A rare congenital valve abnormality unexpectedly detected in a patient with aortic dissection

Authors:
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Clinical Presentation A 50-yo male with a long standing history of arterial hypertension was admitted with sudden onset epigastric pain radiating to the back, associated with distal paresthesia. An urgent CT angiography showed a type B aortic dissection extending from descending aorta to celiac tripod. As blood pressure control was effectively obtained, with symptoms’ regression and no signs of peripheral ischemia, the patient was admitted to the ITU. Diagnostic Technique and their most important findings On transthoracic echocardiography, the function of both ventricles was preserved and a dilated aortic root with trivial regurgitation was noted. The suspicion of an aneurysm of the coronary aortic cusp or of the interventricular septum was also raised. To better understand aortic root anatomy, a 1.5 T CMR was performed showing tricuspid aortic valve with normal cusps and confirming the presence of an interventricular membranous septal (IVMS) aneurysm (16x17 mm) (Fig.1A, asterisk). The right heart was of normal size and function, while the left ventricle was moderately dilated with normal function. On tissue characterization, no myocardial edema, fat infiltration or fibrosis were noted. Surprisingly, a Double-Orifice Mitral Valve (DOMV) characterized by two separated and asymmetric annuli was noted (Fig.1B and C, arrows), with no evidence of significant regurgitation. A 3D transthoracic and transesophageal echocardiography was then performed for functional assessment of the valvular abnormality. No shunting was noted through the IVMS aneurysm (Fig.1D and E, asterisk). DOMV was confirmed (Fig.1E and F, arrows) with evidence of a medial orifice with 2 commissures and a lateral one with 4 commissures (Fig.1G, dashed arrows). Both orifices presented trivial regurgitation with no significant stenosis (mean trans-valvular gradient 1 mmHg). The patient remained asymptomatic with no evidence of progressive aortic false lumen dilatation, so he was discharged with a planned close ambulatory follow-up. Learning Points from this case DOMV is an extremely rare congenital anomaly, usually associated with other cardiac abnormalities, and generally detected in early childhood. Its prevalence in the adult population is unknown and its diagnosis prone to be missed, as it is frequently an isolated finding, with no clinical relevance, incidentally detected during echocardiography or CMR performed for other clinical indications.
Abstract: A rare congenital valve abnormality unexpectedly detected in a patient with aortic dissection.

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