Rare disease

An embolus in the right atrium caught in the Chiari network and resistant to thrombolysis

Motovska Zuzana,¹ Widimsky Petr,¹ Bilkova Dana,¹ Penicka Martin,¹ Linkova Hana,¹ Kautznerova Dana,² Kolesar Miroslav,³ Koldová Ludmila,³ Kvasnicka Jan⁴

¹Third Medical Faculty of Charles University, Prague, Czech Republic

²Institute of Clinical and Experimental Medicine, Prague, Czech Republic

³Cardiocentre University Hospital Vinohrady, Prague, Czech Republic

⁴Thrombosis Center of General University Hospital, Prague, Czech Republic

Correspondence to Motovska Zuzana, zuzana.motovska@icex.cz

Summary

This case report describes a patient with thromboemboli trapped in the Chiari network within the right side of the heart and resistant to thrombolysis. The right atrial masses were completely removed under cardiopulmonary bypass. Histological evaluation confirmed a mixed thromboemboli, with thrombus structures showing signs of organisation and surrounded by a fibrous capsule. The plasma level of the plasminogen activator inhibitor type-1 (PAI-1) was 50% higher than the normal upper limit. In this presented case, the Chiari network displayed a protective function but the expansion and organisation of the thromboembolus found there made it resistant to lytic treatment. Another important factor that could have influenced the resistance to thrombolysis was the high level of PAI-1.

BACKGROUND

The ECG presence of right heart thromboemboli during a pulmonary embolism is described in the literature as occurring in about 4% of patients, with a high related rate of massive pulmonary embolism and short-term mortality.^{1 2} Unfortunately, there is no consensus regarding the treatment strategy for such patients. However, favourable results have been reported with thrombolysis.³

Here, a well-documented case report is presented, which describes a patient with thromboemboli trapped in the Chiari network within the right side of the heart and resistant to thrombolysis.

CASE PRESENTATION

A 43-year-old woman presented in hospital with pain and asymmetric swelling of her left leg, which had been accompanied by shortness of breath during physical activity over the previous 2 weeks. At presentation, she was a smoker, had a long history of taking contraceptive medication and she was on substitution treatment for hypothyreosis. Her history also included two spontaneous abortions. The previous year, she had been admitted to hospital for pyelonephritis complicated by septic shock; during this hospital stay the patient underwent transthoracic ECG where a Chiari network in the right atrium was found. The patient's mother had suffered from a postoperative pulmonary embolism at the age of 50 years.

On physical examination, the patient was stable with a blood pressure of 140/80 mm Hg, heart rate 80 bpm; her height was 165 cm and weight was 68 kg. Except for the left leg oedema, with positive Homan's sign, there were no other pathological findings. No signs of right heart failure were present. A laboratory blood examination found d-dimers, mild leucocytosis and above normal levels of C reactive protein. A ventilation-perfusion lung scan

confirmed a bilateral pulmonary embolism. The source of the embolus was established by Doppler ultrasonography, which documented a femoral-popliteal venous thrombosis in the left leg.

The patient was haemodynamically stable, so treatment with a full-anticoagulation dose of unfractionated heparin (UFH) was started. A transthoracic ECG was performed, which showed two large stationary masses within the right atrium. Their structure and density were suggestive of emboli (figure 1, video 1). One mass was protruding from the right atrium through the tricuspid valve into the right ventricle, but without causing a gradient; systolic functions of both ventricles were normal and there were no signs of right heart overload. Transoesophageal ECG described the hypoechogenic masses as well-circumscribed (23 × 15 mm and 14 × 20 mm) with homogenous structure. The masses appeared to stem from the posterior wall of the right atrium and were caught in the Chiari network (video 2).

Video 1 Hypoechogenic mass protruding from the right atrium through the tricuspid valve into the right ventricle without a change in gradient. The systolic function of both ventricles was normal. There were no signs of right heart overload. 10.1136/bcr.09.2008.1019v1

Video 2 Transoesophageal ECG described the hypoechogenic masses as well-circumscribed with homogenous structure. The masses appeared to stem from the posterior wall of the right atrium and were caught in the Chiari network. 10.1136/bcr.09.2008.1019v2

Both structures were assessed as thromboemboli lodged in the right atrium. Thrombolytic treatment with an intravenous infusion of 100 mg recombinant tissue-type plasminogen activator (rt-PA), over a period of 2 h, with continuous ECG monitoring was performed. Following rt-PA treatment, full dose UFH was restarted. In subsequent

Figure 1 Transthoracic ECG showing two large stationary masses. Their structure and density were suggestive of emboli.

Figure 2 MRI confirmed a thromboembolic aetiology of the right atrial masses.

Figure 3 The right atrial masses were completely removed under cardiopulmonary bypass.

control ECGs, the two masses found to persist without any change in their shape or size. MRI further confirmed the thromboembolic aetiology of these right atrial masses (figure 2). With respect to the high risk of a fatal pulmonary embolism and the inefficacy of the most potent lytic strat-

egy, surgical embolectomy was indicated. Preoperatively, a control Doppler ultrasonography documented the persistence of thrombotic materials in the veins of the left leg. Therefore, a temporary inferior vena cava filter was implanted prior to surgery.

Figure 4 Histological evaluation confirmed a mixed thromboemboli, with thrombus structures showing signs of organisation and surrounded by a fibrous capsule.

The right atrial masses were completely removed under cardiopulmonary bypass (figure 3). Histological evaluation confirmed a mixed thromboemboli, with thrombus structures showing signs of organisation and surrounded by a fibrous capsule (figure 4).

The patient, with a positive family history of pulmonary embolism, had further known risk factors for venous thromboembolism—that is, cigarette smoking and use of oral contraceptive. Because of the high suspicion of thrombophilia, the patient was tested for thrombophilic disorders. A heterozygous methylenetetrahydrofolate reductase gene polymorphism was found and the plasma level of the plasminogen activator inhibitor type-1 (PAI-1) was 50% higher than the normal upper limit.

The patient was discharged home in good condition. She was placed on standard warfarin anticoagulation treatment. The patient also stopped cigarette smoking and discontinued with her contraception. One month after discharge, a control angiography of the vena cava showed persistence of thrombotic masses so the preoperatively implanted temporary vena cava filter was replaced by a permanent one.

DISCUSSION

Chiari anomalies in the human right atrium have been detected in 10.5% of autopsied hearts.⁴ The Chiari network is a web-like structure with a variable number of thread-like components. It is an embryological remnant, resulting from the incomplete resorption of the right sinus venosus valve and persistence of a filamentous tissue network.⁵ These filaments are attached to the wall of the right atrium in close proximity to the entrance of the inferior vena cava.⁴

Chiari malformation is not pathological and is rarely of clinical importance.^{5,6} The spider-web-like structure predisposes it to act as a filter for the inferior vena cava.⁷ However, the Chiari network can serve as a site of thrombus formation and also as a site where an embolus from the

inferior vena cava can become lodged. Endocarditis that develops solely within a Chiari network has been described as well.⁸

In this presented case, the Chiari network displayed a protective function but the expansion and organisation of the thromboembolus caught there made it resistant to lytic treatment. Another important factor that could have influenced the resistance to thrombolysis was the high level of PAI-1. PAI-1 is the primary physiological inhibitor of plasminogen activation in blood. When PAI-1 plasma levels are elevated, the action of tissue plasminogen activator is depressed⁹ resulting in thromboembolic disorders.¹⁰ In addition, elevated pretreatment levels of PAI-1 may reduce the efficacy of thrombolytic treatment by preventing or retarding clot dissolution.¹¹

The influence of impaired glucose metabolism or lipid disorders on PAI-1 plasma concentration was excluded in this patient. PAI-1 plasma levels partly depend on gene regulation.^{9,12} However, at present, genetic and environmental determinants of PAI-1 expression are incompletely understood.¹³ The human PAI-1 gene is located on chromosome 7 and contains nine exons and eight introns,¹² and several polymorphisms have been described within the gene. The patient's DNA was tested for a common single-base-pair polymorphism (4 or 5 guanine bases) in the promoter region of the gene (4G/5G), but the presence of this variant allele was not confirmed: the patient was homozygous for the 5G allele (5G/5G genotype).

Unsuccessful results with thrombolysis have been described in 8% of patients with an acute massive pulmonary embolism.¹⁴ In these patients, emergency surgical embolectomies have led to better in-hospital outcomes when compared to repeated thrombolysis attempts.¹⁴ In cases where a patient, after surgical embolectomy, is at increased risk for recurrent pulmonary embolism, the insertion of a vena cava filter is of potential benefit.¹⁵

Learning points

- ▶ The Chiari network may help prevent massive pulmonary embolisms through filtration of blood from the inferior vena cava.
- ▶ The Chiari network can serve as a site of thrombus formation and also as a site where an embolus from the inferior vena cava can become lodged.
- ▶ In the presented case, because of the combination of the thromboembolus trapped in the Chiari network and concomitant elevation of PAI-1, the thromboembolus had grown and organised and, thereby, became resistant to thrombolytic treatment.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Ferrari E**, Baudouy M, Cerboni P, *et al*. Clinical epidemiology of venous thromboembolic disease. Results of a French Multicentre Registry. *Eur Heart J* 1997;**18**:685–91.
2. **Torbicki A**, Gal N, Covezzoli A, *et al*. Right heart thrombi in pulmonary embolism: results from the International Cooperative Pulmonary Embolism Registry. *J Am Coll Cardiol* 2003;**41**:2245–51.
3. **Rose PS**, Punjabi NM, Pearse DB. Treatment of right heart thromboemboli. *Chest* 2002;**121**:806–14.
4. **Bhatnagar KP**, Nettleton GS, Campbell FR, *et al*. Chiari anomalies in the human right atrium. *Clin Anat* 2006;**19**:510–16.
5. **Schneider B**, Hofmann T, Justen MH, *et al*. Chiari's network: normal anatomic variant or risk factor for arterial embolic events? *J Am Coll Cardiol* 1995;**26**:203–10.
6. **Panidis IP**, Kotler MN, Mintz GS, *et al*. Clinical and echocardiographic features of right atrial masses. *Am Heart J* 1984;**107**:745–58.
7. **Goedde TA**, Conetta D, Rumisek JD. Chiari network entrapment of thromboemboli: congenital inferior vena cava filter. *Ann Thorac Surg* 1990;**49**:317–18.
8. **Payne DM**, Baskett RJ, Hirsch GM. Infectious endocarditis of a Chiari network. *Ann Thorac Surg* 2003;**76**:1303–5.
9. **Huber K**, Christ G, Wojta J, *et al*. Plasminogen activator inhibitor type-1 in cardiovascular disease. Status report 2001. *Thromb Res* 2001;**103** (Suppl 1):S7–19.
10. **Jorgensen M**, Bonnevie-Nielsen V. Increased concentration of the fast-acting plasminogen activator inhibitor in plasma associated with familial venous thrombosis. *Br J Haematol* 1987;**65**:175–80.
11. **Barbash GI**, Hod H, Roth A, *et al*. Correlation of baseline plasminogen activator inhibitor activity with patency of the infarct artery after thrombolytic therapy in acute myocardial infarction. *Am J Cardiol* 1989;**64**:1231–5.
12. **Kohler HP**, Grant PJ. Plasminogen-activator inhibitor type 1 and coronary artery disease. *N Engl J Med* 2000;**342**:1792–801.
13. **Ridker PM**, Hennekens CH, Lindpaintner K, *et al*. Arterial and venous thrombosis is not associated with the 4G/5G polymorphism in the promoter of the plasminogen activator inhibitor gene in a large cohort of US men. *Circulation* 1997;**95**:59–62.
14. **Meneveau N**, Sronde MF, Blonde MC, *et al*. Management of unsuccessful thrombolysis in acute massive pulmonary embolism. *Chest* 2006;**129**:1043–50.
15. **Streiff MB**. Vena caval filters: a comprehensive review. *Blood* 2000;**95**:3669–77.

This pdf has been created automatically from the final edited text and images.

Copyright 2010 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.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Zuzana M, Petr W, Dana B, Martin P, Hana L, Dana K, Miroslav K, Ludmila K, Jan K. An embolus in the right atrium caught in the Chiari network and resistant to thrombolysis. *BMJ Case Reports* 2010;10.1136/bcr.09.2008.1019, date of publication

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