

 **Case Report** 

Surgical Experience of a Case of Primary Leiomyosarcoma of the Left Common Iliac Artery That Presented as Acute Heart Failure Involving an Arteriovenous Fistula

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Primary leiomyosarcoma of the left common iliac artery with development of an arteriovenous fistula is extremely rare. Here we report the case of a 78-year-old woman with primary vascular leiomyosarcoma that presented as acute heart failure involving an arteriovenous fistula. She underwent surgery; however, after the diagnosis of leiomyosarcoma, she did not seek active treatment. She died 8 months after the surgery. This case report supports the need to document and follow-up a primary leiomyosarcoma of the iliac artery, especially when a case includes the presentation of an arteriovenous fistula (AVF). Vascular leiomyosarcoma should be included in the differential diagnosis of an AVF.


Keywords: primary leiomyosarcoma, arteriovenous fistula (AVF), common iliac artery

Introduction

A primary leiomyosarcoma of a major peripheral artery is very rare. Furthermore, the development of an arteriovenous fistula (AVF) in such a case is extremely rare. Here, we report a case of a primary leiomyosarcoma of the left common iliac artery that presented as acute heart failure involving an AVF. This case report supports the need to

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document and follow-up a primary leiomyosarcoma of the iliac artery, especially when a case includes the presentation of an AVF.

Case Report

A 78-year-old woman complaining of shortness of breath visited our hospital. Her dyspnea had worsened considerably during one week. Four days prior to her visit, she was diagnosed with bronchitis at a local clinic, and an antitussive and expectorant were prescribed; however, dyspnea and edema of the lower extremities developed. Investigation of the patient's history revealed that she had been taking medication for the treatment of hypertension and hyperlipidemia since she was 50 years old, and, at 70 years of age, she had experienced a dorsal compression fracture because of osteoporosis. Her body temperature was 36.9°C, her pulse was regular, her heart rate was 100 beats/min, and her blood pressure was 163/83 mmHg. Physical examination revealed edema of both feet. She was 153 cm tall and weighed 63 kg. Abnormal laboratory findings included high levels of D-dimer (8.04 µg/mL; normal range, <1.00 µg/mL) and brain natriuretic peptide (BNP, 542.5 pg/mL; normal range, <18.4 pg/mL). A chest X-ray image revealed cardiomegaly and bilateral lung congestion (cardiothoracic ratio 54%; normal range: <50%). Electrocardiography revealed normal sinus rhythm, with a negative T-wave at leads V1–V4. Echocardiography revealed hyperkinetic movement, with moderate tricuspid regurgitation, without the collapse of the inferior vena cava (IVC). Enhanced computed tomography (CT) indicated an AVF associated with a left iliac pseudoaneurysm (Figs. 1a and 1b). In addition, left hydronephrosis, uterine myoma, pulmonary artery thromboembolism of the right lower lobe branch, pleural effusion, and pulmonary congestion with right predominance were detected. We diagnosed dyspnea and edema of the lower extremities as acute right congestion heart failure with an AVF related to

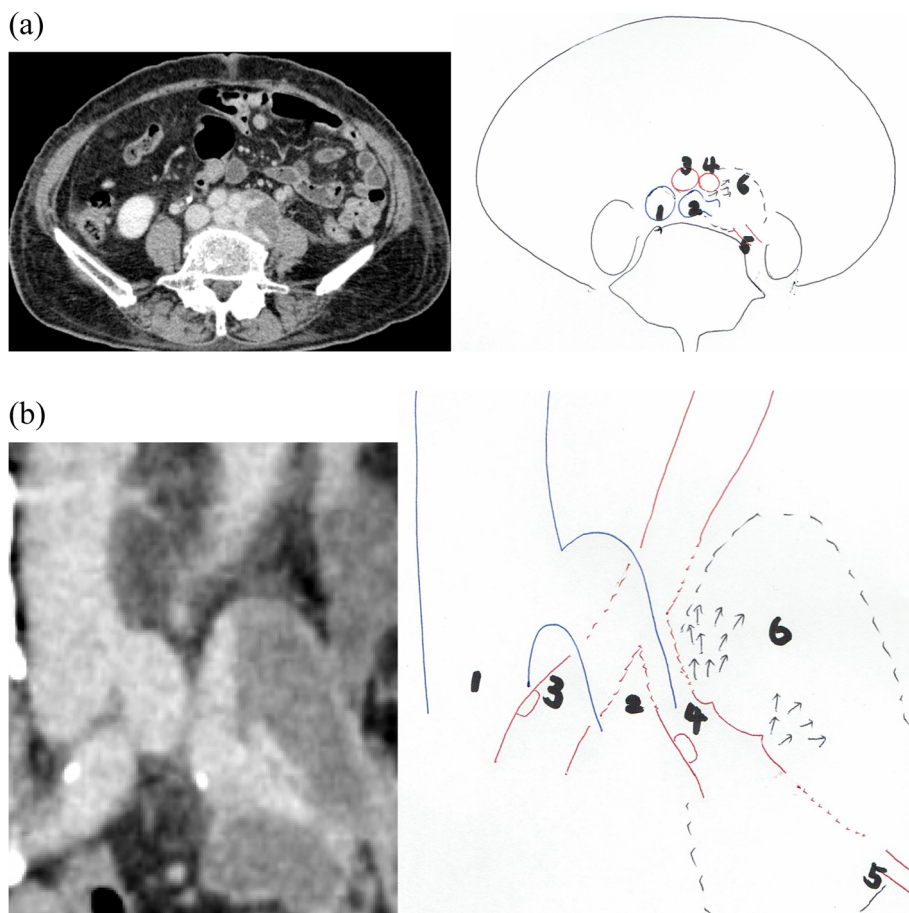


Fig. 1 Enhanced computed tomography (CT) (a: axial image, b: coronal image) revealed an arteriovenous fistula from the left iliac artery to the vein. The mass was suspected to be a pseudoaneurysm of the left iliac artery. Each schema was attached (1: right common iliac vein, 2: left common iliac vein, 3: right common iliac artery, 4: left common iliac artery, 5: left external iliac artery, 6: AV shunt).

the left common iliac pseudoaneurysm.

The patient was treated with intravenous diuretics, and further examinations (magnetic resonance imaging and vascular ultrasound) were conducted. However, a final diagnosis was not obtained, and we selected urgent abdominal surgery for AVF treatment.

Urgent surgery was performed through median laparotomy, following urethroscopy performed by a urologist. The findings indicated only pressure from outside the urinary tract, without a tumor in the left ureter. A ureteral stent was deployed in the left ureter for marking during surgery. After laparotomy, severe adhesion around the left iliac pseudoaneurysm was noted. We initially taped the terminal abdominal aorta and right common iliac artery using the tape. We could not tape the left common iliac artery. Further, we attempted to tape the IVC. But we had to abandon taping the IVC because we were afraid of the possibility of injury due to severe dilatation of the IVC. We subsequently cut open the sigmoid mesocolon and secured the peripheral sides of the left internal and exter-

nal iliac arteries. We judged that only digital compression would help to control bleeding from the AVF. Following heparin injection, we clamped the terminal abdominal aorta, right common iliac artery, and left internal and external arteries. On opening the wall of the pseudoaneurysm, a large amount of venous blood flowed, and shock was noted. During massive blood transfusion, we continued digital compression. We finally achieved complete hemostasis with various medical styptics. The AVF site had multiple fistulae of the venous wall. We submitted a specimen of the arterial wall for histological examination. Finally, we performed bypass from the right common femoral artery to the left common femoral artery, by using an 8-mm ringed prosthesis (FUSION Vascular Graft, Maquet, Germany) following the achievement of a stump for each artery (proximal sites of the left common iliac artery, external iliac artery, and internal iliac artery). After protamine administration, hemostasis was reviewed. After washing the area with a lot of warm saline, we closed the abdominal wall. The operation time was 636 min, and the

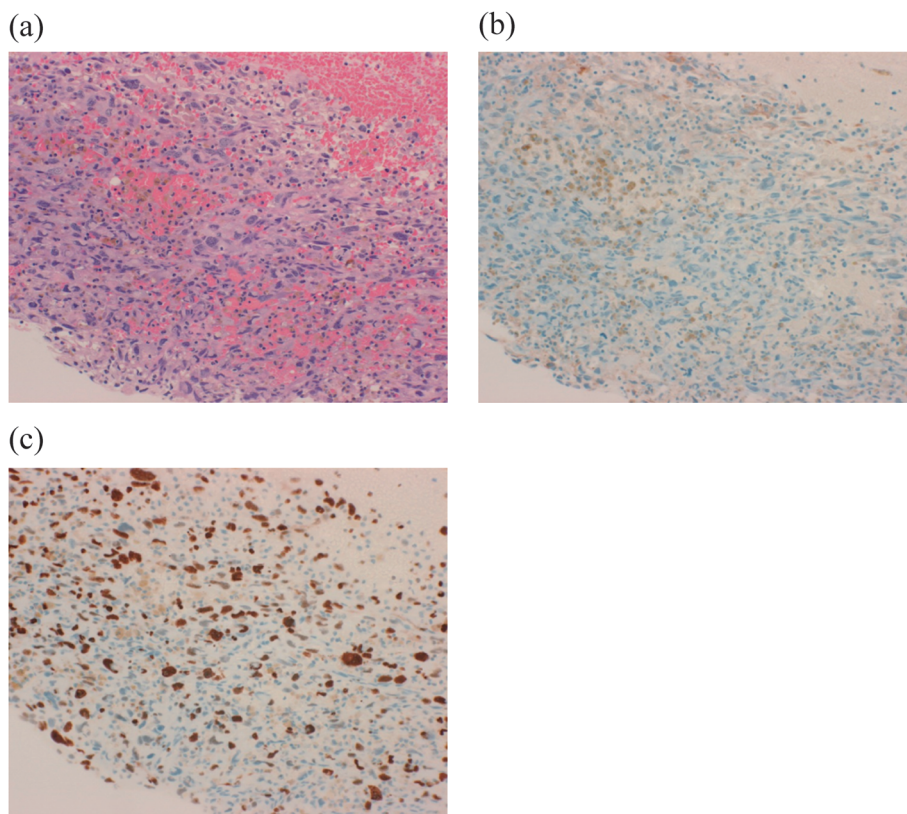


Fig. 2 Microscopic examination of the resected specimen (a): Hematoxylin and eosin staining of the aneurysmal wall revealed the proliferation of atypical spindle-shaped tumor cells, near the margin of the thrombi, that had hyperchromatic pleomorphic nuclei and abundant eosinophilic cytoplasm (magnification: 20 \times). Immunohistochemical examination of the resected specimen (b, c): Atypical spindle cells were positive for smooth muscle actin (b). The negative expression of CD34 and S-100 excludes an epithelial cell origin (data not shown). Therefore, we confirmed that these proliferating tumor cells were derived from smooth muscle cells. The MIB-1 labeling index was greater than 80% (very high) for the proliferating atypical cells of the resected aneurysmal wall (c).

amount of blood loss was 5,471 mL despite several blood transfusions being administered. The patient was extubated in the operating room. Her postoperative course was uneventful. Pathology examination of the surgical specimen revealed leiomyosarcoma; however, the patient did not seek active treatment. Histological examination of the resected aorta revealed colonization by spindle-shaped cells, with nuclei of various sizes, and accompanying acidophils (Fig. 2a). Immunohistology findings were as follows: smooth muscle actin (+), CD34 (-), c-kit (-), S-100 (-), and a MIB-1 index of more than 80% (Figs. 2b and 2c). The final diagnosis made by a pathologist was of primary leiomyosarcoma of the left iliac artery. Enhanced CT revealed no bleeding sites and indicated patency of the anti-anatomical bypass graft (Figs. 3a and 3b). On post-operative day 21, the patient was discharged, because she stubbornly refused further active treatment and expected palliative treatment; as a result, we expected her prognosis

to be very poor.

The patient lived well for three months after discharge. However, 3 months later, enhanced CT revealed tumor recurrence. Furthermore, 4 months later, CT revealed not only a large tumor but also multiple pulmonary metastases (Fig. 3c). She died 8 months after the vascular surgery.

Discussion

Auffermann¹⁾ first reported a patient with a leiomyosarcoma of a large artery. Several cases have been subsequently reported; however, primary tumors originating in the wall of large arteries and veins are exceedingly rare.^{2,3)} The exact etiology of leiomyosarcoma of arteries remains unknown. Vascular leiomyosarcoma involving veins is five times more common than leiomyosarcoma involving arteries. With regard to arterial leiomyosarcomas, several reports have documented the involvement of the pulmonary

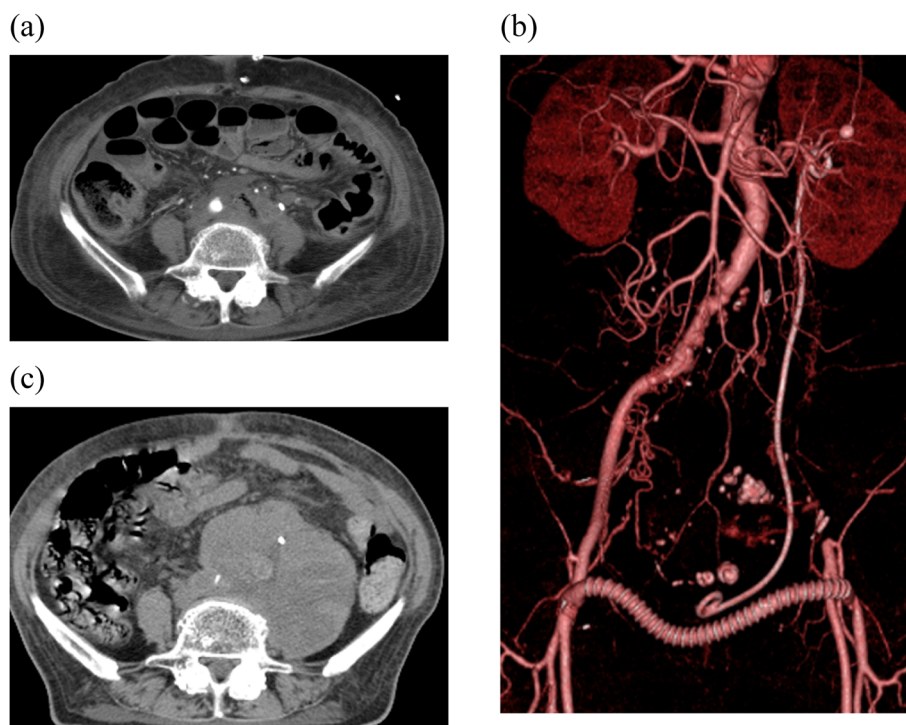


Fig. 3 Enhanced computed tomography (CT) revealed no bleeding sites (a) and indicated the patency of the anti-anatomical bypass graft (b). CT performed 7 months after the surgery revealed the recurrence of a large tumor (c). Additionally, there were multiple pulmonary metastases (data not shown).

artery, whereas several authors have reported the involvement of the aorta and renal, popliteal, splenic, subclavian, carotid, iliac, and femoral arteries.^{2,3} Among case reports in the literature, there is no report regarding a case of leiomyosarcoma with initial symptoms of a spontaneous AVF.

Brewster et al.⁴ reported that spontaneous rupture of an aortic or iliac aneurysm or erosion of an inflammatory or mycotic aneurysm into contiguous veins, including the IVC, iliac vein, and left renal vein, can result in a spontaneously acquired AVF. The authors mentioned that the incidence of AVF is 0.2%–1.3% among patients with abdominal aortic aneurysms and 3%–4% among those with ruptured aneurysms.⁴ No report has been published regarding spontaneous AVF associated with leiomyosarcoma. However, leiomyosarcoma was reported with a pulmonary and postoperative AVF,^{5,6} and a malignant mesenchymal tumor was reported to have occurred with a spontaneous AVF.⁷ The leiomyosarcoma of our case was incidentally diagnosed postoperatively through pathology examination.

Microscopically, spindle cells usually accompany subintimal fibrous changes. For our case, immunohistology examination revealed the presence of smooth muscle actin and a MIB-1 index of >80%. It is important to differentiate low-grade tumors from high-grade tumors, because their clinical management varies. Ki-67 is a widely

studied cell-proliferation marker, and MIB-1 is one of the most sensitive commercially available Ki-67-equivalent antibodies. MIB-1 can react with an epitope of the Ki-67 protein in formalin-fixed, paraffin-embedded sections, after microwave antigen retrieval. The MIB-1 labeling index was calculated as the percentage of MIB-1-positive nuclei. MIB-1 has been used as an operational marker of cell proliferation for various types of human tumors.⁸ For the present case, considering the absence of a primary tumor in other organs, the final diagnosis was of primary leiomyosarcoma of the left iliac artery.

According to Sakpal et al.,³ patient prognosis regarding vascular leiomyosarcoma is related to the stage at presentation and surgical resectability. Delay in diagnosis often makes en bloc surgical resection challenging. For our case, the patient rejected additional treatment; as a result, her prognosis was very poor.

Conclusion

We report a very rare case of primary leiomyosarcoma of the iliac artery that presented as acute heart failure involving an AVF. This case report supports the documentation of primary leiomyosarcoma in the iliac artery, especially when presenting with an AVF. Our patient died 8 months after surgery as a result of tumor recurrence. Vascular

leiomyosarcomas should be included in the differential diagnosis, especially when vascular surgery is required for an AVF.

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Disclosure Statement

The authors declare no conflicts of interest.

Additional Remarks

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Author Contributions

Conception, design, and draft of the manuscript: TA
Principal operating surgeon: TA
Consultant in charge of patient's care: TA, KD, HG
Assistance in the operation and collection of various imaging data: KD, HG

Revision of the manuscript: TA, MT

Approval of the final manuscript: all authors

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