Cor triatriatum: a rare cause of embolisation

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▸ Three video clips showing the presence of cor triatriatum are available at <www.hkmj.org>.

Transthoracic echocardiogram was arranged for a 42-year-old woman who had been diagnosed with ischaemic stroke in April 2013. While no thrombus was found, a membranous structure with two moderately sized fenestrations near the anterior and lateral border of the left atrium was noted by portable transthoracic echocardiogram (Fig a, Video 1). Flow accentuation with gradient up to 7 mm Hg across the fenestrations and spontaneous echo contrast were also documented by formal transthoracic echocardiogram (Fig b, Video 2) and transoesophageal echocardiogram (Fig c, Video 3). Computed tomography of the heart with contrast was arranged and the findings concurred with the echocardiogram findings (Fig d). In addition to cardiac structural abnormalities, the patient was noted to have atrial fibrillation. The overall picture was compatible with cor triatriatum, atrial fibrillation, and history of embolic stroke. She refused both anticoagulation and surgical excision of the cor triatriatum membrane at the time of diagnosis. However, 2 months later, the patient developed acute ischaemia of the right arm. Computed tomography revealed a 4-cm filling defect at the right proximal brachial artery. Echocardiogram did not reveal any intra-cardiac thrombus. Surgical embolectomy was successful in restoring the distal pulses. Excision of the cor triatriatum along with the modified Cox Maze III procedure and left atrial appendage plication were subsequently performed.

Cor triatriatum is an uncommon congenital anomaly. The left atrium is subdivided into a proximal and a distal chamber by a fenestrated fibromuscular membrane. The reported incidence is around 0.1% of all congenital cardiac diseases.[1] The most commonly associated structural abnormalities in adults include secundum atrial septal defect, mitral regurgitation,

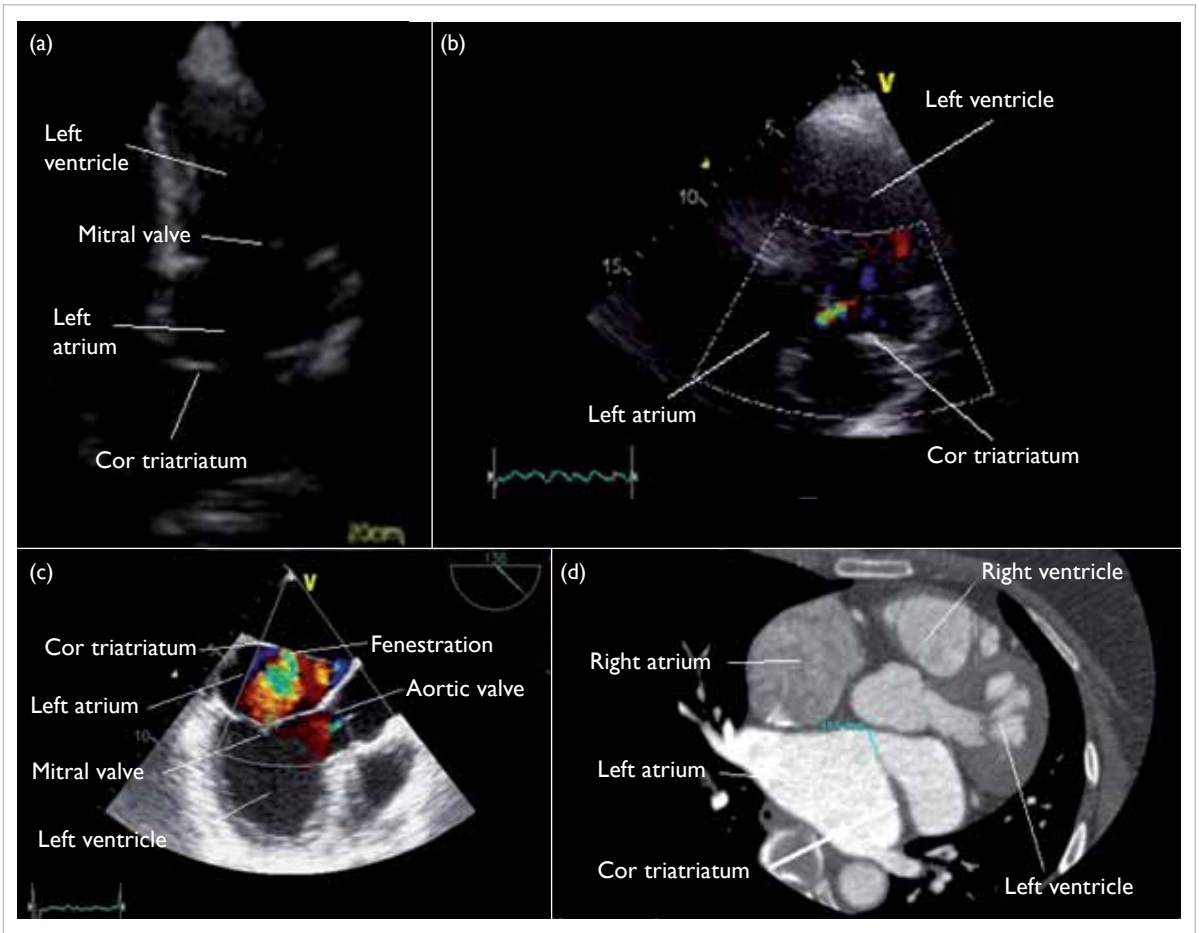

FIG. (a) Cor triatriatum shown by portable echocardiogram at apical four-chamber view. (b) Flow accentuation across cor triatriatum at a modified four-chamber view. (c) Flow accentuation across cor triatriatum by transoesophageal echo at mid-oesophageal long-axis view. (d) Cor triatriatum shown on computed tomography

and left superior vena cava with unroofed coronary sinus.[2] Patients present with symptoms comparable to mitral stenosis due to their similar haemodynamic effects with dyspnoea, orthopnoea, and haemoptysis. Cardioembolic stroke is another recognised complication and echocardiographic features of embolic stroke, including left atrial thrombus and spontaneous echo contrast, are often found.[3] Transthoracic echocardiogram is the initial investigation of choice due to its accessibility. Transoesophageal echocardiogram is needed to define the structure precisely and to screen for other co-existing congenital abnormalities. Computed tomography of the heart can supplement the echocardiographic findings before definitive treatment.[4] Open surgical resection of the accessory membrane is indicated in patients with obstructive symptoms. The procedure is performed through median sternotomy and atriotomy according to the morphology of the membranous defect and co-existing abnormalities. The operative result is excellent when patients present early and there are no co-existing cardiac anomalies.[5]

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VIDEO CLIPS at <www.hkmj.org>:
(1) The presence of cor triatriatum in the left atrium is shown by a portable echocardiogram machine at the apical four-chamber view. (2) Flow accentuation across the fenestration in the cor triatriatum is shown by a standard echocardiogram machine at a modified apical four-chamber view. (3) The presence of cor triatriatum is shown by transoesophageal echocardiogram at mid-oesophageal long-axis view.

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