

Phlegmasia cerulea dolens as an initial manifestation of a fistula between a ruptured iliac artery aneurysm and the iliac vein

Kazumasa Hanada, MD, Kota Yamamoto, MD, PhD, Takafumi Akai, MD, PhD, Ryosuke Taniguchi, MD, PhD, Toshio Takayama, MD, PhD, and Katsuyuki Hoshina, MD, PhD, Tokyo, Japan

ABSTRACT

Phlegmasia cerulea dolens (PCD) is caused by obstruction of limb venous return that may result in venous gangrene and limb loss. We present a case of a fistula between a ruptured right common iliac artery aneurysm and the left common iliac vein (ilioiliac arteriovenous fistula [AVF]), which initially manifested as left PCD and acute renal failure. Resection of the aneurysm and repair of the AVF immediately improved the PCD and acute renal failure. We should be aware that an iliac AVF might present as PCD and should keep this in mind. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:41-4.)

Keywords: Phlegmasia cerulea dolens; Arteriovenous fistula; Acute renal failure

Phlegmasia cerulea dolens (PCD) is characterized by sudden swelling, ischemic pain, and limb cyanosis. It is caused by obstruction of venous return, including microvascular collateral vessels, usually because of massive venous thrombosis, and it has high amputation and mortality rates.¹ An arteriovenous fistula (AVF) between the iliac artery and vein is a rare cause of PCD. A common iliac artery aneurysm (CIAA) may cause abdominal pain, ureteral stenosis, and deep venous thrombosis, and its rupture is usually a life-threatening emergency. CIAA rupture into the iliac vein or inferior vena cava results in an AVF.²

CASE REPORT

We present a case of PCD as an initial manifestation of a fistula between a ruptured right CIAA and the left common iliac vein (CIV) that recovered immediately after surgery (Fig 1, A and B). The patient has provided consent for the publication of the case details and images.

A 44-year-old man with a history of hypertension complaining of left limb swelling and pain that had developed for several hours (Fig 1, A) was admitted to the emergency room. He was a Japanese grocer, who smoked cigarettes and frequently drank alcohol. He had no history of recent surgery, trauma, or arteriosclerosis. There was no family history of cardiovascular disease.

On examination, his blood pressure was 119/83 mm Hg, pulse rate was 118 beats/minute, and body temperature was 36.4°C.

His abdomen was soft and flat without pain. A pulsatile mass was palpable in his right lower abdominal quadrant. A continuous bruit was noted at the lower abdominal area. His left limb was swollen and cyanotic with pain. Distal pulses of the left limb (the dorsalis pedis and posterior tibial arteries) were barely palpable, and the left femoral and popliteal artery pulses were 1+. All the other pulses were 2+. Blood tests showed an increased white blood cell count ($27.4 \times 10^9/L$), C-reactive protein level (4.8 mg/dL), and creatinine level (259 $\mu\text{mol/L}$). The estimated glomerular filtration rate was 17.2 mL/min/1.73 m². Chest radiography did not show pulmonary congestion. Transthoracic echocardiography findings did not indicate cardiac failure. Acute renal failure (ARF) of unknown etiology discouraged us from using contrast medium. Plain computed tomography scanning was performed, which showed bilateral CIAAs. The right CIAA was 56 mm in diameter and was suspected of compressing the left CIV (Fig 2). The aorta was 27 mm in diameter and the left CIAA was 25 mm. There were no other aneurysms. Duplex ultrasound examination showed a fistulous flow into the left CIV as a color mosaic. There was no sign of deep venous thrombosis. The preoperative diagnosis was PCD owing to a fistula between the ruptured right CIAA and left CIV. We suspected an infected arterial aneurysm because of age, sudden onset, and inflammation.

Emergency open surgery was performed for life and limb salvage. Under general anesthesia, a midline laparotomy was performed and the entire abdomen was explored. No active bleeding or hematoma was observed. Control of the infrarenal aorta and the bilateral external and internal iliac arteries was obtained (Fig 3, A). Considering the possibility of infection, we prepared for a retroperitoneal detour from the left retroperitoneum to the right external iliac artery across the front of the bladder. On opening the right CIAA, we identified an AVF (10 × 8 mm) with venous back bleeding from the left CIV. Direct closure of the fistula was performed. Because there were no obvious signs of infection, an aortobi-iliac Dacron graft was placed in situ (Fig 3, B). Intraoperative blood loss was 1790 mL, and the operative time was 234 minutes.

The condition of the left limb improved immediately after the surgery (Fig 1, B). For the prevention of thrombosis, intravenous

From the Division of Vascular Surgery, Department of Surgery, Graduate School of Medicine, The University of Tokyo.

Author conflict of interest: none.

Correspondence: Kota Yamamoto, MD, PhD, Division of Vascular Surgery, Department of Surgery, Graduate School of Medicine, The University of Tokyo, 7-3-1 Hongo, Bunkyo-ku, Tokyo 113-8655, Japan (e-mail: kmaysyamamo@gmail.com).

The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

© 2018 The Author(s). Published by Elsevier Inc. on behalf of Society for Vascular Surgery. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jvscit.2018.11.004>



Fig. 1. Photographs of the patient's limbs. **A**, Preoperatively, the left limb is swollen and cyanotic. **B**, Postoperatively, the condition of the left limb shows improvement.

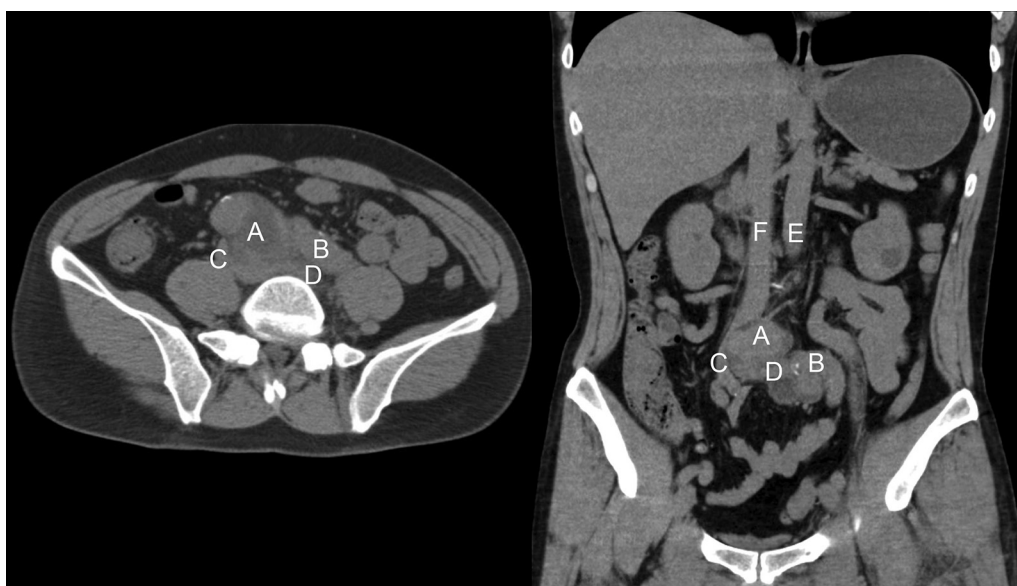


Fig. 2. Preoperative noncontrast-enhanced computed tomography. **A**, Right common iliac artery aneurysm (CIAA). **B**, Left CIAA. **C**, Right common iliac vein. **D**, Compressed left common iliac vein. The border is unclear. **E**, Aorta. **F**, Inferior vena cava.

heparin therapy was administered until postoperative day 4. The creatinine level and estimated glomerular filtration rate improved to within the normal ranges after 3 days. Preoperative blood culture and intraoperative aortic culture were negative. Rare infectious

microorganisms, such as *Coxiella burnetii*, *Mycobacterium*, and fungi, were also negative. No complications occurred, and he was discharged on postoperative day 17. In Japan, patients tend to stay in the hospital until they experience relief.

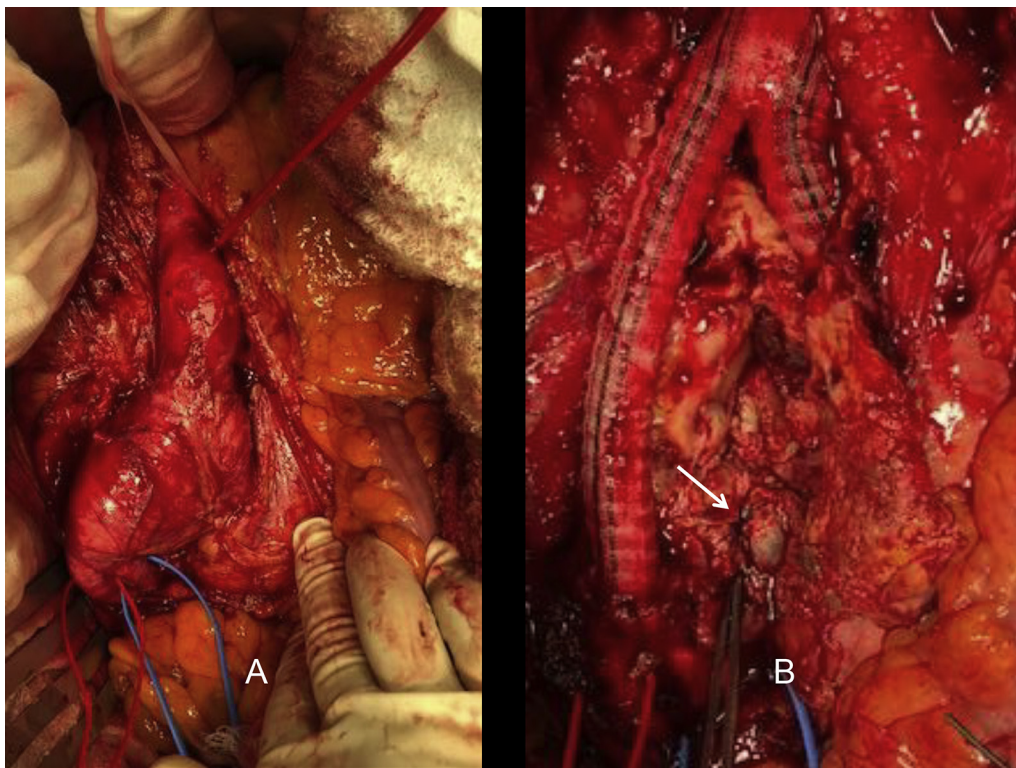


Fig. 3. Intraoperative photographs. **A**, Right common iliac artery aneurysm (CIAA). **B**, After reconstruction. Closure of the left common iliac vein is indicated by the *arrow*.

DISCUSSION

According to previous reports, 29% to 62% of CIAAs were symptomatic and 5% to 29% ruptured. The median diameter of ruptured CIAAs was 6 cm.² An iliac AVF is caused by aneurysmal rupture, trauma, surgery, malignancy, or infection, and it occurs in less than 1% of all cases of CIAAs and in 3% to 4% of cases of ruptured aneurysms.^{2,3} It is classically reported to involve high-output heart failure, abdominal and/or lower back pain, a pulsatile mass with a bruit, and signs of local venous hypertension. However, these features have been reported in only 20% to 50% of patients.^{4,5} An iliac AVF can cause ARF owing to both decreased renal perfusion from low systemic vascular resistance and decreased glomerular filtration pressure from increased renal congestion associated with increased inferior vena cava pressure.⁶ Only repair of the AVF can improve this type of ARF. Surgical mortality in patients with a symptomatic AVF can be as high as 22% to 51%.⁷ Contrast-enhanced computed tomography angiography is the most used modality for diagnosis and treatment planning. Magnetic resonance angiography is useful for diagnosis if time permits. A duplex color ultrasound scan can promptly detect an AVF without invasion.⁸

PCD classically results from venous thrombosis in relation to malignancy and thromboembolic disorders. PCD is best treated with prompt anticoagulation and a

strategy of thrombus removal within 24 hours.⁹⁻¹¹ However, an AVF with or without thrombosis can also cause PCD. A case of PCD caused by an aortocaval fistula (ACF) with right iliac vein thrombosis was reported.¹² Closure of the fistula, implantation of an aortobi-iliac Dacron graft, and thrombectomy resolved the PCD. In our case, thrombosis was not detected, but clinical findings in the left limb suggested PCD. The obstruction of venous outflow could be explained by both compression (the right CIAA compressed the left CIV) and the AVF blocking venous return. Resection of the CIAA and repair of the fistula resolved the occlusion.

We considered endovascular aneurysmal repair (EVAR), but the ARF prevented the selection of EVAR. If renal function allows, EVAR is minimally invasive and can resolve the iliac AVF and the PCD in the short term. If a mycotic aneurysm is found to be associated with the AVF, elective open repair can be planned later. However, we should remember that an endoleak (type II, particularly owing to venous bleeding through the AVF) might enlarge the ruptured aneurysm or induce persistent AVF.¹³

We considered the possibility of infection and prepared a retroperitoneal detour; however, we did not use it, because there was no clear intraoperative sign of infection. A case of an infected CIAA with an ACF has been reported, and it was reconstructed with a reversed

femoral vein graft.¹⁴ If infection is suspected intraoperatively, a detour or an autologous graft or homograft can be considered.

In conclusion, we should keep in mind that an iliac AVF might present as PCD and ARF. We should perform contrast-enhanced computed tomography angiography or magnetic resonance angiography according to the degree of renal dysfunction and urgency. We should confirm the presence of an AVF by auscultation or color Doppler ultrasound examination and, if possible, examine the AVF with a duplex scan. After the diagnosis, we should perform repair. Open surgery or EVAR can be selected taking into account preoperative considerations.

REFERENCES

1. Perkins JM, Magee TR, Galland RB. Phlegmasia caerulea dolens and venous gangrene. *Br J Surg* 1996;83:19-23.
2. Huang Y, Gloviczki P, Duncan AA, Kalra M, Hoskin TL, Oderich GS, et al. Common iliac artery aneurysm: expansion rate and results of open surgical and endovascular repair. *J Vasc Surg* 2008;47:1203-10; discussion: 1210-1.
3. Davis PM, Gloviczki P, Cherry KJ Jr, Toomey BJ, Stanson AW, Bower TC, et al. Aorto-caval and ilio-iliac arteriovenous fistulae. *Am J Surg* 1998;176:115-8.
4. McAuley CE, Peitzman AB, deVries EJ, Silver MR, Steed DL, Webster MW. The syndrome of spontaneous iliac arteriovenous fistula: a distinct clinical and pathophysiologic entity. *Surgery* 1986;99:373-7.
5. Brewster DC, Cambria RP, Moncure AC, Darling RC, LaMuraglia GM, Geller SC, et al. Aortocaval and iliac arteriovenous fistulas: recognition and treatment. *J Vasc Surg* 1991;13:253-64; discussion: 264-5.
6. Yamazaki K, Sakakima M, Nagakura Y, Hashimoto H, Tashiro T, Miwa M, et al. Left common iliac artery aneurysms with arteriovenous fistula that caused acute kidney injury. *Jpn J Nephrol* 2017;59:1252-7.
7. Goto T, Enmoto T, Akimoto K. Diagnosis of an ilio-iliac arteriovenous fistula by multidetector row computed tomography and surgical repair. *Interact Cardiovasc Thorac Surg* 2009;8:387-9.
8. Huang PL, Chua S, Guo GB, Fu M. Mycotic aneurysm leading to iliac arteriovenous fistula diagnosed by vascular duplex color scan. *J Ultrasound Med* 1998;17:513-6.
9. Kearon C, Kahn SR, Agnelli G, Goldhaber S, Raskob GE, Comerota AJ. Antithrombotic therapy for venous thromboembolic disease: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines (8th Edition). *Chest* 2008;133(6 Suppl):454S-55S.
10. Jaff MR, McMurtry MS, Archer SL, Cushman M, Goldenberg N, Goldhaber SZ, et al. Management of massive and submassive pulmonary embolism, iliofemoral deep vein thrombosis, and chronic thromboembolic pulmonary hypertension: a scientific statement from the American Heart Association. *Circulation* 2011;123:1788-830.
11. Meissner MH, Gloviczki P, Comerota AJ, Dalsing MC, Eklof BC, Gillespie DL, et al. Early thrombus removal strategies for acute deep venous thrombosis: clinical practice guidelines of the Society for Vascular Surgery and the American Venous Forum. *J Vasc Surg* 2012;55:1449-62.
12. Ben Abdallah I, El Batti S, da Costa JB, Julia P, Alsac JM. Phlegmasia cerulea dolens as an unusual presentation of ruptured abdominal aortic aneurysm into the inferior vena cava. *Ann Vasc Surg* 2017;40:298.e1-4.
13. Orion KC, Beaulieu RJ, Black JH 3rd. Aortocaval fistula: is endovascular repair the preferred solution? *Ann Vasc Surg* 2016;31:221-8.
14. Sasahashi N, Hamazaki M, Asada H, Kataoka T, Hamanaka K, Nishiyama K. Infected iliac artery aneurysm with aortocaval fistula. *Acute Med Surg* 2016;3:400-3.

Submitted Jun 12, 2018; accepted Nov 5, 2018.