Incomplete Shone’s complex: adult age diagnosis

Ricardo C. Rodrigues, André Correia, Gomes Serrão, Paula Faria, Susana Gomes & Décio Pereira

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A 25-year-old male with previous history of heart surgery was referred for a control echocardiogram. He had been operated when he was 5 years old for repairation of aortic coarctation and the excision of a subaortic membrane, and was then lost to follow-up. No other changes were detected previously or during surgery. The patient was clinically stable without medication and the physical exam was unremarkable. The echocardiogram showed normal left ventricular function, but bicuspid aortic valve (figure 1 A), conditioning mild aortic stenosis, and a parachute mitral valve (figure 1 B, C) with single papillary muscle (figure 1 D, E – arrow) were present, with slight increase in transmural velocity and mild regurgitation. No residual coarctation was present.

Shone’s complex is a rare congenital heart disease consisting of several levels of left-sided obstructive lesions including supravalvar mitral ring, parachute mitral valve, subaortic stenosis and coarctation of aorta, being classified as complete (if all levels are present) or incomplete (if only 2 or 3 lesions are present). Our patient had a previous surgical intervention and no correction was made for two undiagnosed lesions. Furthermore, the main critical problem associated with this condition appears to be mitral valve obstruction which was not significant in our patient. A conservative approach was decided and at 3-year follow-up no events occurred.

This case highlights the importance of exhaustive preoperative echocardiographic evaluation and reminds us that, in the presence of two levels of left-side cavities obstruction, other possible related anatomical lesions must be excluded.

CONFLICT OF INTEREST: none.

Keywords Shone’s complex – congenital heart disease.
Fig. 1