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Cor Triatriatum and Recurrent Thromboembolic Stroke

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Ethics Approval: I can confirm that the information provided in the above article is approved by the ethical board of my institution and does not require patient consent.

ABSTRACT

A woman with recurrent thromboembolic stroke was found to also have cor triatriatum. When the patient first presented with weakness, she was thought to have ischaemic stroke because she had conventional risk factors, but she was later confirmed to have cor triatriatum. The main method of treatment is surgery. However, if surgery is contraindicated, anticoagulation can be used as second-line treatment, but this can be difficult. This report describes the follow-up of a middle-aged female patient with cor triatriatum over 6 years during which she experienced multiple strokes despite different methods of anticoagulation.

LEARNING POINTS

- Cor triatriatum is a rare heart condition which may not be detected by routine transthoracic echocardiography and so requires transthoracic echocardiography and CT angiography.
- Surgical membrane resection is the main treatment option but thromboembolic stroke should be considered when surgery is not possible.
- Anticoagulation may not be as effective at preventing embolic stroke in this rare heart defect as it is in other conditions.

KEYWORDS

Cor triatriatum, thromboembolic stroke, thromboembolic stroke

INTRODUCTION

Cor triatriatum is a rare congenital heart defect where the left atrium (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is subdivided by a thin membrane resulting in three atrial chambers [1]. This condition accounts for 0.1% of all congenital cardiac malformations and may be associated with other cardiac defects. It is primarily diagnosed through imaging such as echocardiography, CT and MRI.

CASE DESCRIPTION

A 48-year-old woman presented to the emergency department with sudden onset left extremity weakness in 2015. She had a number of comorbidities such as hypertension, type 2 diabetes mellitus on insulin, high BMI and mood problems, but she had good functional capacity, was living with her partner and two children and was independent in daily activities. Her family history was not significant for stroke or coronary heart disease and she had a brother who did not have any cardiac problems. Physical examination revealed she had left-sided 4/5 hemiparesis but was otherwise alert with normal heart sounds, a regular rhythm, and blood pressure within the normal range with no murmurs or thrills.

The stroke CT of the head showed low attenuation changes in the right corona radiata with minimal stenosis in carotid arteries. The patient was treated for ischaemic stroke with dual antiplatelet therapy for 3 weeks followed by life-long clopidogrel, and diabetes and hypertension control were optimized. She underwent stroke rehabilitation resulting in good functional recovery.

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Despite optimal HbA1C levels, blood pressure and lipid control, the patient again presented 7 months later with unsteadiness and vertigo. MRI of the head revealed multiple infarcts of different ages in different vascular territories (foci of restricted diffusion in subcortical white matter of the frontal lobes bilaterally, a mature infarct in the right cerebral hemisphere with T2 hyperdense signal change in the right thalamus, cerebral peduncle and pons). An intracranial CT angiogram showed atheromatous calcification of the cavernous segments of the internal carotid arteries bilaterally and the V4 segment of the left vertebral artery. Intracranial arteries were otherwise normal with no features of vasculitis.

A series of blood tests were carried out including lipid profile, vasculitis screen, thrombophilia screen (lupus anticoagulant antibodies, anticardiolipin antibodies protein C and S), myeloma screen, HIV, syphilis, Lyme antibodies and Fabry disease screen, which were all unremarkable.

The baseline ECG was in sinus rhythm with no episodes of arrhythmia shown on a 24-hour Holter monitor.

Two-dimensional transthoracic echocardiography (TTE) revealed a nearly obliterated left ventricular cavity with mild mitral regurgitation but normal LV function and a negative bubble study (*Fig. 1*).

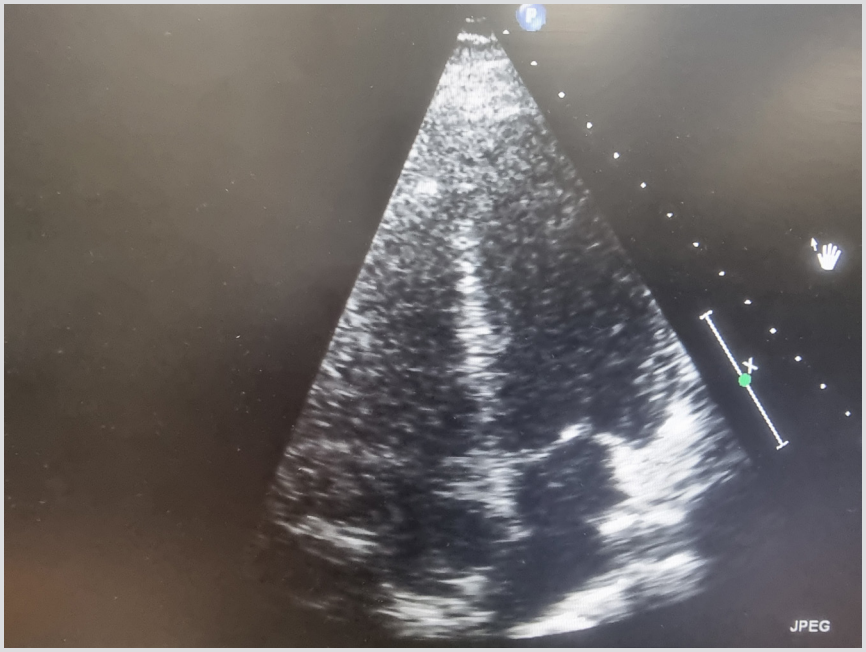

Figure 1. 2D Transthoracic echocardiogram revealed a nearly obliterated left ventricular cavity with mild mitral regurgitation

Transoesophageal echocardiography was attempted but failed because of the presence of oesophageal web. Therefore, CT coronary angiography (CTCA) was carried out and revealed findings in the left atrium consistent with a band extending from the intra-atrial septum to the anterior aspect of the left upper superior venous ostium (*Fig. 2*). These findings were suggestive of cor triatriatum[2]. There was no evidence of intra-cardiac thrombus. Doppler study showed flow through this membrane, and no significant jets or pressure gradient were demonstrated.

These findings were discussed with the congenital cardiology disease multidisciplinary team (MDT). Given the patient's multiple comorbidities, the absence of flow obstruction, the risk of atrial fibrillation with removal of the membrane which would not reduce the risk of stroke, and the presence of conventional risk factors for recurrent stroke, surgery was considered inappropriate at that time and it was decided to treat the patient medically with anticoagulation and annual surveillance of the left atrium for flow obstruction.

Following the MDT decision, the patient was initially started on warfarin, but because of menorrhagia and difficult INR control with poor compliance, warfarin anticoagulation was stopped and restarted many times. The situation was discussed with Haematology in order to consider alternatives such as direct oral anticoagulants (DOACs), but they are not licensed for use to prevent stroke in patients with cor triatriatum.

The patient experienced new right-sided weakness 2 years later in 2019 with MRI of the head findings consistent with acute infarcts in the left corona radiata with extensive high T2 signal abnormalities in cerebral white matter in keeping with severe small vessel disease. Again, no episodes of arrhythmia were demonstrated on Holter monitoring with CTCA findings again consistent with a non-obstructive membrane

in the upper left atrium with no thrombus. Given the poor compliance with warfarin, off-label apixaban 5 mg twice a day was started after discussion with Haematology.

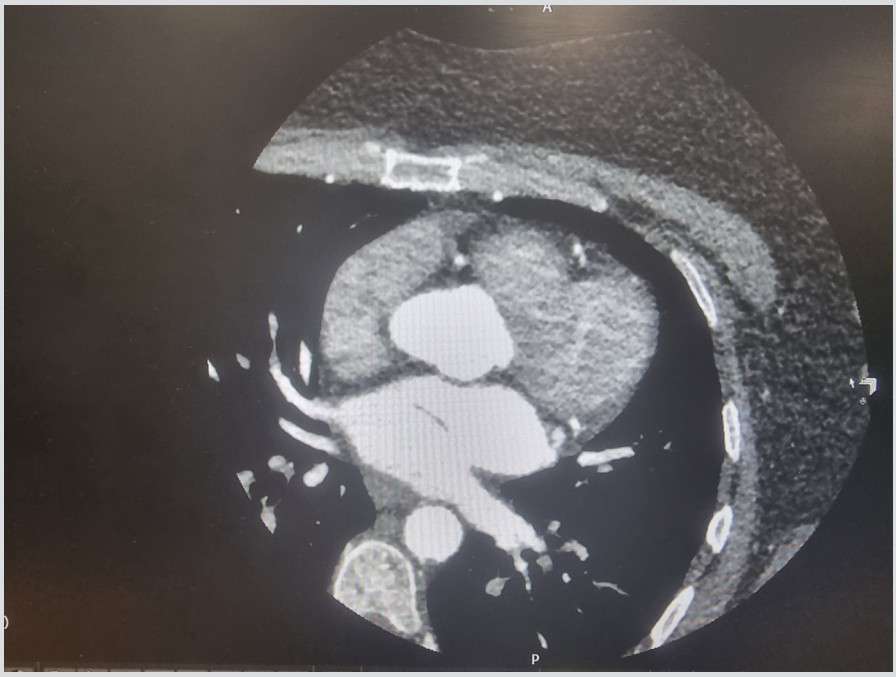

Figure 2. CT angiogram showing a non-obstructive membrane in the left atrium from the intra-atrial septum to the venous ostium with no thrombus

Even after changing to apixaban and with good compliance, the patient had two further episodes of ischaemic stroke in 2020 and 2021. MRI of the head in 2021 revealed established ischaemic damage in the corona radiata and thalami bilaterally and left cerebellum with an acute infarct in the right corona radiata and micro haemorrhages in the right cerebellum (*Fig. 3*). Following the 2021 stroke, anticoagulation treatment was changed to a therapeutic dose of subcutaneous enoxaparin injected daily.

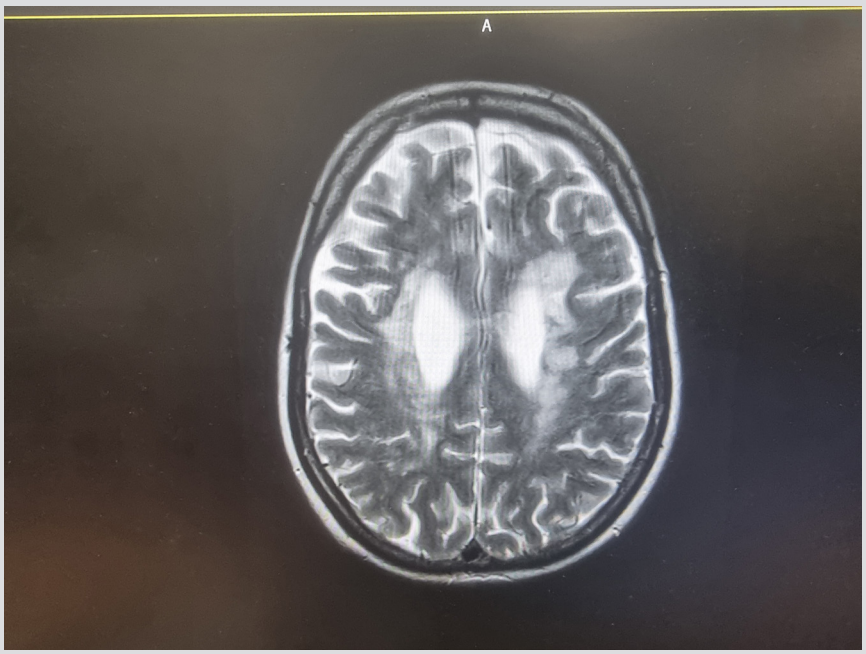

Figure 3. MRI of the head showing ischaemic damage of different ages in multiple vascular territories

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OUTCOME AND CONCLUSION

The long-term use of enoxaparin injections is not the most convenient anticoagulation for the patient but she has not had any further strokes since late 2021.

Regular surveillance of cor triatriatum, flow obstruction monitoring by the cardiology team and regular follow-up by the neurology team is maintained. Repeat MRI of the head in January 2022 in the neurology outpatient clinic showed no change in overall appearance. A repeat echo demonstrated normal left ventricular function with no flow obstruction.

The neurology team believes if the patient has further strokes in the future or if follow-up MRI shows ischaemia progression, that enoxaparin is probably not better than apixaban and further changes in anticoagulation treatment would be of limited benefit and potentially cause haemorrhagic complications requiring changing back to a DOAC for patient convenience.

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