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Interrupted Aortic Arch Associated with AP Window and Complex Cardiac Anomalies: Multi Detector Computed Tomography Findings

Aortopulmoner Pencere ve Kompleks Kardiyak Anomalilerin Eşlik Ettiği Kesintili Arkus Aorta MDBT Bulguları

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Abstract

Interrupted aortic arch is a rare congenital malformation of the aortic arch defined as a loss of luminal continuity between the ascending and descending portions of the aorta. In a simple interrupted aortic arch, only a ventricular septal defect and patent ductus arteriosus are observed. We present a rare complex form of type A interrupted aorta with an aortopulmonary window, an atrial septal defect, a ventricular septal defect, and a patent ductus arteriosus on multidetector computed tomography (MDCT) imaging.

Key Words: Aortic interruption, Aortopulmonary window, Multidetector computed tomography

Özet

Kesintili arkus aorta, çıkan ve inen aorta arasındaki lümen sürekliliği kaybı olarak tanımlanan nadir bir konjenital anomalidir. Basit kesintili arkus aortada sadece ventriküler septal defekt ve patent duktus arteriozus görülür. Aortopulmoner pencere, atrial septal defekt, ventriküler septal defekt ve patent duktus arteriozus ile birlikte tip A kesintili arkus aortanın nadir bir kompleks formu olgusunun multidedektör bilgisayarlı tomografi bulguları sunulmuştur.

Anahtar Kelimeler: Kesintili arkus aorta, Aortopulmoner pencere, Multidedektör bilgisayarlı tomografi

Interrupted aortic arch is a rare congenital malformation that occurs in 3 per million live births [1-4]. This anomaly is defined as a loss of luminal continuity between the ascending and descending portions of the aorta [5]. Interrupted aortic arch typically occurs in association with an intracardiac malformation, such as a ventricular septal defect, patent ductus arteriosus, a bicuspid aortic valve, left ventricular outflow obstruction, or an aortopulmonary window [1-2]. In the presence of an interrupted aortic arch, the descending aorta receives its blood supply from a patent ductus arteriosus and systemic collaterals. The infusion of prostaglandin E1 to maintain ductal patency is an important component of preoperative stabilization. We report a rare complex form of type A interrupted aorta with an aortopulmonary window, an atrial septal defect, a ventricular septal defect, and a patent ductus arteriosus on multidetector computed tomography imaging (MDCT).

Case Report

A seven-day-old infant was admitted to our hospital with tachypnea and difficulties in feeding. After echocardiography, an atrial septal defect, a ventricular septal defect, a patent ductus arteriosus, and discontinuity of the aorta on the distal left subclavian artery (interrupted aortic arch or severe coarctation) were detected. MDCT imaging was performed to clearly visualize the lesions and to depict the associated lesions. MDCT revealed a type A interrupted aortic arch with an atrial septal defect, a ventricular septal defect, a patent ductus arteriosus, and a large communication between the ascending aorta and the proximal main pulmonary artery that presented an aortopulmonary window (Figure 1). The ductus arteriosus was widely evident and continued into the descending aorta. Cardiomegaly was also observed. Because the site of the interruption of the aortic arch in the present case was distal to the left subclavian artery, the interrup-

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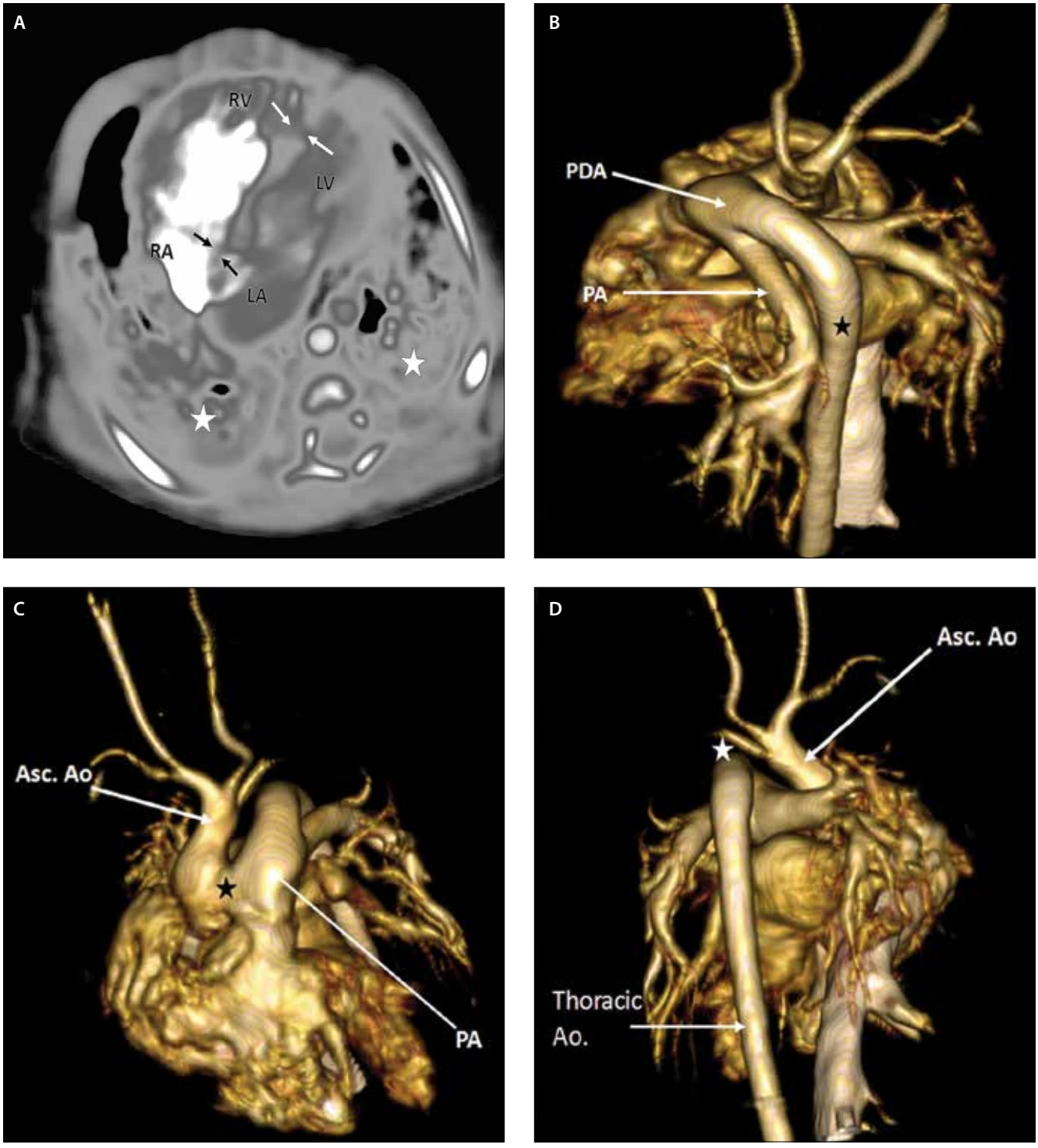

Figure 1. *A) Axial maximum intensity projection image showing an atrial septal defect (black arrows), a ventricular septal defect (white arrows), and lung consolidation (asterisks) (RA: right atrium, LA: left atrium, RV: right ventricle, LV: left ventricle). B) Left posterolateral view with three-dimensional volume rendering showing a patent ductus arteriosus and continuation of the ductus arteriosus into the thoracic aorta (asterisk) (PA: pulmonary artery). C) Anterior view with three-dimensional volume rendering showing a large aortopulmonary window (asterisk) between the ascending aorta (Asc. Ao) and the pulmonary artery (PA). D) Posterior view with three-dimensional volume rendering showing a type A interrupted aortic arch (asterisk) (Asc. Ao: ascending aorta).*

tion was identified as a type A interrupted aortic arch. An infusion of prostaglandin E1 was administered to maintain ductal patency for preoperative stabilization. Surgical correction of this anomaly to connect the two separate portions of the aorta, close the atrial and ventricular septal defects, and ligate the patent ductus arteriosus was performed during the neonatal period.

Discussion

Interrupted aortic arch is a rare congenital malformation of the aortic arch defined as a loss of luminal continuity between the ascending and descending portions of the aorta. This anomaly is associated with a very poor prognosis without surgical treatment [1]. The disruption between the ascending and descending aorta is classified according to the state of the interruption [6-7]:

- Type A: The interruption occurs on the distal left subclavian artery (30% of cases).
- Type B: The interruption occurs between the left carotid artery and the left subclavian artery (43%).
- Type C: The interruption occurs between the innominate artery and the left carotid artery (17%).

Interrupted aortic arch is a rare and usually fatal cardiovascular anomaly that requires early medical and surgical care of affected infants. In a simple interrupted aortic arch, only a ventricular septal defect and patent ductus arteriosus are observed. The complex form is associated with truncus arteriosus, transposition of the great arteries, a double-outlet right ventricle, an aortopulmonary window, and a single functional ventricle. With rare exceptions, patients with an interrupted aortic arch have either a ventricular septal defect (80%-90%) or an aortopulmonary window (10%-20%) [8-9]. In our case, we present a rare complex form of a type A interrupted aorta with an aortopulmonary window, an atrial septal defect, a ventricular septal defect, and a patent ductus arteriosus on MDCT imaging.

The diagnosis of interrupted aortic arch may be suspected based on the presenting symptoms of the infant. During the preoperative evaluation, echocardiography may not differentiate between interrupted aortic arch and severe aortic coarctation with a hypoplastic arch. In these cases, the assessment is complemented by computed tomography or magnetic resonance imaging. The morphologic features of interrupted aortic arch, its anatomic relationship, and the potential complex associated findings can be demonstrated using these techniques because of their multiplanar capabilities. Combined multiplanar reformatting and three-dimensional imaging using MDCT is a noninvasive imaging method that can clearly display an interrupted aortic arch and collateral vessels and the anatomical structures. The advantages over magnetic resonance angiography include shorter acquisition times, which reduces the need for sedation and provides the ability to scan extremely ill patients who cannot tolerate the long imaging times required for magnetic resonance imaging examinations, and the simultaneous evaluation of the airway and the lung [10]. Coronary angiography effectively shows the anatomical structure, but it is time consuming and invasive.

An infant with an interrupted aortic arch requires an alternate route to obtain adequate blood flow in the lower body. In such cases, the descending aorta receives its blood supply from a patent ductus arteriosus and systemic collaterals. An infusion of prostaglandin E1 to maintain ductal patency is an important component of preoperative stabilization [11]. Once the diagnosis is suspected and confirmed, treatment and surgical intervention are vitally important. The immediately required treatment includes the administration of a prostaglandin infusion to allow blood flow to the lower body until surgery is performed to re-establish the continuity of the aortic arch. Open-heart surgery connects the two separate portions of the aorta, closes the ventricular septal defect, and ligates the patent ductus arteriosus.

In conclusion, in such congenital cardiovascular diseases with potential complex associated findings, such as a type A interrupted aortic arch that requires immediate treatment and surgical intervention, MDCT should be used for a fast and reliable diagnosis.

Conflict of interest statement: The authors declare that they have no conflict of interest to the publication of this article.

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