A Case of Anomalous Left Coronary Artery Arising From the Pulmonary Artery in Adulthood: Multidetector Computed Tomography Coronary Angiography Findings

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Abstract

Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) is a rare but very serious congenital coronary artery anomaly. Multidetector computed tomography (MDCT) coronary angiography has recently become the gold standard for depicting anatomical variations and anomalies of the coronary arteries because the origin and course of anomalous arteries can be demonstrated very accurately by this technique. In this report, we present a case of 22-year-old female who was admitted to our emergency department with cardiac arrest. In the course of diagnosis, MDCT coronary angiography revealed a left coronary artery arising from the pulmonary artery as well as marked dilatation of the coronary arteries.

Key Words: ALCAPA, Anomalous left coronary artery, MDCT coronary angiography, Pulmonary artery

Introduction

Anomalous origin of the left main coronary artery (LCA) from the pulmonary artery (PA) is a rare malformation also known as Bland-White-Garland syndrome. Very few individuals with anomalous left coronary artery arising from the pulmonary artery (ALCAPA) survive until adulthood without surgical repair; furthermore, approximately 90% die suddenly at a mean age of 35 years [1]. In the most common form of this disease, the LCA arises from the PA, and the right coronary artery (RCA) arises normally from the aorta. This configuration creates a collateral circulation between the RCA and LCA and a coronary “steal” phenomenon into the PA [2]. In this article, we present clinical and MDCT coronary angiography findings of a 22-year-old female patient with ALCAPA with no history of chest pain.

Case Report

A 22-year-old female with no history of chest pain was admitted to our emergency department with sudden cardiac arrest. Following the reinstitution of the cardiopulmonary circulation and medical management, a transthoracic echocardiographic examination was performed, revealing an obvious dilatation of the coronary arteries. The patient was referred to the radiology department on suspicion of coronary artery anomaly and scanned with a 16-detector CT scanner (Philips Medical Systems MX 8000 IDT Multislice CT System-V 2.5). The informed consent of the patient’s parents was obtained before radiological examination. Scan parameters were 120 kV, 340 mAs, 420 msec rotation time with a slice thickness of 1 mm and increments of 0.5 mm, using a detector collimation of 16x0.75 mm (pitch: 0.2). The heart rate was 67 beats per minute during acquisition. One hundred milliliters of non-ionic, iodinated, low-osmolar contrast medium was injected through the antecubital vein at a rate of 5 ml/sec. Subsequently, a bolus of 40 ml saline solution was administered at a rate of 2.5 ml/sec to avoid possible contrast artifacts at the right atrial entrance. An automatic bolus tracking method was used to optimize visualization.

MDCT coronary angiography images demonstrated a LCA originating from the inferior side of the main PA. MDCT showed enormous sinus vessels originating from a normally situated right coronary artery with collateral circulation to the LCA. The lumen of the LCA was fully enhanced, and a little enhancement was noticed in the lumen of the PA, showing retrograde blood flow toward the PA. The RCA arose from the aorta. All coronary arteries and their branches were ectatic and showed mildly tortuous courses. Collateral circulation between the RCA and LCA as well as both a coronary “steal” phenomenon (from RCA to LCA) and a pulmonary “steal” phenomenon (from LCA to main PA) were readily demonstrated (Figure 1a-c).
The patient died from massive cerebral and cerebellar infarction 12 hours after the MDCT coronary angiography examination. Therefore, surgical correction could not be performed.

Discussion

Coronary artery anomalies are common in the general population. They may accompany other congenital heart diseases and can cause myocardial ischemia, a decrease in life expectancy, or sudden death. Most patients with coronary artery anomalies are asymptomatic [1]. ALCAPA is one of the most serious congenital coronary artery anomalies. It has an estimated prevalence of one in 300,000 live births [3]. Most of the affected patients show symptoms in infancy and early childhood. Approximately 90% of untreated infants die in the first year of life, and only a few patients survive until adulthood [3].

Abnormal embryonic development may lead to positional anomalies of the coronary arteries. The coronary artery buds appear on about the 12th day of life after the division of the truncus arteriosus, which leads to the separation of the aorta and pulmonary artery [4]. Coronary artery anomalies can arise secondary to either malrotation of the spiral septum dividing the truncus or malpositioning of the coronary buds themselves. Soloff described the four possible anomalous coronary artery connections to the pulmonary artery [5]. The most common is anomalous left coronary artery from the pulmonary artery (ALCAPA), followed by anomalous right coronary artery from the pulmonary artery (ARCAPA), origin of both coronaries from the pulmonary trunk, and origin of an accessory coronary artery from the pulmonary trunk. The relatively high incidence of ALCAPA is explained by the proximity of the left coronary bud to the pulmonary artery sinus.

Conventional angiography is a traditional technique that has been used for many decades to diagnose coronary artery anomalies [2]. However, MDCT has recently become accepted as the main technique for evaluation of coronary anomalies [6, 7]. Several case reports describing MDCT coronary angiography findings in ALCAPA have been published previously [1, 7]. In our experience, unlike conventional angiography, MDCT coronary angiography allows the accurate and noninvasive depiction of coronary artery anomalies. MDCT has been determined to be superior to conventional angiography for delineating the origin, course, and termination of abnormal coronary arteries [6, 7]. Familiarity with the CT appearances of various coronary artery anomalies and an understanding of the clinical significance of these anomalies are essential for making a correct diagnosis and planning treatment.

Most infants with ALCAPA die within the first year of life without treatment, and survival beyond infancy is dependent upon adequate collateral circulation from the RCA or another source. These lesions are rarely tolerated well in the neonate, as the pulmonary vascular resistance is still high, leading to high-pressure antegrade flow down the LCA [1]. In our case, the existence of collateral vasculature between the LCA and RCA, which caused the coronary steal phenomenon and supplied normally oxygenated blood to the RCA's territory, was the mechanism of survival until adulthood.

Treatment of ALCAPA consists of surgical re-creation of dual coronary perfusion. In the infantile type of ALCAPA, reimplantation of the anomalous LCA directly into the aorta or creation of an intrapulmonary conduit from the left coronary ostia to the aorta, known as the Takeuchi procedure, can be implemented. In the adult type of ALCAPA, ligation of the LCA from the pulmonary artery combined with coronary artery bypass grafting can be performed [3]. Our case was asymptomatic, so this serious coronary anomaly remained undiagnosed until the patient had a heart attack. Magnetic resonance imaging (MRI) was performed following MDCT. There were extensive ischemic areas in both the cerebrum and the cerebellum on MRI. Because the patient died 12 hours after MDCT coronary angiography, no surgical intervention could be performed.

In conclusion, ALCAPA is very rare in adults without a previous clinical history, as in the presented case. However, a radiologist encountering ectatic coronary arteries in an MDCT coronary angiography scan should always consider ALCAPA as a possible diagnosis. Determining the origins, courses and terminations of the coronary arteries is very important in the diagnosis of this syndrome.
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References


