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An unexpected direction: a case report of a double aortic arch in an asymptomatic woman in adulthood

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Introduction: Complete vascular rings represent about 0.5-2% of all congenital cardiovascular malformations, with the double aortic arch (DAA) being the most common of the complete vascular rings, causing tracheoesophageal compression. The right (posterior) arch is usually dominant (70%), although the two arches can have the same size (5%). The left (anterior) arch is dominant in only approximately 25% of cases. In most cases, this anomaly is diagnosed during childhood due to symptoms caused by oesophageal or tracheal compression. For this reason, case reports of adults are rare. This report describes a case of a 61-year-old woman with DAA with dominant left arch, diagnosed accidentally by thoracic CT angiography.

Case Report Description: A 61 years old woman with a previous story of hypertension and type 1 diabetes presented to the emergency service with dyspnoea and thoracic pain. She also referred a history of intermittent dysphagia and cough with at least 12 years of progression. All the parameters of the physical examination were within normal limits. The electrocardiogram showed a normal sinus rhythm with no evidence of acute ischemia and her blood analyses did not show any abnormalitie. She also performed a thoracic CT angiography, which excluded signs of pulmonary embolism, but revealed a vascular ring suggesting a double aortic arch with permeability in both right and left arches as well as their collaterals. The Cardiac MRI was performed with the purpose of excluding ischemia, confirming the double aortic arch with left dominance. The right arch, posterior to the oesophagus and trachea, and the left arch, in an anterior position, showed an anatomic compression of the oesophagus as well as the proximal trachea, capable of eliciting the symptoms mentioned. Other congenital anomalies were excluded. The echocardiography did not demonstrate any additional cardiac malformation. Endoscopy shows a pulsatile extrinsic compression of the esophagus (aortic ring). The patient is currently being studied and closely monitored in the Cardiology consultation.

Discussion: The most common type of complete vascular ring is the double aortic arch, which accounts for 70% of the complete rings. In most cases, there are two permeable arches, usually with right dominance (70% of the cases). Rarely, both arches are symmetrical. Symptoms usually appear in the fifth month of life. In most cases, only supportive treatment is required.

Conclusion: This case illustrates the atypical features of this congenital malformation, namely the diagnosis during adulthood as well as the left dominance.

Abstract P725 Figure. A double aortic arch

