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Double aortic arch and tetralogy of fallot with pulmonary atresia

Çift aortik arkus ve pulmoner atrezili fallot tetralojisi

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Double aortic arch (DAA) is the most common form of complete vascular ring and results in respiratory and feeding problems because the trachea and the esophagus are circumscribed by the ring. It often occurs as an isolated anomaly; however, DAA may be associated with other congenital cardiovascular anomalies such as the tetralogy of Fallot (TOF), transposition of the great arteries, coarctation of the aorta, ventricular septal defect, and patent ductus arteriosus.[1-3] The coexistence of TOF and DAA is a rare entity.[2,4] We herein present our experience with a patient who was diagnosed with symmetrical DAA in conjunction with TOF along with pulmonary atresia, which resulted in tracheoesophageal compression symptoms.

CASE REPORT

A six-day-old girl weighing 2.37 kg was referred to our hospital with a diagnosis of pulmonary atresia and TOF. She was intubated immediately after birth due to severe respiratory problems. A physical examination showed the typical findings of TOF, and echocardiography confirmed the diagnosis of TOF with pulmonary atresia. Through a median sternotomy, a shunt was inserted between the right innominate artery and the right pulmonary artery using a 3.5 mm polytetrafluoroethylene (PTFE) graft. An intraoperative evaluation detected the presence of DAA, with both arches being patent and about equal in size. Postoperative multislice computed tomography (MSCT) confirmed the diagnosis of symmetrical DAA. The right subclavian and right common carotid arteries originated from the right aortic arch, whereas the left common carotid and left subclavian arteries originated from the left aortic arch (Figure 1). The trachea and esophagus were circumscribed and compressed by the vascular ring, resulting in tracheal stenosis (Figure 2). In the second operation, a left posterolateral thoracotomy was performed in which the right arch

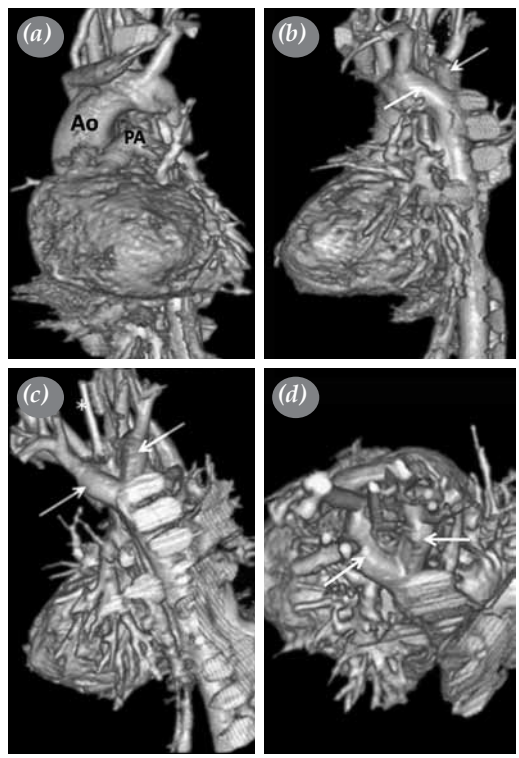

Figure 1. (a-d) Multislice computed tomography shows the pulmonary atresia and the anatomy of the double aortic arch (DAA) (white bold arrows), with the right subclavian and right common carotid arteries originating from the right arch, and the left common carotid and left subclavian arteries originating from the left aortic arch. **(c, d)** It also reveals the DAA which separated at the distal portion of the ascending aorta and joined at the proximal portion of the descending aorta. Ao: Aorta; PA: Pulmonary artery; *: PTFE graft.

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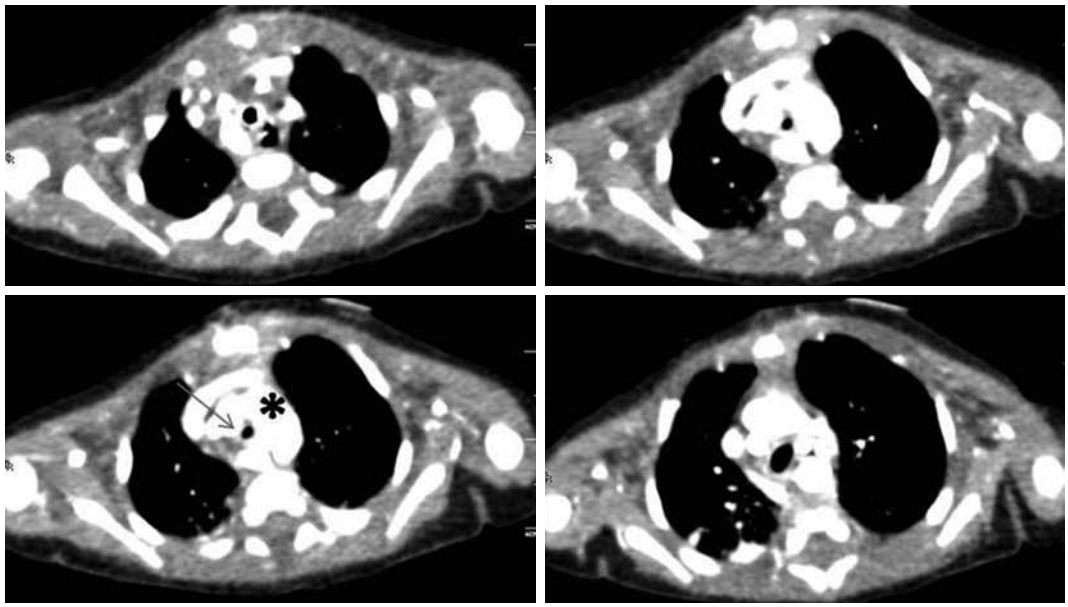

Figure 2. Multislice computed tomography images show that the trachea and esophagus were surrounded and compressed by the vascular ring (asterisk), resulting in tracheal stenosis (arrow).

was divided at the point where it joined the descending aorta, and the trachea and esophagus were then released. The patient's respiratory problems continued after the operation, and control MSCT showed that persistent tracheal stenosis. The patient died one month later as a result of postoperative respiratory failure.

DISCUSSION

Double aortic arch can be classically separated into three groups: dominant right aortic arch, dominant left aortic arch, and balanced aortic arch. Most of the patients have the dominant right arch type of DAA. The incidence rate of patients who have dominant right and left arches is only approximately 5%,[1-3] but symmetrical DAA was detected in our patient.

Surgical access depends on identifying the dominant arch. Compression of the esophagus and trachea is alleviated with the division of the atretic or small diameter aorta and the persistent ductus arteriosus (or ligamentum arteriosum).[1-3] When both arches have equal diameters, the division of the right arch is preferable in order to achieve normal anatomy. In our patient, we made the surgical decision based on the MSCT findings. We then performed a left thoracotomy and divided the right aortic arch at the point where it joined the descending aorta.

Any signs of tracheoesophageal compression in a newborn should suggest the possibility of a vascular ring. Successful results can be obtained by early diagnosis and timely surgical treatment in symptomatic patients. Furthermore, the presence of other cardiac pathologies and structural disorders resulting from compression on the tracheobronchial system are causes of significant morbidity and mortality. Multislice computed tomography accurately defines the vascular abnormality and shows the relationship of the arch with the neighboring structures. In addition, this case also shows that echocardiography has its limitations in the detection of extracardiac anomalies.

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