

Abnormal Origin of the Left Subclavian Artery from the Left Pulmonary Artery in a Patient with Double Outlet Right Ventricle

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Anomalous aortic origin of the left subclavian artery (LSCA) from the left pulmonary artery (LPA) is a rare congenital cardiac malformation. We describe a case of LSCA from the LPA via ductus arteriosus in association with a double-outlet right ventricle, which never has been reported previously in Korea.

Key words: 1. Aorta, arch
2. Congenital heart disease
3. Embryology

CASE REPORT

Fetal echocardiography performed at 28 weeks of gestational age in a full-term 2,780-g male neonate showed a double-outlet right ventricle (DORV). The patient was born through a cesarean section with Apgar scores of 7 and 9 at 1 and 5 minutes, respectively. A physical examination demonstrated an acyanotic neonate without respiratory distress. The heart rate was 170 beats/min, and the respiratory rate was 54 beats/min. The blood pressure was 71/42 and 43/21 mmHg in the right and the left upper extremities, respectively. The left radial pulse was weak, but the right radial and femoral pulses were normal. The oxygen saturation was 95% in room air. Both lung sounds were clear, and the abdominal examination was unremarkable.

Two-dimensional echocardiography demonstrated the right aortic arch, anterior malalignment ventricular septal defect, overriding aorta (60%), and pulmonary and infundibular stenosis. These findings were consistent with DORV, Fallot

type. Patent ductus arteriosus (PDA) arose from a tortuous abnormal artery with bidirectional shunting. The left subclavian artery (LSCA) was not seen arising from the aorta. However, three-dimensional 64-row multidetector computed tomography (MDCT) showed the LSCA arising from the left pulmonary artery via ductus arteriosus (Fig. 1). Associated non-cardiac anomalies were also determined and included inguinal hernia, polydactyly, and syndactyly. The fluorescent *in situ* hybridization test was positive for the DiGeorge syndrome (22q11 deletion).

The patient underwent surgical repair through median sternotomy. The patient's age at the time of surgery was 28 days, and his weight was 3.8 kg. The aberrant LSCA was divided, and its pulmonary stump was oversewn. After appropriate trimming, the LSCA was directly reimplemented into the left common carotid artery (LCCA) with 8-0 polypropylene sutures (Surgipro; Tyco Healthcare USSC, Norwalk, CT, USA). We conducted near-infrared spectroscopy during the operation. The left cerebral oxygen saturation was maintained

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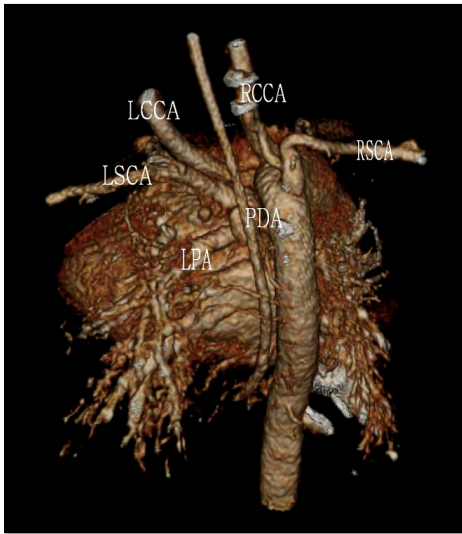


Fig. 1. Preoperative three-dimensional 64-row multidetector computed tomography showed the LSCA arising from LPA via PDA. LSCA, left subclavian artery; LPA, left pulmonary artery; PDA, patent ductus arteriosus; LCCA, left common carotid artery; RCCA, right common carotid artery; RSCA, right subclavian artery.

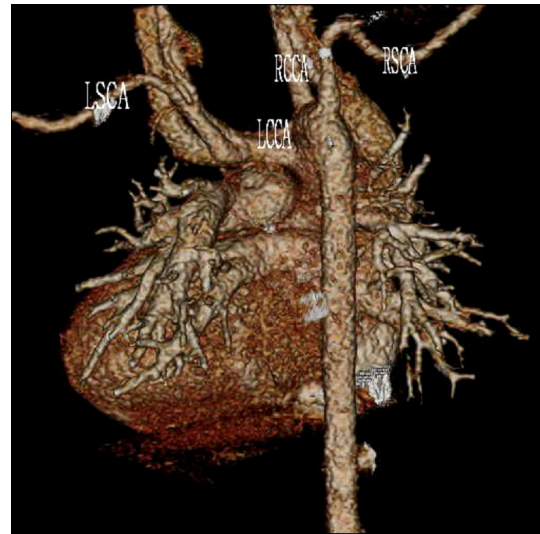


Fig. 2. Postoperative three-dimensional 64-row multidetector computed tomography revealed LSCA arising from the LCCA without stenosis. LSCA, left subclavian artery; LCCA, left common carotid artery; RCCA, right common carotid artery; RSCA, right subclavian artery.

above 80% of the baseline value during the clamping of the LCCA. After the operation, the blood pressure was symmetric in both arms. The patient was discharged from the hospital without any problems.

Fifteen months later, the patient underwent intraventricular tunnel repair of the DORV at 16 months of age. Three-dimensional 64-row MDCT at the time revealed the LSCA arising from the LCCA without stenosis (Fig. 2). The patient is now 18 months old and has normal left arm function and growth.

DISCUSSION

The right aortic arch with isolation of the LSCA is an uncommon arch anomaly in which the LSCA arises exclusively from the pulmonary artery via ductus arteriosus (DA) or ligamentum arteriosum without communication with the aorta [1]. The development of the aortic arch and its branches takes place during the third week of gestation. A common arterial trunk arises from the primitive heart and divides into six paired aortic arches that fuse and form bilateral dorsal aorta which, in turn, fuse caudally into the descending aorta. The

persistence or regression of these arches may lead to various arch anomalies [1-3]. Edwards' embryologic model of aortic arch malformation explains this LSCA isolation by the interruption of the left aortic arch at two locations: 1) between the left carotid and subclavian arteries, and 2) between the left ductus arteriosus and the left dorsal aortic root [3].

The isolation of the LSCA is commonly associated with congenital heart disease (CHD) and 22q11 deletion. Luetmer et al. reported associated CHD in 23 of the 39 cases (59%) of isolated LSCA. Further, the tetralogy of Fallot was the most common (14 of the 23 cases). Double-outlet right ventricle and d-transposition of the great artery have also been reported in a few cases [4-6].

The clinical presentations of patients with isolated LSCA depend on the patency of the DA. The isolation of the LSCA is usually asymptomatic and is usually discovered during the evaluations of the associated cardiac anomalies or when reduced blood pressure is detected in the left arm. In these patients, both congenital subclavian and pulmonary artery steal syndromes have been reported. If the direction of blood flow is from the left vertebral artery to the pulmonary artery (pulmonary steal syndrome) or the subclavian artery

(subclavian steal syndrome), the patient has vertebrobasilar insufficiency. Symptoms may include the disturbance of vision, faintness, syncope, and headache that can be exacerbated by exercising the left upper limb, resulting in increased left arm circulation. Patients with the pulmonary steal phenomenon are at risk of pulmonary overcirculation. Ischemic symptoms of the left arm, including pain, weakness, coldness, and a reduced limb length, may be present [3-7]. In a review of the 39 cases described by Luetmer and Miller [4], we found that 5 patients had ischemic symptoms of the left arm and 5 had vertebrobasilar insufficiency. They reported that the ages of the symptomatic patients ranged from 22 to 53 years, and the symptom duration before diagnosis ranged from less than 1 year to 11 years. The authors suggested that initially asymptomatic patients may lose the ability to compensate for the steal phenomenon and that eventually symptoms may develop with age [4].

The optimal therapeutic approach is still controversial. Simple ligation of the LSCA, surgical reimplantation, device occlusion of the PDA, and follow-up have been described as therapeutic options. Most authors have suggested that simple ligation of the LSCA or PDA is effective. However, persistent isolated LSCA, which was not surgically reimplanted into the aorta, provides an anatomic substrate for a subclavian steal syndrome [4-7]. Although this anomaly may be clinically asymptomatic, we recommend surgical correction before significant vertebrobasilar insufficiency or arm ischemia occurs. In our case, the LSCA was directly anastomosed with the LCCA to prevent symptomatic subclavian or pulmonary steal. Further, the patient now has normal growth of the left arm.

We think that very low operative mortality rates and high success rates justify a surgical approach in patients with isolated LSCA. Further, surgical reimplantation provides com-

plete abolition of the anatomic substrate for the subclavian steal syndrome.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Weinberg PM, Natarajan S, Rogers LS. *Aortic arch and vascular anomalies*. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, editors. *Moss and Adams heart disease in infants, children, and adolescents: including the fetus and young adult*. 8th ed. Philadelphia: Lippincott Williams & Wilkins; 2013. p. 758-98.
2. Uchino A, Saito N, Okada Y, et al. *Variation of the origin of the left common carotid artery diagnosed by CT angiography*. *Surg Radiol Anat* 2013;35:339-42.
3. Edwards JE. *Anomalies of the derivatives of the aortic arch system*. *Med Clin North Am* 1948;32:925-49.
4. Luetmer PH, Miller GM. *Right aortic arch with isolation of the left subclavian artery: case report and review of the literature*. *Mayo Clin Proc* 1990;65:407-13.
5. Mosieri J, Chintala K, Delius RE, Walters HL 3rd, Hakimi M. *Abnormal origin of the right subclavian artery from the right pulmonary artery in a patient with D-transposition of the great vessels and left juxtaposition of the right atrial appendage: an unusual anatomical variant*. *J Card Surg* 2004;19:41-4.
6. McMahon CJ, Thompson KS, Kearney DL, Nihill MR. *Subclavian steal syndrome in anomalous connection of the left subclavian artery to the pulmonary artery in d-transposition of the great arteries*. *Pediatr Cardiol* 2001;22:60-2.
7. Carano N, Piazza P, Agnetti A, Squarcia U. *Congenital pulmonary steal phenomenon associated with tetralogy of Fallot, right aortic arch, and isolation of the left subclavian artery*. *Pediatr Cardiol* 1997;18:57-60.