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# Rare Presentation of High Grade Retroperitoneal Angiosarcoma in a Patient Status Post Endovascular Aortic Repair (EVAR)

Jairo Espinosa M.D.

*Western Michigan University Homer Stryker M.D. School of Medicine*

Alan Hifko

*Western Michigan University Homer Stryker M.D. School of Medicine*

Julia Miladore M.D.

*Western Michigan Homer Stryker M.D. School of Medicine*

John T. Collins

*Borgess Medical Center*

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# Rare Presentation of High Grade Retroperitoneal Angiosarcoma in a Patient Status Post Endovascular Aortic Repair

Jairo A. Espinosa MD, Alan Hifko, Julia Miladore MD, John T. Collins MD FACS

## INTRODUCTION

Primary angiosarcoma of the aorta is a rare malignancy arising from the aorta's endothelial lining. The most common presentation is claudication, however in the presence of an endograft a tumor originating in the aneurysmal sac may remain undetected and mimic common graft pathology<sup>1,2</sup>. Diagnosis is typically delayed due to vague clinical presentation and imaging being indistinguishable from common complications of endografts such as leaks, infections or hematomas. We report a case of primary angiosarcoma of the aorta after endovascular repair of an abdominal aortic aneurysm. This case report describes the sixth case in the literature of primary angiosarcoma arising after the placement of an endovascular stent<sup>2-5</sup>.

## CASE PRESENTATION

A 75 year-old male with a previous history of endovascular aortic repair for a AAA six years prior presented to the emergency room with signs and symptoms concerning for bowel obstruction. He elicited abdominal pain, nausea, emesis, obstipation and was anemic and had leukocytosis. A CT angiogram of the abdomen and pelvis displayed a large mass encasing the infra-renal aorta, without active extravasation, that was externally compressing the second portion of the duodenum and causing a gastric outlet obstruction.



Figure 1. CT findings 6 months prior to presentation(a) and at presentation with large mass encapsulating abdominal aorta (b).

The mass extended into both iliac arteries with extension into a right iliac artery aneurysm. Review of the patient's chart revealed that this mass was not present six months prior. Patient was treated non-operatively with placement of a nasogastric tube. Concerns for a Type I or III Endoleak delayed the eventual CT-guided biopsy of the mass. Pathology revealed a malignant spindled and pleomorphic mass with extensive hemorrhage and necrosis consistent with high grade angiosarcoma. Specimen tested positive for vimentin, CD68, ERG, and CD31 on immunohistochemical staining. Patient did not wish to undergo aggressive treatment and was discharged to a hospice facility where he expired days later.

## CONCLUSION

Angiosarcoma is a rare malignancy representing 2% of all soft tissue sarcomas; associated with a poor prognosis with a median survival time of seven to eight months in advanced tumors.<sup>6,7,8</sup> Tumors larger than 5cm and those located in the retroperitoneum, such as in the case report that we present, are associated with a poor prognosis.<sup>9</sup> Doxorubicin is the mainstay of treatment for locally advanced tumors.<sup>10</sup> Paclitaxel has also been shown to be effective.<sup>6</sup> We present this care report to increase cognizance of this rare entity in the setting of a previously placed aortic endograft and avoid unnecessary surgical interventions.

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