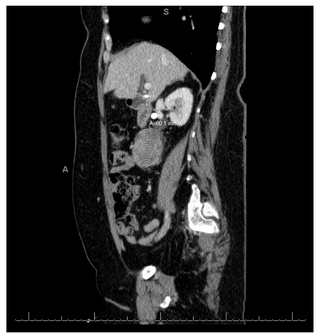


Fig 1 and 2) CT scan of Right Gonadal Vein Leiomyosarcoma

LEIOMYOSARCOMA (LMS)

-Due to the aggressive nature and most common positioning being retroperitoneal and involving major blood vessels, venous leiomyosarcomas pose a unique challenge for both early diagnosis and treatment. Though these tumors are commonly found incidentally in asymptomatic patients, most patients present with advanced, symptomatic disease. One can easily understand an increased morbidity and mortality associated with large tumors involving the vena cava. However, according to Gage, et. Al. venous leiomyosarcomas not involving the vena cava still only hold a 32% four-year survival rate. Vascular leiomyosarcomas affect around one in one hundred thousand people. This disease carries a high mortality rate due to its stealthily and invasive nature. Currently no routine screening tests exist leaving incidental finding or late presentation the current options for diagnosis. Tumor markers, like CA-125 among others, and CT imaging illuminate this sarcoma. Studies show the benefit to treatment of vLMS. When compared to uterine LMS, the five-year survival rate is less in vLMS at 35% compared to 42% in uterine LMS but with proper treatment of each, the ten-year survival rate with vLMS is greater than that of uterine LMS (41% vs 27% respectively). Therefore, when identified early enough, treatment is effective. With the radiation exposure CT's present, this might only be deemed necessary if there is a genetic component at play. This component is real and plays a large factor in the rate of Leiomyosarcomas in individuals based on family history of cancer. Based on data from over 2.9 million patients, there were 867 diagnoses of LMS. Of those 867 diagnoses, 291 have a family history of cancer, that being 33.5% and 90 have a mother who had cancer 10.5%. These averages are much higher than that of the general public of 4% and 1.2% respectively

CONCLUSION

-Final pathology confirmed negative margins had been achieved and the diagnosis of right gonadal vein leiomyosarcoma was made with final dimensions being 64mm x52mm x42mm. The patient had an uneventful post-operative course and has since been seen in follow up for nearly two years with no evidence of recurrence.

-Due to the aggressive nature and retroperitoneal location, venous leiomyosarcomas pose a unique challenge for both diagnosis and treatment. This case is unique with the origin of the LMS being the gonadal vein. This has rarely been reported in the literature, but one publication reported venous leiomyosarcomas not involving the vena cava, such as this case, only hold a 32% four-year survival rate. Although case reports are rare, it is crucial to consider venous leiomyosarcomas in the differential diagnosis when evaluating retroperitoneal tumors due to their significant lethality. More reports are needed in the future to add to the body of literature surrounding this rare diagnosis.

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