

Arteria lusoria and hypertrophic obstructive cardiomyopathy. A unique coexistence

Tętnica błędząca i kardiomiopatia przerostowa zawężająca. Rzadkie współistnienie

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Coexistence of hypertrophic obstructive cardiomyopathy (HOCM) and aberrant subclavian artery (arteria lusoria) presents a unique combination. We describe two female patients in whom both anomalies were confirmed by cardiac magnetic resonance (CMR). Patient 1: A 57-year-old patient presented with New York Heart Association (NYHA) class III heart failure (HF). Transthoracic echocardiography revealed systolic anterior motion of the mitral leaflet and maximal left ventricular outflow tract (LVOT) gradient of 110 mmHg. CMR showed asymmetric hypertrophy up to 25 mm in the anteroseptal basal segments. Additionally, right arteria lusoria was diagnosed (Fig. 1). The patient underwent alcohol ablation of the septal branch. However, a significant gradient at the level of both papillary muscles and LVOT remained at 7 months. Thus, extended surgical myectomy was carried out with a satisfactory result. Patient 2: A 51-year-old patient presented with NYHA class III HF. Medical history included diabetes mellitus, arterial hypertension, and paroxysmal atrial fibrillation. Echocardiography showed maximal LVOT gradient of 90 mm Hg, pulmonary hypertension of 120 mm Hg (irreversible in the right heart catheterisation), and significant mitral regurgitation. CMR revealed asymmetric left ventricular hypertrophy in the anteroseptal basal segments. Again, right arteria lusoria was diagnosed. Conservative approach was the treatment strategy. Arteria lusoria is a frequent finding, occurring in 1% of the population. The prevalence of hypertrophic cardiomyopathy is estimated to be 0.2%, and LVOT obstruction is observed in the minority of these patients. Nonetheless, according to our knowledge no coexistence of both anomalies has been reported so far. Different molecular genetics do not allow us to suggest common pathogenesis of these disorders. Arteria lusoria used to be associated with different heart and vessel anomalies. In an impressive study of approximately 11,000 pathological specimens, arteria lusoria was identified in 128 patients, among whom 117 patients had a congenital heart disease [Zapata H et al. *Pediatric Cardiology*, 1993; 14: 159–161]. The most frequently diagnosed conditions were truncus arteriosus and left heart obstruction. In none of them HOCM was identified. Similar results were observed in a database of nearly 16,000 paediatric echocardiography exams [Ramaswamy P et al. *Am J Cardiol*, 2008; 101: 677–682]. Arteria lusoria was found in 226 patients in whom intracardiac lesions were common. Most of these defects were conotruncal anomalies. HOCM was not identified in this study. Nevertheless, the limitations of these two large studies must be taken into account. Many of the conditions found in this pathological specimens are lethal, and HOCM is not strictly a lethal disorder. Both of these studies included large, predominantly paediatric populations. For HOCM identification, however, older age is optimal. From a practical point of view, the presence of arteria lusoria may cause heart cannulation via right radial artery challenging. Wire entry into the ascending aorta may be difficult. Its identification was instrumental to avoid coronary cannulation via the right radial artery in the first patient. Arteria lusoria may be associated not only with conotruncal anomalies, but also with LVOT obstruction.

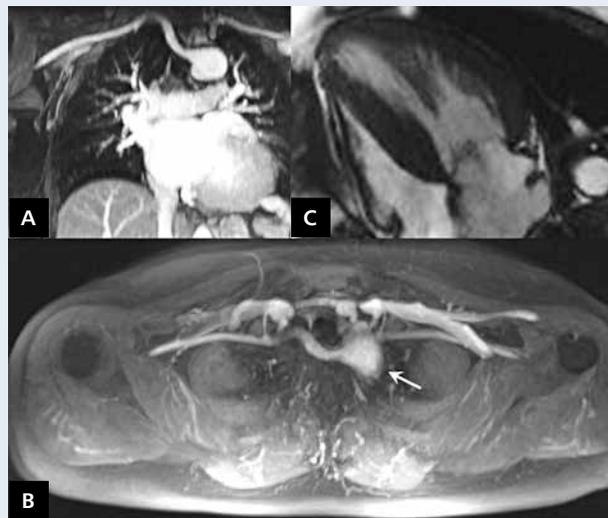


Figure 1. Cardiac magnetic resonance; **A, B.** Arteria lusoria in coronal plane and in axial plane, respectively; arrow shows the cross-section of the distal aortic arch; **C.** Hypertrophied interventricular septum

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