Unusual association of diseases/symptoms

Chiari network: a protective filter against pulmonary embolism in a case of polycythaemia

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Summary

The Chiari network, a net-like structure within the right atrium, has been described as an embryonic remnant of no clinical significance. Its role in facilitating paradoxical arterial embolic events and as a potential site for thrombus formation however, has been reported in the literature. This case illustrates the protective filter-effect of the Chiari network in a 79-year-old Caucasian man with an unknown diagnosis of polycythaemia rubra vera who presented with clinical signs and symptoms of deep vein thrombosis. Transoesophageal echocardiogram confirmed entrapment of a thrombus within the Chiari network, which was not visualised following treatment on repeat scanning 1 month later. Here the authors describe how the Chiari network may have prevented the occurrence of a fatal massive pulmonary embolism in a patient with a high pro-thrombotic tendency, and review the current literature regarding the action of this anatomic variant as risk factor or protector.

BACKGROUND

The Chiari network is typically an incidental finding discovered during echocardiography or cardiac MRI. In the majority of cases it serves no clinical purpose, however, adverse associations have been reported including the development of arrhythmias, paradoxical arterial embolic events and infective endocarditis. The role of the Chiari network in thrombo-embolic disease is controversial – it may initially prevent the transmission of a thrombus by entrapment within the network, alternatively it could act as a nidus for thrombus formation, encouraging clot growth and development. We report a case where the Chiari network served as a protective intrinsic filter in the prevention of massive pulmonary embolism (PE).

CASE PRESENTATION

A 79-year-old Caucasian man, with a medical history of hiatus hernia and hypertension, presented with sudden onset pleuritic chest pain, associated with shortness of breath. He had attended his general practitioner due to pain and swelling of his right leg which had not been investigated prior to his admission. He was a non-smoker and had no known identifiable risk factors for thrombo-embolic disease.

On examination, he was haemodynamically stable with oxygen saturations of 91% on room air. There were no signs of right heart failure and examination of all other systems was unremarkable, apart from mild swelling and tenderness of the right lower leg. Twelve lead resting-electrocardiogram showed sinus rhythm with old left bundle branch block. Arterial blood gas confirmed marked type 1 respiratory failure with a PO_2 of 6.5kPa.

Subsequent blood investigations revealed a haemoglobin of 20.7 g/dl (13.5–17.0 g/dl) with a raised haematocrit of 0.62 (0.39–0.50); Troponin I was undetectable and D-dimer was elevated at 560 ng/ml (0–255 ng/ml). He was commenced on treatment with low molecular weight

heparin for probable right leg deep vein thrombosis (DVT) and subsequent PE.

An inferior vena cava (IVC) filter was inserted following a venous duplex ultrasound of the lower limbs which showed a large mobile thrombus in the left common and small femoral veins. There was no evidence of residual thrombus in the right leg. Transthoracic echocardiogram revealed normal left and right ventricular systolic function and an estimated pulmonary artery systolic pressure of 36 mm Hg. A long mobile structure was visualised within the right atrium, suggestive of part of Chiari network.

Further investigation with transoesophageal echocardiogram (TOE), confirmed a thrombus at the IVC/RA junction within the Chiari network (figure 1, video 1; early section). The patient was subsequently anticoagulated with warfarin. Lung ventilation-perfusion scan the following day revealed multiple small segmental pulmonary emboli.

Video 1 TOE video showing Chiari network. Initially with clot attached, second portion shows repeat TOE following treatment.

OUTCOME AND FOLLOW-UP

The patient was discharged when stabilised on warfarin therapy with an out-patient haematology follow-up for the management of his newly diagnosed polycythaemia rubra vera. There were no significant sequelae of PE or DVT on discharge. One month later, a repeat TOE confirmed a small Chiari network with no significant thrombus (figure 2, video 1; latter section) and the IVC filter was subsequently removed. The patient remained on life-long treatment with warfarin due to the high pro-thrombotic risk from his haematological disorder.

DISCUSSION

The Chiari network, an embryonic remnant from the persistence of the right valve of the sinus venosus, was first described in 1897.^{1 2} It is reported to occur in 2% of the

Figure 1 Transoesophageal echocardiogram (TOE) image on presentation. A large clot can be seen in the right atrium attached to a Chiari network. (A, large clot tethered to Chiari network; AV, aortic valve; LA: left atrium, RA: right atrium).

population, although more recently was observed in 13.6% of 213 cadaver hearts and 10% of the 38 hearts that were autopsied.³ No prevalence with age or gender has been reported.⁴ There has been much speculation surrounding the purpose and exact role of the Chiari network, with the consensus that it is of little pathological consequence.

The echocardiographic findings of a Chiari network include a highly mobile fenestrated structure attached to the right atrium, in close proximity with the junction of the IVC and superior vena cava. It has been noted to have the appearance of a ring in certain views on TOE and has been described as having a characteristic ‘whip-like’ motion.² It originates from the Eustachian valve, an embryological remnant of the IVC, which runs along the posterior wall of the right atrium and has the role of directing blood from the IVC toward the foramen ovale in fetal life. The distal attachments of the Chiari network within the right atrium are variable.⁵ It remains an important differential to be considered in the finding of a right atrial abnormality which should be distinguished from masses such as thrombus, flail tricuspid leaflet or vegetation.²

The Chiari network has previously been associated with cardiac anomalies including the persistence of a patent foramen ovale (PFO) into adulthood and the formation of an atrial septal aneurysm. In a study of over 1400 patients, 80% of the 29 patients who had a Chiari network confirmed by TOE had a PFO compared with 28% of the control patients. Similarly, one quarter had evidence of an atrial septal aneurysm compared with 1.6% in the control group. Its presence was recognised as a potential risk factor for paradoxical arterial embolic events since the network

may encourage blood flow away from the IVC and toward the inter-atrial septum causing right-to-left shunting.⁴

In addition to its role as a risk factor for arterial embolic events, it has been suggested that the Chiari network may act as a nidus for thrombus formation. By initially entrapping the fragments of thrombi from a DVT an even bigger clot may develop, which once the filter-like effect of the Chiari network is overcome, could result in a fatal massive PE.⁶ Furthermore, Motovska *et al*, reported a case of an embolus caught within the Chiari network, which became resistant to thrombolysis due to the high level of expansion and organisation of the thrombus within the network. A high plasma level of plasminogen activator inhibitor type 1 was also considered to have had an influence on the failure of thrombolytic therapy. This led to the need for emergency surgical treatment with embolectomy.⁷

The protective role conferred by Chiari network in the entrapment of thrombo-emboli and thus preventing the formation of PE has also been described in the literature.^{8,9} Van Opstal *et al* described a large mobile mass in the right atrium which was caught within the Chiari network, although the patient subsequently had a massive PE which was successfully thrombolysed.¹⁰ The action of the Chiari network as an intrinsic filter or risk factor for thrombus formation may therefore seem paradoxical.

Massive pulmonary embolus is a potentially fatal condition, which when left untreated is associated with a mortality of 52%.¹¹ Thrombosis remains the commonest cause of mortality in patients with polycythaemia rubra vera.¹² This case supports the action of this embryonic remnant as a protective filter, which may have prevented the

Figure 2 TOE 1 month later, the clot has disappeared and the underlying Chiari network can be seen clearly. (AV, aortic valve; B, Chiari network; LA, left atrium; RA, right atrium).

occurrence of a massive pulmonary embolus in a patient with a high pro-thrombotic tendency.

Learning points

- ▶ An underlying cause should be sought in patients presenting with idiopathic, suspected bilateral DVT.
- ▶ Despite its potential action as a nidus for clot formation, the Chiari network may also act as a protective filter by entrapment of thrombus.
- ▶ The finding of a Chiari network on echocardiography may be of particular clinical significance in patients with a pro-thrombotic tendency.

Competing interests None.

Patient consent Obtained.

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