A right-sided aortic arch and aberrant left subclavian artery with proximal segment hypoplasia

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Abstract

The right-sided thoracic aortic arch is a rare congenital malformation occurring during embryologic development. A majority of the cases present with two anatomic variations, one of which is an aberrant left subclavian artery (LSA) originating off the distal descending aortic arch. This aberrant LSA courses across and compresses thoracic structures on its way to the left thoracic outlet. While this aberrant vessel causes mostly benign symptoms, patients may first present with rupture of a thoracic aneurysm. This thoracic aneurysm is commonly known as Kommerell’s diverticulum. This paper presents a case of an aberrant LSA originating off Kommerell’s diverticulum with a proximal long-segment hypoplasia, a very rare anatomic variation.

Keywords: Right-sided arch • Aberrant subclavian artery • Kommerell’s diverticulum • Isusoria

INTRODUCTION

A right-sided aortic arch and thoracic aorta is a rare congenital malformation occurring during the 4th and 5th weeks of embryogenesis. This defect has been reported to occur in 0.05–0.1% of the general population [1], and approximately half of these cases are associated with an aberrant left subclavian artery (LSA). To date, approximately 50 cases of right-sided aorta and aberrant LSA have been published in the literature [2]. These patients usually remain asymptomatic and the anomalies are found as incidental findings on imaging for other reasons. However, when present, symptoms develop secondary to the aberrant subclavian artery compressing adjacent structures. These same structures may also be compressed from aneurysm of the aortic wall, commonly known as Kommerell’s diverticulum. This paper will present a case of a symptomatic right-sided aortic arch, right-sided thoracic aorta and a left aberrant and hypoplastic subclavian artery originating from Kommerell’s diverticulum. This anatomy of the aberrant subclavian artery has not been previously described.

MATERIAL AND METHODS

A review of the Medline database for similar cases with the search queries ‘right-sided arch’, ‘right-sided aorta’, ‘aberrant LSA’, ‘Isusoria’, ‘dysphagia’ and ‘hypoplastic subclavian artery’ was performed. References for these reports were also reviewed for similar cases to the one presented.

CASE REPORT

A 39-year-old woman presented with chief complaint of recent changes in her voice. She also complained of left temporal migraine headaches, occurring approximately twice per week and having difficulty swallowing vitamins and pills since childhood. She denied any symptoms of claudication in her left arm both at rest and during exercise. She stated that since childhood her voice has ‘always been raspy’ but getting increasingly horse over the past year. Relevant past medical history included symptoms of heartburn and dysphagia starting in childhood.

Physical examination revealed right arm blood pressure of 137/98 mm Hg and left arm blood pressure of 129/89 mm Hg along with a pulse rate of 76 with a strong right radial pulse and barely palpable left radial pulse. Neck and chest revealed no signs of Jugular venous distension (JVD) and normal heart and lung sounds. The left arm did not show any atrophic changes.

A computer tomography scan with contrast revealed a right-sided aortic arch and aneurysm at the origin of an aberrant LSA with a proximal long-segment hypoplasia and post-aneurysm ectasia (Fig. 1). The oesophagus is compressed by Kommerell’s diverticulum, and the hypoplastic LSA travels behind the esophagus on its way to the left thoracic outlet.

DISCUSSION

This congenital anomaly occurs during the 4th and 5th weeks of embryogenesis when the six pairs of aortic arch vessels
connecting the ventral aorta to both dorsal aortae begin to migrate and form their respective structures. With normal embryologic development, the left 4th aortic arch becomes a continuation of the ascending aorta and develops into the normal left descending aortic arch. The development of a right-sided arch occurs when the right 4th arch becomes the right aortic arch, and the right dorsal aorta persists as the thoracic aorta with regression of the left dorsal aorta. The arch can be in the normal pre-tracheal configuration or a retrooesophageal configuration giving rise to various forms of vascular ring malformations. When an aberrant LSA is present, the vessel runoff from the aortic arch from proximal to distal occurs as left carotid, right carotid, right subclavian and lastly the LSA which courses from the right thorax to the left. The course of the LSA can vary as posterior to the oesophagus, anterior to the oesophagus or anterior to both the trachea and oesophagus [1].

The anatomy seen in our patient is an interesting embryologic variant that can help further our understanding of the genetic and embryologic development of the thoracic vasculature. Some authors believe that the weakness causing the diverticulum is a consequence of being derived from embryologic cells that would normally regress during development. However, mechanical forces experienced at the LSA bifurcation including increased fluid turbulence and tensional forces from left arm motion may also contribute to the progression of this relatively benign condition.

Many minimally invasive surgical options were discussed with the patient; however, she remained hesitant for any surgical intervention at this time and may opt for long-term surveillance.

Conflict of interest: none declared.

REFERENCES