

Successful staged management of a spontaneous iliac vein rupture associated with May–Thurner syndrome: a case report

Yuji Nishimoto *, Masanao Toma , Kohei Iwasa, and Yukihito Sato

Department of Cardiology, Hyogo Prefectural Amagasaki General Medical Center, 2-17-77 Higashinaniwa-cho, Amagasaki 660-8550, Japan

Received 19 March 2021; first decision 20 April 2021; accepted 22 July 2021; online publish-ahead-of-print 4 August 2021

Background

Spontaneous iliac vein ruptures have only been reported in approximately 50 cases. An accurate preoperative diagnosis is difficult even with contrast-enhanced computed tomography (CT), and the operative mortality and morbidity rates are quite high. The cause of spontaneous iliac vein ruptures and their optimal diagnosis and management remain unclear.

Case summary

A 69-year-old woman without a history of prior trauma presented with low back pain, left lower limb swelling, and hypovolaemic shock. An initial contrast-enhanced CT revealed a large retroperitoneal haematoma without arterial extravasation. Her blood pressure dropped again under a noradrenaline administration. A second venous phase contrast-enhanced CT revealed venous extravasation in the external iliac vein with a suspected compression of the common iliac vein (May–Thurner syndrome) and deep vein thrombosis (DVT). Her haemodynamics were stabilized whilst a laparotomy was arranged. An inferior vena cava (IVC) filter was placed due to concerns about rebleeding with initiating anticoagulation therapy. Given the failed conservative management, elective endovascular treatment (EVT) was performed including percutaneous Fogarty venous thrombectomy and placement of self-expanding and covered stents. After the intervention, the lower limb swelling significantly improved under oral anticoagulation therapy, and the IVC filter was retrieved. At the 3-month follow-up, the lower limb swelling completely disappeared, and the contrast-enhanced CT demonstrated the complete disappearance of the retroperitoneal haematoma and DVT.

Discussion

This case provided not only the potential value of the venous phase contrast-enhanced CT in diagnosing a spontaneous iliac vein rupture but also the potential benefit of conservative management followed by elective EVT.

Keywords

Spontaneous iliac vein rupture • Retroperitoneal haematoma • May–Thurner syndrome • Endovascular procedures • Thrombectomy • Case report

Introduction

Spontaneous iliac vein ruptures, which present with a life-threatening retroperitoneal haematoma, have only been reported in approximately 50 cases.¹ Most patients are middle-aged women, most

present with a left-sided iliac vein rupture and concomitant deep vein thrombosis (DVT), and approximately one-third have May–Thurner syndrome, which is defined as a symptomatic proximal compression of the iliac vein between the overlying iliac arteries and lumbar vertebrae.² An accurate pre-operative diagnosis is difficult even with

* Corresponding author. Tel: +81-6-6480-7000, Fax: +81-6-6480-7001, E-mail: yuji.nishimoto@gmail.com

Handling Editor: Konstantinos Stathogiannis

Peer-reviewers: Domenico Filomena; Ugur Canpolat; Erica Tirr

Compliance Editor: Gemina Doolub

Supplementary Material Editor: Ayse Djahit

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Learning points

- An accurate pre-operative diagnosis of a spontaneous iliac vein rupture is difficult even with contrast-enhanced computed tomography (CT), and the operative mortality and morbidity rates are quite high.
- Dynamic contrast-enhanced CT, including venous phase, has a potential diagnostic value.
- Conservative management followed by elective endovascular treatment can be effective.

contrast-enhanced computed tomography (CT), and the operative mortality and morbidity rates are quite high (16.7% and 50%, respectively).³ The cause of spontaneous iliac vein ruptures and their optimal diagnosis and management remain unclear. We report a case of a spontaneous iliac vein rupture with May–Thurner syndrome, which was accurately diagnosed with venous phase contrast-enhanced CT and successfully managed with conservative management followed by endovascular treatment (EVT).

Timeline

Day 0	Patient was referred with low back pain, left lower limb swelling, and hypovolaemic shock. Prompt fluid resuscitation and blood transfusions were performed.
10 min	An initial contrast-enhanced computed tomography (CT) revealed a large retroperitoneal haematoma without arterial extravasation.
2 h	The blood pressure dropped again under a noradrenaline administration.
3 h	A second venous phase contrast-enhanced CT revealed venous extravasation in the external iliac vein with suspected May–Thurner syndrome.
4 h	The haemodynamics stabilized whilst a laparotomy was arranged. An inferior vena cava (IVC) filter was placed.
Day 7	Parenteral anticoagulation therapy was initiated. The left lower limb swelling did not improve.
Day 11	The follow-up CT showed a further increased retroperitoneal haematoma.
Day 14	Elective endovascular treatment was performed.
Day 20	The lower limb swelling significantly improved under oral anticoagulation therapy. The follow-up CT showed a decreased retroperitoneal haematoma and nearly dissolved venous thrombi.
Day 21	The IVC filter was retrieved.
Day 28	Patient was discharged home.
3 months	The lower limb swelling had completely disappeared. The follow-up CT demonstrated the complete disappearance of the retroperitoneal haematoma and venous thrombi.

Case presentation

A 69-year-old woman without a history of prior trauma or gynaecological disease was referred to our emergency department with left lower limb pain and swelling when she woke up, followed by low back pain and light-headedness the next day. On the initial assessment, she was hypotensive and tachycardic. Her heart rate was 156 b.p.m., blood pressure 99/73 mmHg, respiratory rate 31 per min, and the saturation was not measured on 10 L of oxygen. The physical examination was significant for perspiration and left leg swelling. Fortunately, prompt fluid resuscitation and blood transfusions temporarily stabilized the haemodynamics. The blood test results revealed an elevated D-dimer level (55.4 µg/mL, normal value; <1.0 µg/mL) and high lactate concentration (12.5 mmol/L; normal value; 0.5–2.0 mmol/L). A ruptured aortic aneurysm (AAA) was suspected and an initial arterial phase contrast-enhanced CT was organized. This showed no AAA or active arterial extravasation but did demonstrate a large retroperitoneal haematoma (Figure 1A; Video 1). Her blood pressure dropped again under a noradrenaline administration. A second dynamic contrast-enhanced CT, including venous phase, was performed due to leg swelling suggesting a DVT and revealed an increased retroperitoneal haematoma (Figure 1B; Video 2) with a suspected compression of the common iliac vein (CIV) between the left internal iliac artery and fifth lumbar vertebra (May–Thurner syndrome) (Figure 1D), venous extravasation in the external iliac vein (EIV) (Figure 1B and C), DVT below the CIV (Figure 1C, E, and F), and no pulmonary embolism. Haemostasis may have been achieved by a tamponade effect of the pelvis, and her haemodynamics were stabilized whilst a laparotomy was arranged. As such, we opted to manage the patient conservatively in the first instance, and she did not undergo surgery. An inferior vena cava (IVC) filter was placed because we were hesitant to initiate anticoagulation therapy due to a life-threatening bleeding event. Low-dose unfractionated heparin was initiated after confirming that there was no progression of the anaemia for 7 days after the initial presentation because the left lower limb swelling was very severe and could cause post-thrombotic syndrome without anticoagulation therapy. However, not only did the left lower limb swelling not improve but also the retroperitoneal haematoma further increased in the follow-up CT. A team discussion with the vascular surgeons concluded a percutaneous Fogarty venous thrombectomy would be performed to remove the thrombi as much as possible, followed by the placement of self-expanding stents including a covered stent due to the complicated co-existence of a compression of the CIV, DVT below the CIV, and rupture in the EIV (Figure 2A and B; Supplementary material online, Video S1).

After placement of a 5-Fr sheath in the left common femoral vein under ultrasonography guidance, venography was performed via the left great saphenous vein and revealed a lot of thrombi throughout the iliac vein (Figure 3A). A guidewire easily passed the lesions, and intravascular ultrasound (IVUS) confirmed the compression of the CIV (Figure 3B–D; Supplementary material online, Video S2). The 5-Fr sheath was percutaneously replaced with a 22-Fr DrySeal sheath (W. L. Gore, Flagstaff, AZ, USA) under local anaesthesia, and a thrombectomy using a 4-Fr Fogarty balloon catheter (Edwards Lifesciences, Irvine, CA, USA) with a softly inflated balloon was performed (Figure 3E). A sufficient amount of thrombi could not be removed (Figure 3F) and angiography revealed venous extravasation (Figure 3G). A 9 mm

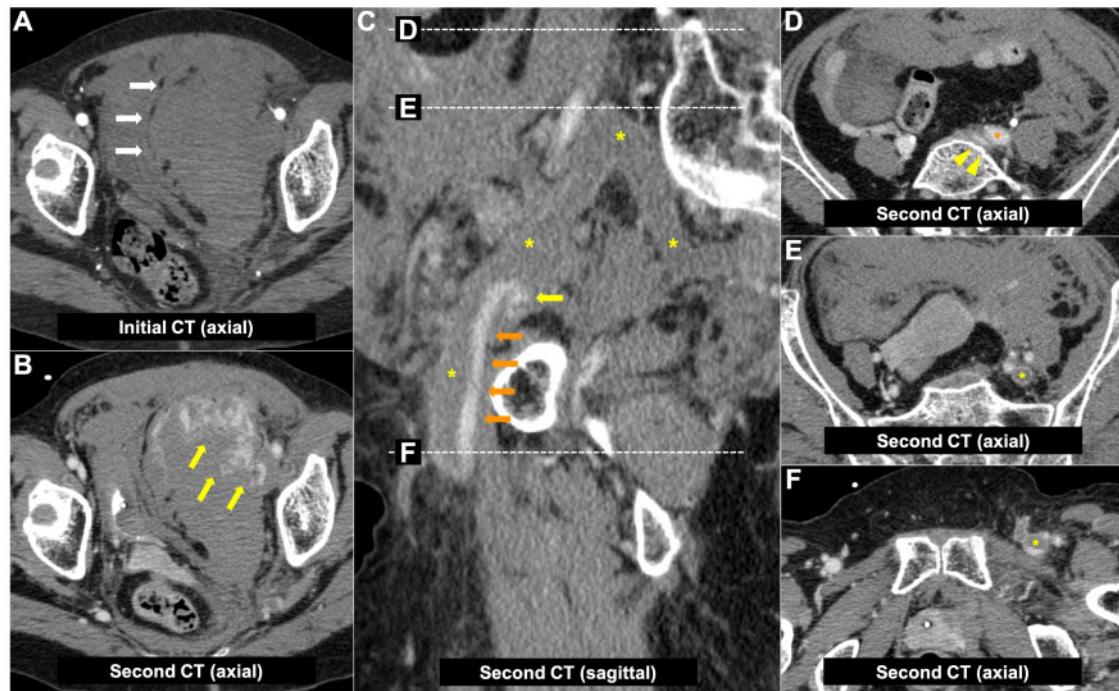


Figure 1 The initial and second contrast-enhanced computed tomography. (A) The initial arterial phase contrast-enhanced computed tomography (axial image) reveals a large retroperitoneal haematoma without arterial extravasation (white arrows). (B) The second venous phase contrast-enhanced computed tomography (axial image) reveals an increased retroperitoneal haematoma with venous extravasation (yellow arrows). (C) The second venous phase contrast-enhanced computed tomography (sagittal image) shows a deep vein thrombosis (yellow asterisks) below the common iliac vein, remaining venous flow (orange arrows) from the femoral vein to the suboccluded iliac vein, and venous extravasation (yellow arrow). (D–F) The second venous phase contrast-enhanced computed tomography (axial images) show a suspected compression of the common iliac vein between the left internal iliac artery (orange asterisk) and fifth lumbar vertebra (yellow arrowheads) and deep vein thrombosis (yellow asterisks) below the common iliac vein. CT, computed tomography.

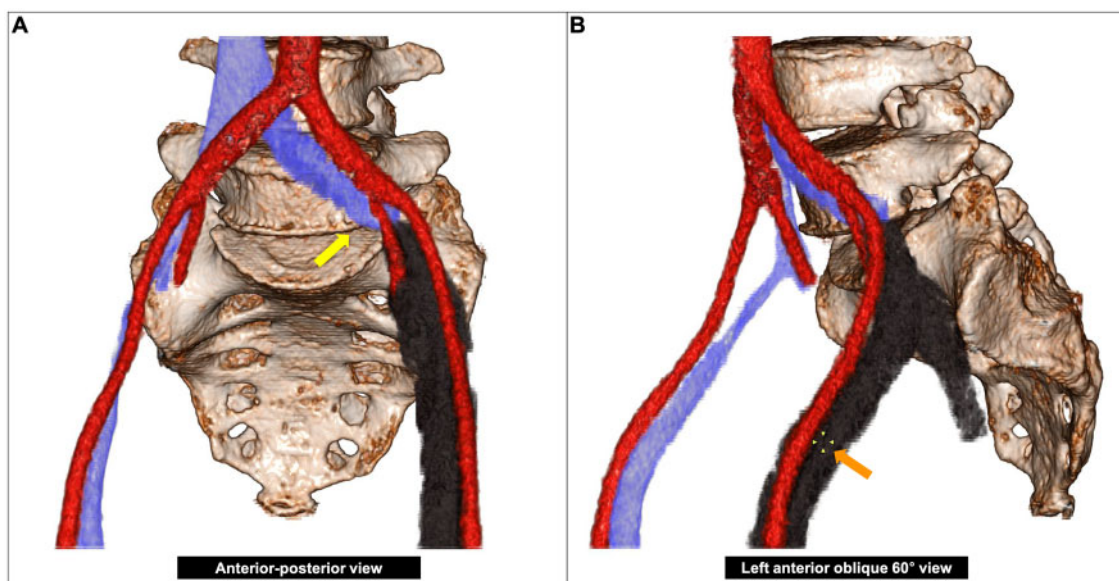


Figure 2 Pre-procedural venous phase contrast-enhanced computed tomography with 3D constructions. (A) Pre-procedural venous phase contrast-enhanced computed tomography with 3D constructions (Anterior-posterior view). The red indicates arteries, blue indicates veins, and black indicates deep vein thrombosis. The yellow arrow indicates the site of the suspected compression of the common iliac vein between the left internal iliac artery and fifth lumbar vertebra. (B) Pre-procedural venous phase contrast-enhanced computed tomography with 3D constructions (Left anterior oblique 60° view). The yellow arrowheads and orange arrow indicate the site of the rupture in the external iliac vein.

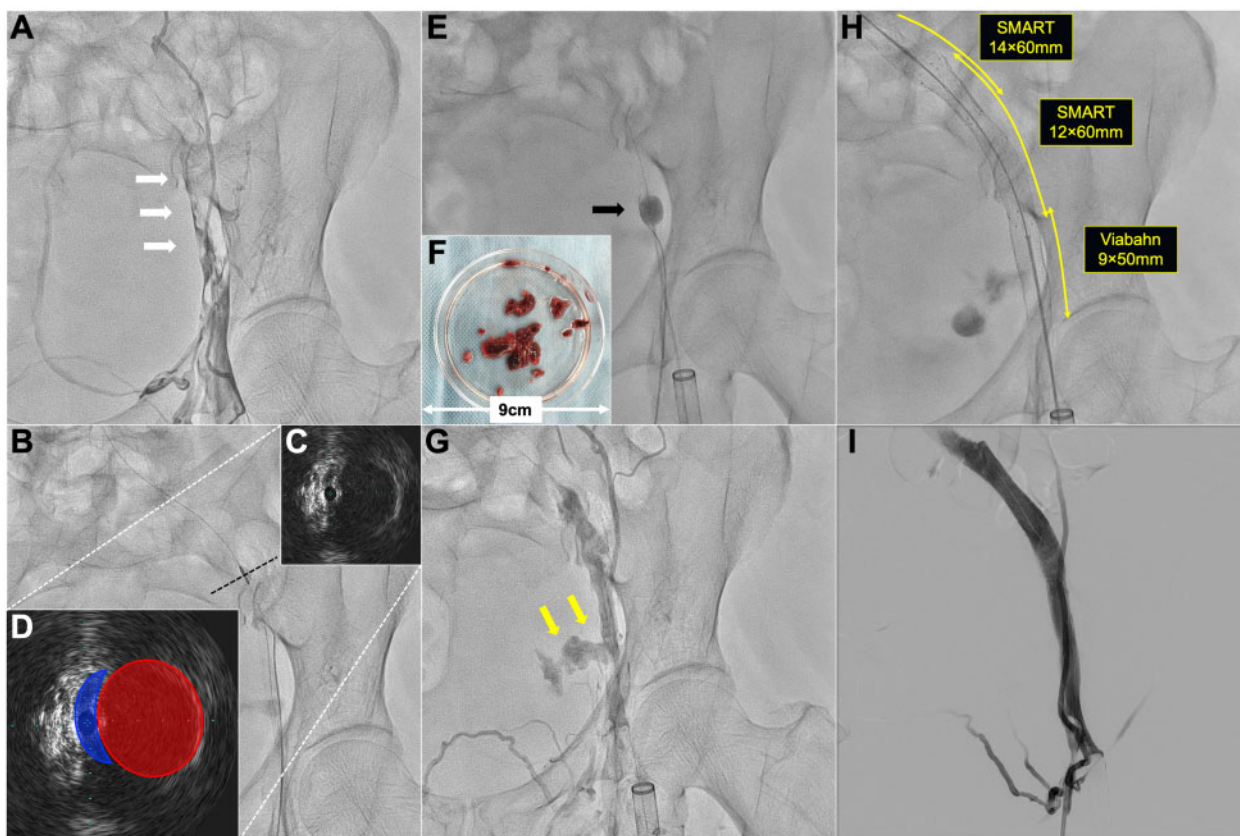


Figure 3 Elective endovascular procedures. (A) Venography via the left great saphenous vein reveals a lot of thrombi (white arrows) throughout the iliac vein. (B–D) Intravascular ultrasound confirms the compression of the common iliac vein (the black dotted line indicates the part observed by intravascular ultrasound, the red shadow indicates the iliac artery, and the blue shadow indicates the iliac vein). (E) A thrombectomy using a 4-Fr Fogarty balloon catheter (black arrow) with a softly inflated balloon is performed. (F) An insufficient amount of thrombi removed. (G) Angiography reveals active venous extravasation (yellow arrows). (H) A 9 mm × 50 mm covered stent is deployed for haemostasis in the external iliac vein, then 14 mm × 60 mm and 12 mm × 60 mm nitinol self-expanding stents with a strong radial force are deployed in the common iliac vein. (I) A final angiogram confirms a successful reperfusion.

× 50 mm covered stent (Viabahn; W. L. Gore, Flagstaff, AZ, USA) was deployed for haemostasis in the EIV. Then, 14 mm × 60 mm and 12 mm × 60 mm nitinol self-expanding stents with a strong radial force (SMART; Cordis, Fremont, CA, USA) were placed in the CIV (Figure 3H). A final angiogram confirmed a successful reperfusion (Figure 3I). Complete haemostasis was achieved by manual compression with a HemCon Patch (Tricol Biomedical, Portland, OR, USA) for 15 min after removing the sheath.

After the intervention, anticoagulation therapy was switched from unfractionated heparin to Rivaroxaban 15 mg twice daily for 3 weeks, followed by 15 mg once daily, which was the standard approved dose in Japan. The left lower limb swelling significantly improved, and the contrast-enhanced CT 6 days after the intervention confirmed fully dilated stents, a decreased retroperitoneal haematoma, and nearly dissolved DVT. The IVC filter was retrieved 7 days after the intervention. On a 3-month follow-up evaluation, the left lower limb swelling had completely disappeared (Figure 4B), and the contrast-enhanced

CT demonstrated the complete disappearance of the retroperitoneal haematoma and DVT (Figure 4A; Video 3).

Discussion

The three lessons learned from this case were as follows: (i) dynamic contrast-enhanced CT, including venous phase, should be considered in patients with a retroperitoneal haematoma of unknown cause; (ii) EVT may be minimally invasive and reasonable in the management of haemodynamically stable patients with a spontaneous iliac vein rupture; and (iii) May–Thurner syndrome may contribute to a spontaneous iliac vein rupture.

An accurate diagnosis during the initial evaluation remains challenging in patients with spontaneous iliac vein ruptures. Extensive testing is not always possible due to hypovolaemic shock, and the clinical picture mimics a ruptured AAA or abdominal catastrophe due to a gynaecological problem.⁴ Venous extravasation or DVT might be detected in the late arterial contrast-enhanced CT

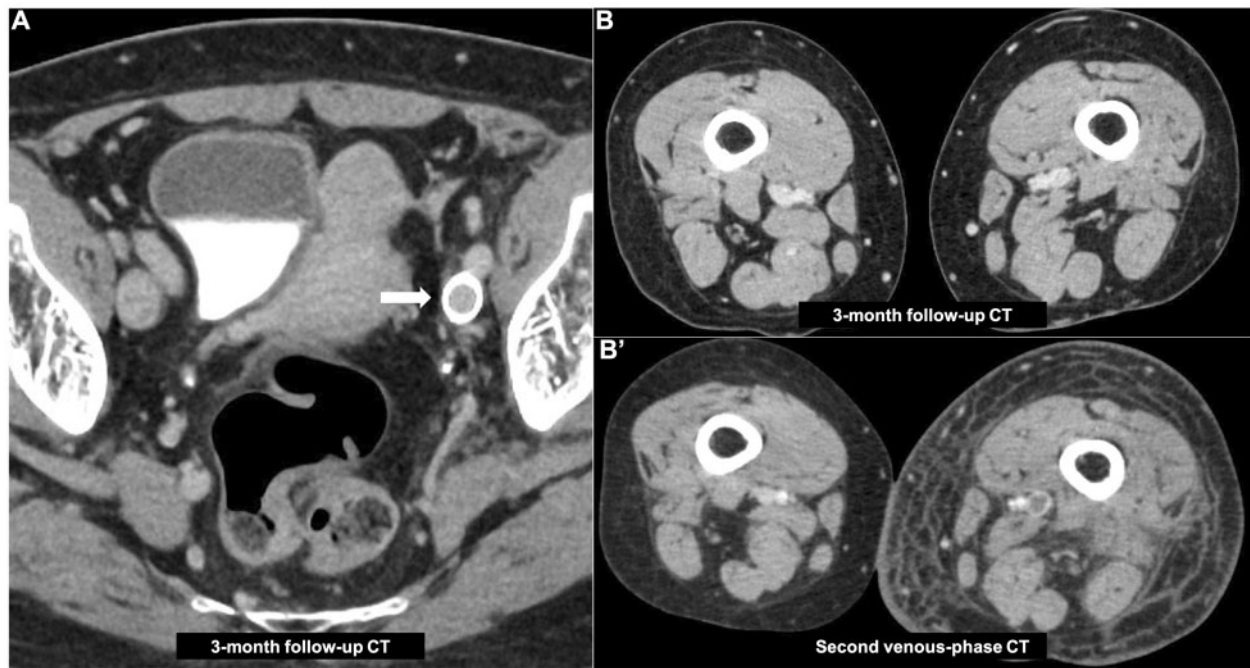


Figure 4 A 3-month follow-up evaluation. (A) The 3-month follow-up venous phase contrast-enhanced computed tomography demonstrates the complete disappearance of the retroperitoneal haematoma and deep vein thrombosis (white arrow). (B) The 3-month follow-up venous phase contrast-enhanced computed tomography (axial image) shows that the left lower limb swelling had completely improved compared to the swelling in the second venous phase contrast-enhanced computed tomography (axial image) on admission (B'). CT, computed tomography.

depending on the condition of the patient, and images of the venous phase are more desirable.⁴ Dynamic contrast-enhanced CT, including venous phase, could be appropriate for differentiating a ruptured AAA and accurately diagnosing a ruptured iliac vein. To the best of our knowledge, this is the first case in which venous phase contrast-enhanced CT demonstrated venous extravasation in a patient with a

spontaneous iliac vein rupture. Magnetic resonance imaging (MRI) with MR venography has a potential benefit not only for diagnosing iliac vein ruptures in haemodynamically stable patients, especially in differentiating a gynaecological disease but also for frequent follow-up imaging without contrast media or ionizing radiation exposure.⁵



Video 1 The initial arterial phase contrast-enhanced computed tomography.



Video 2 The second venous phase contrast-enhanced computed tomography.



Video 3 The 3-month follow-up contrast-enhanced computed tomography.

In the previous reports, most patients underwent open surgery without an accurate diagnosis of the spontaneous iliac vein rupture, leading to a high operative mortality and morbidity.^{2,3} Patients that are haemodynamically stable after fluid resuscitation and blood transfusions could allow for a temporal conservative management as an alternative option to open surgery.⁶ If patients have severe leg swelling due to a concomitant proximal DVT even after the introduction of anticoagulation therapy, elective EVT may prevent the development of post-thrombotic syndrome (PTS).⁷ Furthermore, a surgical repair of the iliac veins alone cannot relieve the compression of the iliac veins in patients with May–Thurner syndrome, suggesting EVT may be reasonable in the management of those patients. Several previous reports have shown that EVT, including the placement of covered stents, is safe and effective not only to stop bleeding and relieve occlusions in the acute phase but also to prevent the development of PTS in the chronic phase,^{1,4,8–10} and the clinical course of our case supported those previous reports.

The cause of a spontaneous iliac vein rupture remains unclear, although previous reports speculated the causes included a loss of an oestrogen effect,^{2,6} a sudden increased venous pressure due to activities such as bending or defecation,¹¹ and May–Thurner syndrome.^{2,3} Our case revealed that the DVT was not the result of the compression of the iliac vein by a retroperitoneal haematoma because the DVT was already present during the rupture of the iliac vein. The venous phase contrast-enhanced CT (Figures 1D and 2A) and IVUS (Figure 3B–D) confirmed that the left CIV was compressed between the left internal iliac artery and fifth lumbar vertebra (May–Thurner syndrome) and that the ruptured site of the left external iliac vein was downstream from the compressed site of the left CIV. The venous phase contrast-enhanced CT (Figure 1C) also suggested that a remaining venous

flow from the femoral vein to the sub-occluded iliac vein caused an increased venous pressure due to an upstream venous occlusion from the May–Thurner syndrome, and might have led to the downstream iliac vein rupture.

Herein, we report the case of a spontaneous iliac vein rupture associated with May–Thurner syndrome. Our case provided not only the potential value of the venous phase contrast-enhanced CT in the diagnosis of a spontaneous iliac vein rupture but also the potential benefit of conservative management followed by elective EVT.

Lead author biography



Yuji Nishimoto, MD is an interventional cardiologist and intensivist at Hyogo Prefectural Amagasaki General Medical Center in Japan. His areas of interest include venous thromboembolism, Cardio-Oncology, and cardiogenic shock.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: Y.N. received lecture fees from Daiichi-Sankyo, Bristol-Myers Squibb, Pfizer, and Bayer Healthcare. All other authors have reported that they have no relationships relevant to the contents of this article to disclose.

Funding: None declared.

Acknowledgements

We are grateful to Mr John Martin for his grammatical assistance.

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