MDCT angiography of isolated right subclavian artery

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SUMMARY: Türkvatan A, Paç A, Kızıltepe U, Ölçer T, Cumhur T. MDCT angiography of isolated right subclavian artery. Turk J Pediatr 2010; 52: 668-672.

Isolation of a subclavian artery is an uncommon congenital anomaly of the aortic arch in which one subclavian artery loses its connection with the aorta and originates from the homolateral pulmonary artery by way of a ductus arteriosus. Isolation of the left subclavian artery in patients with a right aortic arch is well known. However, isolated right subclavian artery with a left-sided aortic arch is an extremely rare condition. In this report, we present multidetector computed tomographic (MDCT) angiography findings of an isolated right subclavian artery associated with a common carotid trunk and an anomalous origin and proximal interruption of the left pulmonary artery.

Key words: aortic arch anomalies, isolated right subclavian artery, tetralogy of Fallot, multidetector computed tomography, angiography.

Isolation of a subclavian artery is an uncommon congenital anomaly of the aortic arch in which one subclavian artery loses its connection with the aorta and originates from the homolateral pulmonary artery by way of a ductus arteriosus[1]. This subclavian isolation is always observed on the opposite side of the position of the aortic arch. Isolation of the left subclavian artery in patients with a right aortic arch is well known. However, isolated right subclavian artery with a left-sided aortic arch is an extremely rare condition. According to Edwards' hypothetical double aortic arch system, isolated right subclavian artery with left aortic arch would be explained by interruption of the right aortic arch at two locations: between the right common carotid and the right subclavian arteries and between the right ductus arteriosus and the right dorsal aortic root[1]. The right subclavian artery may remain connected to the right pulmonary artery via either a ligamentum arteriosum or patent ductus arteriosus. Although conventional catheter angiography has been considered the definitive diagnostic tool, multidetector computed tomographic (MDCT) angiography has increasingly been used for the evaluation of congenital cardiovascular diseases[2,3].

In this report, we present MDCT angiography findings of an isolated right subclavian artery associated with a common carotid trunk and an anomalous origin and proximal interruption of the left pulmonary artery in a three-year-old boy with tetralogy of Fallot. To the best of our knowledge, this is the first report of MDCT angiography findings for an isolated right subclavian artery.

Case Report

A three-year-old boy was admitted due to fatigue and dyspnea during physical exertion. Physical examination revealed a heart rate of 100 beats/min and blood pressure of 79/54 mmHg in the right arm and 81/47 mmHg in the left arm. Both radial pulses were described as identical, and no clinical evidence of reduced blood flow to the right arm was seen. He was mildly cyanotic, and the oxygen saturation was 88% in room air. There were normal first and second heart sounds and 3/6 systolic murmur at the mid-sternal border. Laboratory examination revealed an iron-deficiency anemia. Chest roentgenogram showed left-sided aortic arch, cardiomegaly and volume loss, and decreased pulmonary markings in the left lung. Electrocardiography revealed right

ventricular hypertrophy, and echocardiography showed a large malaligned ventricular septal defect, an overriding aorta, an infundibular and valvular pulmonic stenosis (pressure gradient 52 mmHg), and right pulmonary artery stenosis. The left pulmonary artery and origin of the right subclavian artery could not be visualized by echocardiography. Conventional catheter angiography was performed to establish a definitive diagnosis. A selective right ventriculogram demonstrated findings consistent with a tetralogy of Fallot. The main pulmonary artery was of adequate size. There was a stenosis of the right pulmonary artery. The left pulmonary artery arose from the right pulmonary artery and it could not be visualized after a short segment. The delayed phase of the aortograms showed the right subclavian artery was opacified through the right vertebral artery. A small amount of contrast material passed from the proximal portion of the right subclavian artery to the right pulmonary artery through a patent ductus arteriosus. MDCT angiography was requested to evaluate the anatomy and size of the left pulmonary artery and the exact anatomy of the aortic arch. It was performed with a 16-slice MDCT scanner (Light-speed Ultra, GE Medical System, WI, USA) using a body weight-based low-dose protocol (80 kV and 25 mAs). The imaging data was acquired during an intravenous injection of 20 ml iodinated contrast agent (Iodixanol, Visipaque 320 mgI/ml, GE Healthcare, Milwaukee, WI, USA) at a rate of 2 ml/sec. The scanning delay is determined with a bolus tracking technique. The examination was initiated 4 seconds after the attenuation of region of interest positioned in the ascending aorta reaching 100 HU. For three-dimensional image reconstruction, the raw MDCT data were processed on a separate workstation (Advanced Workstation 4.2, GE Healthcare, Milwaukee, WI, USA). MDCT angiography showed a left aortic arch, which gave rise to two branches. The first branch was the common carotid trunk and the second was the left subclavian artery. The right subclavian artery did not arise from the aortic arch. It had an anomalous origin from the proximal right pulmonary artery through the patent right ductus arteriosus (Figs. 1, 2) Multiple tortuous aortopulmonary collateral arteries arising from the descending aorta were present. The left pulmonary artery arose from the posterior aspect of the right pulmonary artery and could not be seen after a short segment. However, only a 6.5 mm in diameter distal left pulmonary artery, supplied by aortopulmonary collateral arteries, was clearly seen in the left hilum. The anomalous origin and proximal interruption of the left pulmonary artery were diagnosed according to MDCT angiography findings. The patient was considered for staged surgical repair. The first stage of repair, a modified left-sided Blalock-Taussig shunt procedure, was performed to allow the growth of the left pulmonary arteries. The postoperative course was uneventful. Complete surgical correction of tetralogy of Fallot and isolated right subclavian artery was planned for a later date.

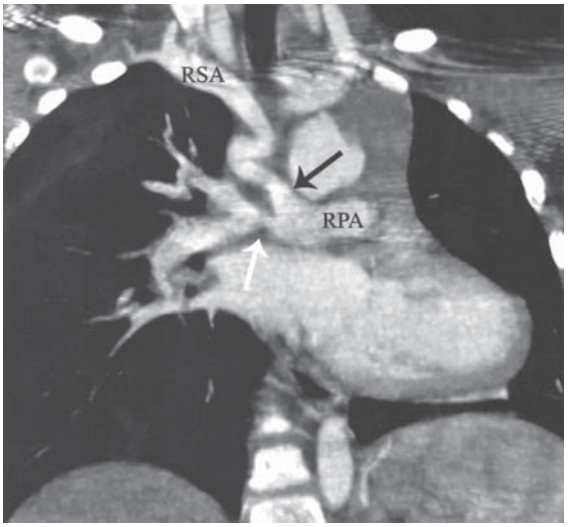

Fig. 1. Oblique coronal multiplanar reformatted image shows the right subclavian artery (RSA) arising from the right pulmonary artery by way of a patent ductus arteriosus (black arrow). There is a stenosis of the right pulmonary artery (white arrow).

Discussion

Isolation of the subclavian artery is generally observed on the side contralateral to the position of the aortic arch, but is well-known as involving the left subclavian artery four times more frequently than the right one, and is associated with intracardiac or aortic arch anomalies[4]. More than half of the reported cases of isolated left subclavian artery are associated with major cardiac malformation, of which tetralogy of Fallot is the most common[5]. An isolated right subclavian artery with aortic

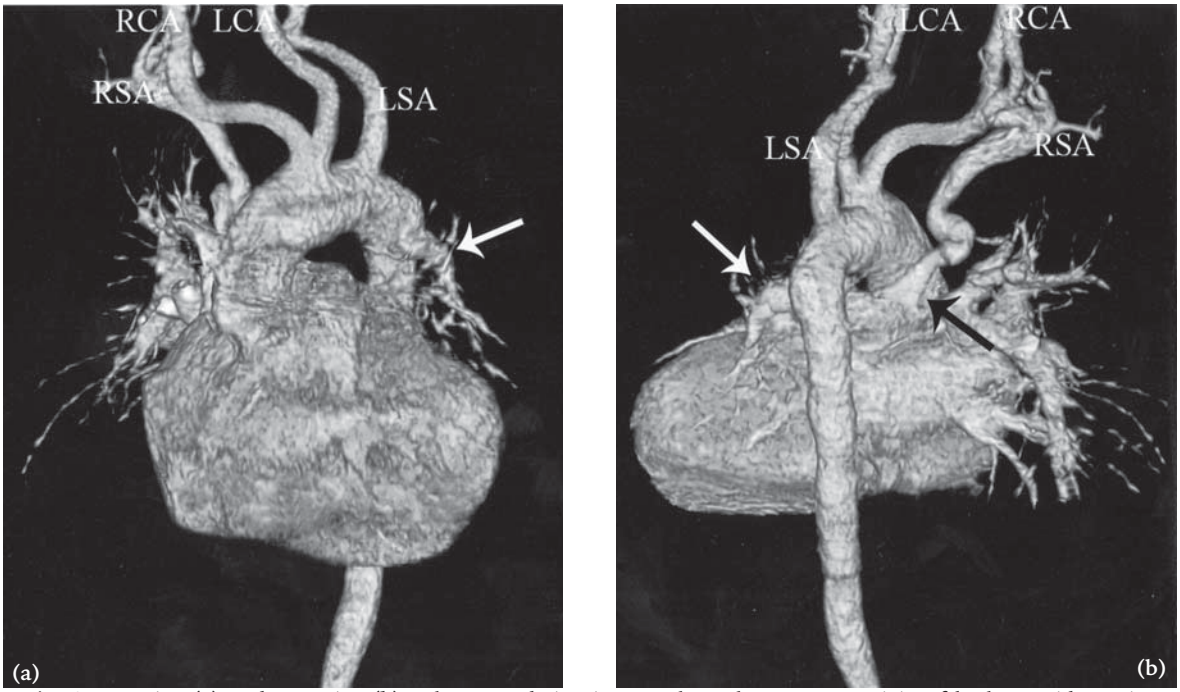

Fig. 2. Anterior **(a)** and posterior **(b)** volume rendering images show the common origin of both carotid arteries from the aortic arch and the isolated right subclavian artery arising from the right pulmonary artery by way of a patent ductus arteriosus (black arrow). Distal segment of the left pulmonary artery (white arrow) is also seen. (RSA: Right subclavian artery, RCA: Right carotid artery, LCA: Left carotid artery, LSA: Left subclavian artery).

arch interruption was first described in 1954[6]. However, the first clinical and angiographic report of an isolated right subclavian artery was published in 1980[7]. To the best of our knowledge, 13 cases of isolated right subclavian artery have been reported to date[1,4,6-14]. The most common association of an isolated right subclavian artery was D-transposition of the great arteries (five cases)[1,4,12-14]. It has been reported as an isolated finding in three cases[1,7,11], and associated with persistent ductus arteriosus (two cases)[4,8] ventricular septal defect[9], interrupted aortic arch[6], and tetralogy of Fallot[10].

Hemodynamic manifestations of an isolated subclavian artery depend on the state of the associated ductus arteriosus, and to some extent, on associated intracardiac anomalies. When the ductus arteriosus is obliterated, the isolated subclavian artery is supplied by collateral arteries from the head and neck through flow reversal in the vertebrobasilar system on the side affected. This results in the so-called congenital subclavian steal syndrome with abnormal blood flow to the affected arm and blood pressure difference between the upper limbs[4,9]. However, if the ductus arteriosus is patent, pulmonary artery steal can occur with flow down the vertebral artery through the ductus into the low-resistance pulmonary artery, and this can lead to congestive heart failure and pulmonary hypertension from the left-to-right shunt[5]. If the ductus arteriosus is patent and the pulmonary arterial pressure is suprasystemic due to a serious associated left-to-right shunt (ventricular septal defect or large contralateral patent ductus arteriosus with elevated pulmonary vascular resistances), the pulmonary arterial blood flow is stolen to the subclavian artery, resulting in lower oxygen saturation of the affected limb[5].

Various imaging modalities contribute to diagnostic information in patients with congenital cardiovascular disease. Traditionally, congenital cardiovascular disease was diagnosed and managed with the use of echocardiography and conventional catheter angiography, which have both advantages and limitations. Doppler echocardiography performs well in defining intracardiac anomalies and estimating hemodynamics. However, it is limited by a small field of view, a variable

acoustic window and difficulty in depicting extracardiac structures. Conventional catheter angiography is an invasive modality that gives important hemodynamic data and defines the anatomy of vessels that are accessible to catheterization. However, it often yields only indirect information regarding arterial anatomy distal to high-grade stenosis or atresia and it uses high doses of ionizing radiation and iodinated contrast material. Magnetic resonance imaging has increasingly been used for the diagnosis of congenital heart disease and aortic arch anomalies. Studies have shown that magnetic resonance imaging may be superior to echocardiography and that it is an excellent noninvasive alternative to catheter angiography since it can provide both anatomic and functional information about cardiovascular anomalies[15]. However, it also has some limitations. Long acquisition times may necessitate the sedation of young children and may present a problem in the examination of patients whose clinical condition is unstable. More recently, MDCT angiography is increasingly being used for rapid, accurate and noninvasive evaluation of aortic arch anomalies[2,3]. It has the advantages of easy availability and very short scanning time. MDCT angiography with three-dimensional reconstruction is especially useful in evaluating extracardiac structures. It enables the clear and complete depiction of size and morphologic configuration of the pulmonary arteries and the presence and location of aortopulmonary collateral vessels in patients with tetralogy of Fallot[2]. The drawbacks of MDCT include patient exposure to ionizing radiation and the risks of iodinated contrast material. However, adjustment of specific technical factors has been shown to minimize the radiation dose in children[2,3].

At present, complete surgical correction of tetralogy of Fallot is the procedure of choice. Alternatively, a palliative technique such as the Blalock-Taussig shunt is applicable in infants who are not candidates for complete repair, and this procedure suffices until the pulmonary artery branches reach an age and size that permit complete repair. The modified Blalock-Taussig shunt allows blood to flow from the subclavian artery to the ipsilateral pulmonary artery via a synthetic tube.

In conclusion, the existence of an isolated subclavian artery has an effect on the surgical approach to the associated congenital heart defects. In patients with tetralogy of Fallot, recognition of the isolated subclavian artery is crucial, because a Blalock-Taussig anastomosis from the subclavian artery to the pulmonary artery is usually carried out on the side opposite the aortic arch. Due to the decreased pressure and flow in an isolated subclavian artery, it cannot be used for this purpose. MDCT angiography enables a thorough preoperative evaluation of the complex cardiovascular anatomy in patients with tetralogy of Fallot.

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