# Challenges in diagnosing aortic leiomyosarcoma post endovascular repair of abdominal aortic aneurysm

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## ABSTRACT

Primary aortic tumors after endovascular aortic repair are rarely reported in the literature. Here, we report an elderly male with abdominal aortic leiomyosarcomas (LMS) after an endovascular aneurysm repair in 2012 for a 5-cm symptomatic abdominal aortic aneurysm using an Endurant II aortic stent graft (Medtronic, Minneapolis, Minn). The autopsy confirmed the aortic LMS after the patient rapidly deteriorated and succumbed to death. The vascular LMS are rapidly progressive and diagnostically challenging malignant soft tissue tumors with poor prognosis, which necessitates a strong clinical suspicion and attentiveness to radiologic signs for prompt diagnosis. (J Vasc Surg Cases and Innovative Techniques 2020;6:666-70.)

**Keywords:** Leiomyosarcoma; Abdominal aorta; Aortic aneurysm; Endovascular procedure; Case report

Vascular leiomyosarcomas (LMS) are rare subtypes (5%) of soft tissue tumors.[1] Virchow described the first large vessel LMS in 1871, and Brodowski described a primary aortic malignancy 2 year later in 1873.[2,3] The majority of vascular soft tissue tumors arise from the inferior vena cava.[4] A unique subset of this rare pathology is aortic sarcomas, and these can occur after endovascular aneurysm repair (EVAR), reports of which are scarce in the published literature. Here, we report a case of abdominal aortic LMS after an EVAR. The patient provided the consent to publish the case details and images.

## CASE REPORT

An 86-year-old man was referred to us in 2017 after he developed type Ib endoleaks in both iliac limbs after an EVAR. He underwent an EVAR in 2012 at another institution for a 5-cm symptomatic abdominal aortic aneurysm using an Endurant II aortic stent graft (Medtronic, Minneapolis, Minn). Although abdominal and low back pain were reported at the time, there was no evidence of impending rupture or evidence of LMS on the preoperative computed tomography angiography (CTA) (Fig 1)There was no significant past medical history before the EVAR. He was regularly followed up after the aneurysmal repair and follow-up CTA scans showed no evidence of any suspicious lesion in his aorta with stable aortic sac (Fig 2).

We found type Ib endoleaks associated with insidious sac expansion (>0.5 cm), which required bilateral iliac extensions. The CTA undertaken to investigate sac expansion did not show suspicious lesions or evidence of LMS at that time. The patient underwent a redo EVAR in our center successfully. In April 2019, follow-up duplex studies documented modulation of the aortic sac with a maximum diameter of 3.9 cm.

Three months later, the patient developed suprapubic and right flank pain. The pain was attributed to a ureteric stone and managed by double J stents. Unfortunately, the abdominal pain got worse after a month, and it was associated with lower back and left leg pain even at rest. During this 3-month period, he lost around 18 kgs weight. A CT scan was performed, which showed an increase in the aneurysmal sac size to 7.4 cm and changes in the contour of the vessel along with thrombosis of the left main limb of the graft, but without any evidence of endoleak (Fig 3, *A, B*). We suspected a graft infection; however, the patient's white cell count was 12,000/mm$^3$, and C-reactive protein level was 56 mg/dL. We managed his left leg with a home-based sequential compression device for peripheral arterial disease (ArtAssist, ACI Medical, San Marcos, Calif) and optimal medical therapy.

A positron emission tomography (PET) scan performed during this time showed high fluorodeoxyglucose avidity in the aorta, and active nodules in the right gluteal region (Fig 3, *C, D*) and right lung. A biopsy of the gluteal lesion documented LMS. Within 1 week, the patient developed a solid lesion in his right calf. The patient and his family opted for no intervention, including aortic biopsy, and decided to continue with palliative treatment only. Therefore, we could not perform an aortic biopsy or plan any surgical intervention, and a probable diagnosis of

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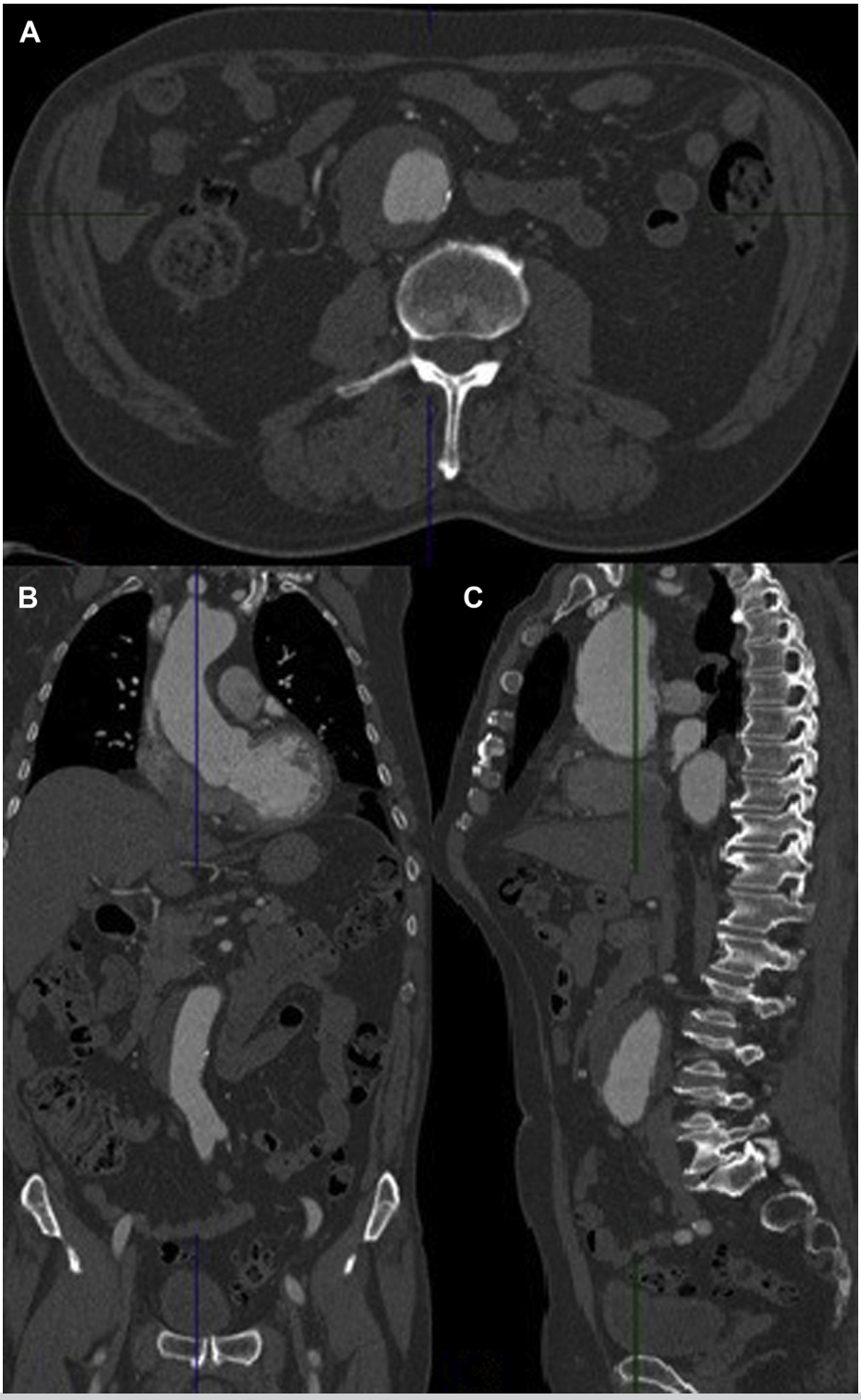


**Fig 1.** Preoperative computed tomography (CT) angiogram (CTA) in 2012 with innocent looking homogenous thrombus within the abdominal aortic aneurysm (AAA) and no alarming findings in the aortic sac or periaortic tissue **(A)** CTA axial view, documenting AAA with thrombus and no alarming findings in the aortic sac. **B,** CTA coronal view, documenting AAA with thrombus along the lateral aortic wall, the thrombus appears homogenous without any suspicion of any abnormality. **C,** CTA sagittal view, documenting AAA with thrombus along the anterior and posterior aortic wall, the thrombus seems to be homogenous without any suspicion of any abnormality.

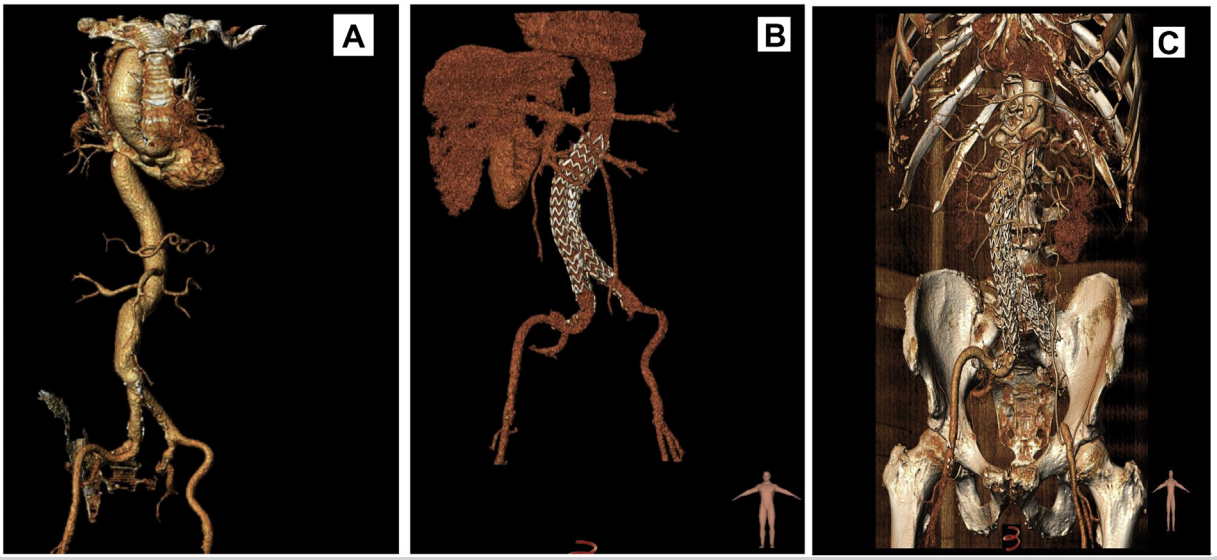


**Fig 2.** Preoperative and postoperative operative three-dimensional (3D) reconstructed scans. **A,** Preoperative 3D reconstruction scan in 2012. **B,** Post endovascular aneurysm repair (EVAR) follow-up scans in 2016, showing normal appearing aorta with the EVAR stent. **C,** Post redo EVAR and representation in 2019, showing lack of contrast through the left limb of the EVAR graft.

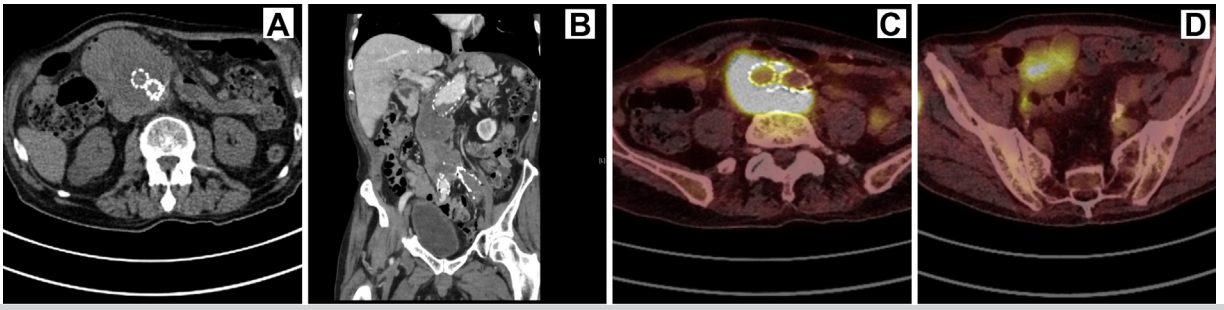


**Fig 3.** Abdominal/gluteal computed tomography (CT) and positron emission tomography (PET) scans. **A,** CT scan showing an increase in the sac size to 7.4 cm. **B,** CT scan showing the changes in the contour of the vessel along with thrombosis of the left main limb of the graft. **C, D,** PET scan showing high fluorodeoxyglucose avidity in the aorta and active nodule in the right gluteal region.

aortic sarcoma was considered after consulting with colleagues at international vascular centers; however, consent was obtained for the future autopsy.

The patient succumbed to death in January 2020, and his autopsy showed a substantial retroperitoneal mass (200 × 150 mm) with extensive necrosis. The tumor encircled the abdominal aorta, which contained the endovascular stent from the levels of renal arteries to the level of the aortic bifurcation. Multiple metastatic nodules were identified in the liver (the most significant being 30 × 25 mm in size) and the right lung. Histologically, the tumor was composed predominantly of spindle cells arranged in fascicles and contained elongated nuclei with nuclear pleomorphism and abundant mitoses (Fig 4, *A*). There was extensive tumor necrosis (Fig 4, *B*) within areas of epithelioid morphology (Fig 4, *C*). Immunohistochemistry revealed tumor cells positive for vimentin and smooth muscle actin (Fig 4, *D*) and negative for other markers, including cKit, S100, CD31, CD99, and CD34.

## DISCUSSION

Primary aortic tumors are rare vascular tumors that affect the descending thoracic aorta (34.9%), abdominal aorta (27.3%), thoracoabdominal aorta (26.5%), and ascending aorta and aortic arch (11.3%).[5] Iwabuchi[6] classified aortic tumors into either luminal or mural sarcomas. However, this histologic classification does not reflect patient prognosis, which is clinically relevant. Clinically, obstructive and embolic manifestations are more common in the intimal type.[7] In contrast, mural lesions have vague symptoms, like back or abdominal pain, or systemic signs, such as dyspnea, dysphagia, or weight loss. Occasionally, they are asymptomatic and can only be diagnosed after a considerable incremental increase in size.[8]

CT scans, magnetic resonance imaging, and/or PET scans are useful in diagnosis. Balaney et al[9] proposed a novel endovascular catheter-based aspiration technique

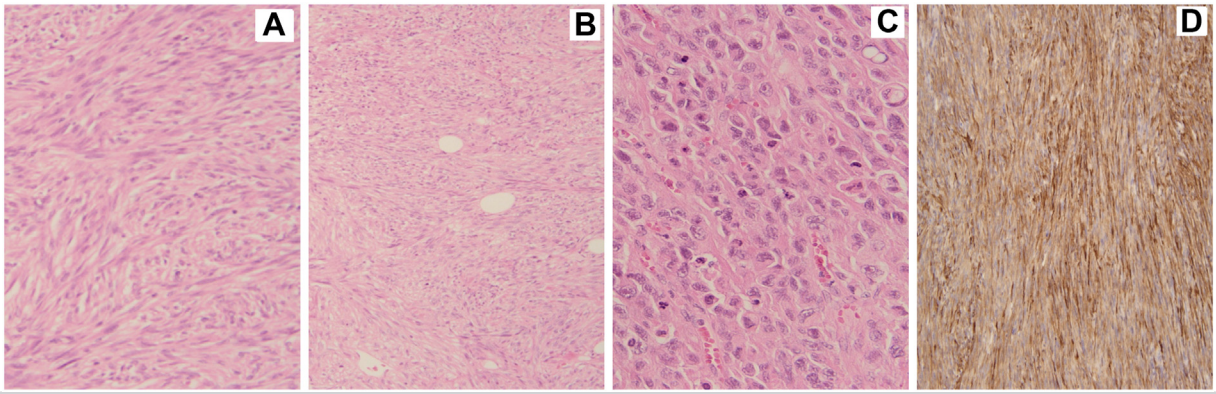


**Fig 4.** Hematoxylin and eosin histopathologic sections. **A,** The tumor is highly cellular and composed of spindle cells in sheath, showing nuclear atypia and abundant mitoses (magnication 40 HPF). **B,** Low-power view of the tumor with adjacent tumor necrosis (magnication 10 HPF). **C,** High-grade epithelioid appearance of the tumor (magnication 40 HPF). **D,** Actin immunohistochemistry showing the spindle and the epithelioid component of the tumor strongly positive for smooth muscle actin (magnication 10 HPF).

as a minimally invasive approach to diagnosis. The recommended therapy involves bloc resection of the tumor-involved aorta and surrounding tissues, followed by interposition grafting; however, because of the late diagnosis, this intervention is usually not feasible.[7]

In our case, the diagnosis was challenging and primarily based on strong clinical suspicion after the initial evaluation. After subsequent development of soft tissue metastases to the buttock and lung and persistent enlargement of the periaortic tissue on surveillance imaging, the buttock biopsy was later proven to represent high-grade angiosarcoma. However, aortic sarcoma was only confirmed after the autopsy owing to the patient's refusal to undergo an aortic biopsy.

The histopathologic pattern for vascular LMS is categorized by a proliferation of atypical smooth muscle cells with a large number of intermingled blood vessels.[10] The mitotic index is high, which is the most crucial pathologic feature on which a prognostic evaluation of vascular LMS can be based. Our patient had a highly aggressive tumor, as demonstrated by rapid clinical deterioration and evidence of de novo distant metastases in multiple areas at autopsy.

The use of foreign material has been associated with sarcoma formation. Jennings et al[11] described 46 cases of sarcoma associated with a variety of foreign bodies, including bullets, laparotomy sponges, and bone wax, with a latency period of 4 months to 63 years and postulated that implanted foreign material can give rise to any form of sarcoma. Burns et al[12] first described an association between a Dacron femoral graft and a rapidly growing fibrosarcoma in 1972. Specifically in relation to primary aortic tumors, several authors have reported a tentative association between Dacron graft implantation and neoplasm formation,[13] and the use of synthetic material has been associated with 65.7% of primary aortic angiosarcomas.[2] Ben-Izhak et al[14] suggested that a chronic inflammatory response around the graft can give rise to cellular malignant transformation. It has been postulated that pore size of implanted materials is related to the tumorigenesis, with smaller pore sizes having greater carcinogenic potential.[15,16] However, this finding has only been demonstrated in animal studies and not humans; it is, therefore, difficult to ascertain whether the type of material, for example, Dacron vs PTFE, influences malignant potential. With respect to aortic endografts, some investigators have suggested an association between endograft infection and subsequent malignancy, although this remains speculative.[13,17-20] Although causality cannot be proven, other authors, like in our case, testify that malignancy was not seen at the time of surgical graft implantation and was not seen on preoperative imaging at the time of endograft implantation. Furthermore, as in this case, the malignancy arises in the aortic wall adjacent to the graft.[21] There is an established association between long chain polymers in the polyester family compounds and the development of aortic sarcomas, and this presents a commonality between open and endovascular grafts.[22] However, nickel and titanium alloys, used in the metallic frame of endovascular stent grafts may also have carcinogenic potential, as described in the orthopedic literature.[23,24] As to whether the combination of polyester and metal increases the malignant potential of endografts compared with open surgical grafts is supposition.

The preliminary manifestation of aortic sarcoma after EVAR is usually missed owing to its similarity on imaging to type V endoleaks and infection of the graft, and both these differentials can delay the diagnosis. However, based on the reported literature, there are several signs that may raise suspicion of aortic sarcoma and serve as adjunctive pieces of evidence for aortic sarcoma rather than an infected graft. These include protrusive vegetation or nodular soft tissue components, which are devoid of atherosclerosis in a suspicious area; heterogeneous

thrombus; sudden enlargement of an excluded sac; and avid fluorodeoxyglucose uptake on PET scan. Appropriate imaging by CT scan-guided biopsy is recommended for diagnostic certainty for this rare pathology.[10,22,23]

## CONCLUSIONS

Aortic sarcomas, after EVAR, are uncommon findings with a poor prognosis. A strong clinical suspicion and attentiveness to radiologic signs may alert the clinician to neoplasm and enable prompt diagnosis. Multimodality imaging is mandatory to distinguish tumor enhancement from endoleaks, high-attenuation thrombus, or infection of the endovascular prosthesis.

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