A rare case of adult incomplete Shone complex

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Aims

To present a rare case of adult incomplete Shone complex with challenges in treatment and the role of multimodality imaging in its diagnosis.

Methods

Shone complex is a rare congenital heart disease that comprises multilevel obstruction of the left side of the heart and includes supramitral membrane, parachute mitral valve, subaortic membrane and coarctation of the aorta. However, the incomplete forms, characterised by two or three of the obstructive components, have been described rarely in adults.

A 50-year-old male patient reported shortness of breath with exertion for a long time; however, his symptoms had been more pronounced over the past 2 years and he was prescribed frusemide that seemed to help with his symptoms. He presented to us for follow-up; echocardiography was performed and showed a subaortic ridge with a left ventricular outflow tract (LVOT) gradient of 40 mmHg and a dysplastic mitral valve with moderate mitral regurgitation and transmitral mean gradient of 9 mmHg with no criteria of rheumatic affection and with a characteristic parachute-like appearance of the mitral valve. Initial assessment of the mitral valve area by traditional planimetry did not explain the high gradient and the flame-shaped turbulent jet at the mitral valve orifice; however, a three-dimensional assessment of the subvalvular structures suggested a possible secondary orifice with an approximate area of 11 mm.

Magnetic resonance imaging was done, which confirmed the diagnosis of a fibrous subaortic ridge, parachute-like mitral valve with severe mitral regurgitation, the anteromedial papillary muscle being underdeveloped and excluded coarctation of the aorta. A diagnosis of incomplete form of Shone complex was then made. The patient was advised regarding the need for surgery; however, he decided to continue on medical treatment as his symptoms were more or less controlled on frusemide and a beta blocker.

The patient had repeated follow-up echocardiography studies with diverse Doppler readings according to heart rate and volume status. Adjustment of diuretic doses was challenging, especially when the patient experienced dehydration due to fever or hot weather, or volume overload due to salty food.

Over 3 years' follow-up, the patient was admitted three times with increasing shortness of breath, melaena and iron-deficiency anaemia. Upper gastrointestinal endoscopy and colonoscopy were normal. Capsule endoscopy was done for suspected associated vascular malformations (angiodysplasia) secondary to possible acquired von Willebrand factor (vWF) deficiency (Heyde syndrome); however, all the results were normal.

Results

Our patient is still well with frequent cardiology clinic follow up.

Conclusions

Our case is one of the rarely reported adult patients with incomplete Shone complex. The presence of a left-sided obstructive lesion should arouse the suspicion of other obstructive lesions at different levels. The feasibility of multimodality imaging will probably allow better assessment of different lesions and probably result in more cases of the adult incomplete variant being reported. ■

Conflict of interest statement

None declared.

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