CASE REPORT

Pulmonary arteriovenous fistula complicated by venous thromboembolism and paradoxical cerebral infarction during early pregnancy

Hiroshi Tajima, Hajime Kasai, Toshihiko Sugiura, Koichiro Tatsumi

Department of Respiriology,
Graduate School of Medicine,
Chiba University, Chiba, Japan

Correspondence to

Dr Hajime Kasai,
daikasai6075@yahoo.co.jp

Accepted 7 December 2017

SUMMARY

A 39-year-old Japanese woman was diagnosed with a pulmonary arteriovenous fistula (PAVF) complicated by paradoxical cerebral infarction during pregnancy. She received an inferior vena cava filter (IVCF), but it was difficult to indwell because of the risk of repeat thromboembolism via PAVF. Therefore, we temporarily occluded the pulmonary artery with a balloon to prevent free thrombi from reaching the IVCF. Detection and treatment of PAVF are recommended in fertile young women. Obstruction of the pulmonary artery trunk with a balloon catheter may be useful during IVCF removal in patients with untreated PAVF.

BACKGROUND

Pulmonary arteriovenous fistula (PAVF) is one symptom of hereditary haemorrhagic telangiectasia (HHT).¹ Since no oxygenated blood will pass through the fistula, PAVF results in progressive hypoxaemia, which leads to general malaise, dyspnoea and cyanosis.² PAVF shows lung field nodules on chest X-ray or CT. There are many differential diagnoses, such as tumour, infection and

pulmonary artery embolism, with this finding. PAVF is distinguished from these diseases by detecting a feeding artery and a draining vein.³ Additionally, PAVF can show central nervous system (CNS) symptoms, such as paradoxical cerebral infarction.⁴ In pregnant patients, it may cause symptomatic exacerbations of PAVF, such as increase in fistula diameter, rupture and cerebral infarction.⁵ A temporary inferior vena cava filter (IVCF) is clinically used to prevent pulmonary thromboembolism (PTE) when adequate anticoagulation treatment is difficult. However, long-term use of IVCF has been reported to induce various complications, such as new thrombus formation or perforation.⁶ Therefore, IVCF should be removed when initial indications no longer exist or when contraindications to anticoagulation have resolved. Here, we present a patient with PAVF who developed paradoxical cerebral infarction during pregnancy and had difficulty with anticoagulation therapy and withdrawal of IVCF.

CASE PRESENTATION

A 39-year-old woman was hospitalised due to loss of consciousness when she was 7 weeks pregnant. MRI revealed right middle cerebral artery infarction; she was advised for follow-up without anticoagulant therapy. Five days after hospitalisation, head CT showed haemorrhage around the infarction. Whole-body CT was performed due to an elevated D-dimer level (10.2 µg/mL, which exceeded the upper limit of the reference range (1.0 µg/mL)). The CT showed deep vein thrombosis, PTE of the right pulmonary artery and a PAVF in the left lower lung lobe. Due to haemorrhagic cerebral infarction, anticoagulant therapy was contraindicated. To prevent critical embolisation of the lung and brain, temporary IVCF (Neuhaus Protect; TORAY, Tokyo, Japan) was inserted.

INVESTIGATIONS

She was transported to our institution for management of the PAVF. At the time of admission, her vital signs were: blood pressure of 97/53 mm Hg, pulse rate of 63 beats/min, pulse oximetry of 98% on 2L/min oxygen inhalation and body temperature of 37.0°C. Her consciousness was clear. She had no angiotelectasias on her skin. Her neurological examination revealed incomplete motor paralysis of the left upper and lower extremities.

Figure 1 Chest X-ray on admission to our institution showing a pulmonary arteriovenous fistula (arrow) behind the cardiac shadow and the inserted temporary inferior vena cava filter (triangle).

To cite: Tajima H, Kasai H, Sugiura T, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-222519

Figure 2 (A) Enhanced CT showing a feeding artery and a draining vein. (B) Another level of the left lower lobe of the lung showing a nodular shadow connected with blood vessels in the left lower lobe of the lung. (C) Reconstructed three-dimensional CT showing that the pulmonary arteriovenous fistula had one feeding artery (1) and one draining vein (2).

She gave no remarkable medical history. However, she had been pregnant five times and had several episodes of nosebleeds during her previous pregnancy. Her family history was notable for a younger brother with PAVF. Chest X-ray and CT showed the presence of a nodular shadow in the left lower lobe of the lung (figure 1). Contrast-enhanced CT revealed the presence of blood vessels connected to this nodular shadow, which suggested the presence of a PAVF. Moreover, the contrast-enhanced CT revealed that the PAVF showed one feeding artery and one draining vein, measuring 4.4 and 5.4 mm in diameter, respectively (figure 2). Laboratory data revealed a D-dimer level of 18.7 µg/mL. Her activated partial thromboplastin time was 35.1 s, which was within the normal range of 25.0–36.0 s.

TREATMENT

We confirmed the diagnosis of PAVF with paradoxical cerebral infarction. We administered intravenous unfractionated heparin, and no subsequent cerebral haemorrhage was noted. Because the patient was observed to be in the acute phase of cerebral infarction, surgery or transcatheter embolisation was not indicated for the management of the PAVF. To prevent life-threatening complications, the patient and her husband decided to terminate the pregnancy. After the abortion, contrast-enhanced CT revealed a thrombus adherent to the IVCF; no other cerebral haemorrhages were noted. Therefore, sufficient anticoagulant

therapy was initiated and we planned withdrawal of the filter. However, as the IVCF itself could be a cause of thrombosis, we were concerned that another cerebral infarction might occur via PAVF during the IVCF removal. Thus, during the procedure, we obstructed the pulmonary artery trunk with a balloon catheter for approximately 20 s to prevent paradoxical cerebral infarction (figure 3).

OUTCOME AND FOLLOW-UP

The IVCF was successfully removed and no recurrence of cerebral infarction or haemorrhage was observed. After IVCF removal, anticoagulation therapy with edoxaban was initiated. We are planning on performing transcatheter embolisation of the PAVF after the patient has completed rehabilitation.

DISCUSSION

The present case demonstrates two notable clinical scenarios. First, detection and treatment of PAVF are recommended in young women presenting with a confirmed or a suspected family history of HHT. Second, to prevent paradoxical cerebral infarction during IVCF removal in a patient with PAVF, obstruction of the main trunk of the pulmonary artery using a balloon catheter may be useful.

HHT is an autosomal-dominant disorder characterised by recurrent nose bleeding, mucocutaneous telangiectasias and visceral involvement, including arteriovenous communications.⁷ HHT has previously been estimated to occur at a rate of 2.5 and 19.4 per 100 000, but may be underestimated.⁸ That is also the case in Japan.⁹ PAVF is a commonly associated symptom of HHT. The prevalence of PAVF in patients with HHT has been estimated to be 15% and 33% in previous studies.¹⁰ Patients with PAVF often develop CNS symptoms, such as headaches, disturbances of consciousness, convulsions, cerebral infarctions and brain abscesses. The incidence of these symptoms has been reported to be 19%–59%.¹¹ Cottin *et al* found that the most common CNS complications were brain abscesses and cerebral ischaemic events, with incidences of 47% and 40%, respectively.¹² These symptoms may be caused by erythrocytosis, paradoxical infarction and thrombi formation in patients with PAVF. Furthermore, during pregnancy, the mothers' enlarging uterus compresses the iliac vein and IVC and the risk of coagulation and thus thrombosis is further increased.¹³ On the other hand, it was reported that symptoms of PAVF may worsen during pregnancy.¹⁴ In an evaluation of 161 pregnancies in 47 patients with HHT, 23 pregnancies with PAVF were observed and 43% had PAVF exacerbations.⁵ The appearance of these symptoms was reportedly more frequent in the second and third trimesters of pregnancy, during which the circulating blood volume increases most significantly.¹⁴ Expansion and rupture of a pulmonary arteriovenous malformation (PAVM) was cited as a mechanism of exacerbation of PAVM during pregnancy, which was due to an increase in circulating blood volume/cardiac output accompanying pregnancy and the expansive action of progesterone on the PAVM. If vascular malformations of organs are not detected or are appropriately treated, the risk of serious complications during pregnancy can be decreased.¹⁵ Therefore, detection of PAVM is important, particularly in women who report a family history of HHT. Transthoracic contrast echocardiography (TTCE) and thoracic CT are recommended for the detection of a PAVM in patients presenting with HHT. The choice between CT and TTCE as screening tools for PAVM may depend on the preferences of physicians and patients, patients' age and gender and the available resources.¹⁶ In this patient, pregnancy

Figure 3 (A) Angiography of the IVC showing the thrombi in the IVCF. (B) Pulmonary angiography showing the expanded balloon catheter during IVCF removal. (Arrow: IVCF, triangle: thrombi, 1: expanded balloon catheter). IVC, inferior vena cava; IVCF, inferior vena cava filter.

itself increased the risk of thrombus formation, prompting the patient and her husband to opt for abortion. Haemorrhagic cerebral infarction developed early in pregnancy, resulting in clinical sequelae. In addition, the need to avoid X-ray exposure during pregnancy added to the difficulty of treatment. When the patient's cerebral infarction further worsened, her prognosis was deemed extremely poor; therefore, we considered reducing the risk of thrombus formation to be of primary importance.

Although temporary IVCF was proposed, thrombi that are trapped during indwelling may cause PTE at the time of removal. Millward suggested that if the size of the captured thrombus in the IVCF is up to 2×1 cm, there is a possibility of a small thromboembolism.¹⁷ In this case, thrombi were detected in the IVCF and it was suggested that free thrombi could cause pulmonary embolism and a recurrence of paradoxical cerebral embolism at the time of the IVCF removal. On the other hand, there was a risk of new thrombi formation and infarction if the IVCF was retained with insufficient anticoagulation therapy. Therefore, we temporarily occluded the main trunk of the left pulmonary artery with a balloon at the time of removal; this prevented free thrombi from causing paradoxical embolism via the PAVF.

In conclusion, we present a case of a pregnant patient with PAVF complicated by paradoxical cerebral infarction and insertion of an IVCF. Detection and treatment of PAVF are recommended in young women presenting with a confirmed or a suspected family history of HHT. During IVCF removal in patients with untreated PAVF, obstruction of the pulmonary artery trunk with a balloon catheter may be useful.

Learning points

- Pulmonary arteriovenous fistula (PAVF) may worsen by pregnancy and it makes the treatment difficult.
- Detection and treatment of PAVF are recommended for young women suspected or having a family history of hereditary haemorrhagic telangiectasia.
- During inferior vena cava filter removal in patients with untreated PAVF, obstruction of the pulmonary artery trunk with a balloon catheter may be useful.

Contributors HT is the guarantor of this manuscript and contributed to the design, data interpretation, writing and reviewing of the entire manuscript. HK contributed

to clinical analysis and reviewing of the entire manuscript. TS contributed to clinical care of the patient and data analysis. KT contributed to clinical care of the patient.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2018. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Gammon RB, Miksa AK, Keller FS. Osler-Weber-Rendu disease and pulmonary arteriovenous fistulas. Deterioration and embolotherapy during pregnancy. *Chest* 1990;98:1522–4.
- 2 Boczek ML. Pulmonary arteriovenous fistulas. *Mayo Clin Proc* 1999;74:1305.
- 3 Godwin JD, Webb WR. Dynamic computed tomography in the evaluation of vascular lung lesions. *Radiology* 1981;138:629–35.
- 4 Swanson KL, Prakash UB, Stanson AW. Pulmonary arteriovenous fistulas: mayo clinic experience, 1982–1997. *Mayo Clin Proc* 1999;74:671–80.
- 5 Shovlin CL, Winstock AR, Peters AM, et al. Medical complications of pregnancy in hereditary haemorrhagic telangiectasia. *QJM* 1995;88:879–87.
- 6 Van Ha TG. Complications of inferior vena caval filters. *Semin Intervent Radiol* 2006;23:150–5.
- 7 Shovlin CL, Guttacher AE, Buscarini E, et al. Diagnostic criteria for hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome). *Am J Med Genet* 2000;91:66–7.
- 8 Kjeldsen AD, Vase P, Green A, et al. [Hereditary hemorrhagic telangiectasia. A population-based study on prevalence and mortality among Danish HHT patients]. *Ugeskr Laeger* 2000;162:3597–601.
- 9 Dakeishi M, Shioya T, Wada Y, et al. Genetic epidemiology of hereditary hemorrhagic telangiectasia in a local community in the northern part of Japan. *Hum Mutat* 2002;19:140–8.
- 10 Shovlin CL, Letarte M. Hereditary haemorrhagic telangiectasia and pulmonary arteriovenous malformations: issues in clinical management and review of pathogenic mechanisms. *Thorax* 1999;54:714–29.
- 11 Khurshid I, Downie GH. Pulmonary arteriovenous malformation. *Postgrad Med J* 2002;78:191–7.
- 12 Cottin V, Chinnet T, Lavolé A, et al. Pulmonary arteriovenous malformations in hereditary hemorrhagic telangiectasia: a series of 126 patients. *Medicine* 2007;86:1–17.
- 13 Macklon NS, Greer IA. Venous thromboembolic disease in obstetrics and gynaecology: the Scottish experience. *Scott Med J* 1996;41:83–6.
- 14 Gershon AS, Faughnan ME, Chon KS, et al. Transcatheter embolotherapy of maternal pulmonary arteriovenous malformations during pregnancy. *Chest* 2001;119:470–7.
- 15 Shovlin CL, Sodhi V, McCarthy A, et al. Estimates of maternal risks of pregnancy for women with hereditary haemorrhagic telangiectasia (Osler-Weber-Rendu syndrome): suggested approach for obstetric services. *BJOG* 2008;115:1108–15.
- 16 Dupuis-Girod S, Cottin V, Shovlin CL. The lung in hereditary hemorrhagic telangiectasia. *Respiration* 2017;94:315–30.
- 17 Millward SF. Günther Tulip retrievable filter: why, when and how? *Can Assoc Radiol J* 2001;52:188–92.

Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow