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Anomalous Origin of the Left Coronary Artery from Pulmonary Artery (ALCAPA)

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Abstract

Congenital abnormalities, including anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), and lethal arrhythmias in a structurally normal heart explain about 5-10% of sudden cardiac death (SCD) cases. In this narrative review, the available literature pertaining to the rare congenital coronary artery malformation, ALCAPA, shall be comprehensively reviewed and summarized.

The majority of patients with ALCAPA present in early life with symptoms of ischemic heart disease often causing death. Alternatively, if sufficient collateral formation between the right and the left coronary arteries is present, these patients may be asymptomatic throughout their entire childhood. Since ALCAPA is rarely seen in pediatric patients and even less in adults, the diagnosis requires a high index of suspicion. The development of novel non-invasive high resolution imaging techniques such as echocardiography, multi detector computed tomography (MDCT) and cardiac MRI has allowed detailed visualization of the origin and course of the anomalous artery. Currently, surgical correction of the malformation is the optimal treatment option which can lead to significant improvement in myocardial ischemia. It is important to note, despite ALCAPA as a rare condition, this differential diagnosis should be always considered in an otherwise healthy young adult individual who presents with SCD, syncope, exercise intolerance or chest pain.

Keywords: ALCAPA; Sudden cardiac death; Anomalous origin of the coronary arteries; Syncope; Exercise intolerance; Dyspnea; Chest pain

Epidemiology

Heart disease represents the leading cause of death in the US as confirmed by the Centers of Control of Disease (CDC) in 2011 [1]. ALCAPA is infrequent (1 of 300,000 births), although suspected to be under diagnosed accounting for 0.25-0.5% of congenital heart disease anomalies and is a rare cause of SCD in completely asymptomatic adults [2,3]. It is also infrequent that an individual with ALCAPA achieves adulthood without corrective surgery, as 90% of these patients would die within the first year of life without surgical correction of the abnormality [4,5].

Etiology

Theories have been described to explain the origin of a coronary artery from the pulmonary trunk specifically related to the embryologic division of the truncus arteriosus. Assuming that the coronary arteries arise as two endothelial buds, displacement of the site of origin of one or both coronary artery buds could assign either of both coronary arteries to that portion of the truncus arteriosus destined to become the pulmonary artery [6]. Another possibility is the abnormal division of the truncus by the developing truncal septum incorporating one of both normally located coronary artery buds into the pulmonary artery [6].

Clinical Presentation

Patients with ALCAPA usually present with symptoms early within the first year of life with chronic ischemia due to this coronary artery malformation leading to congestive heart failure. Patients can also present with significant mitral valve regurgitation. Infants with ALCAPA typically have feeding difficulties, irritability, diaphoresis, tachypnea and tachycardia. Associated chest pain can occasionally be due to myocardial ischemia which may be confused with infantile colic. If there is enough collateral formation between the right and the left coronary arteries, patients can pass through childhood with silent manifestations. Adult patients with this malformation can complain of dyspnea, chest pain, syncope and exercise intolerance, or they can present with SCD due to acute ischemia during exercise or malignant ventricular arrhythmias generated from myocardial scar tissue [7,8]. Physical findings include a gallop or murmur of mitral regurgitation secondary to ischemic papillary muscle dysfunction. A continuous murmur resembling a patent ductus arteriosus with flow from the aorta to the pulmonary circulation can be found reflecting collateral flow. A retrospective study examining coronary arteriography of 5400 adult patients found that all coronary artery anomalies have an 80% association with arteriosclerotic disease and 40% with significant stenosis (>50%) in anomalous coronary arteries [9]. These results can partially explain the potential presentation of ALCAPA as acute coronary syndrome in the adult population.

Diagnosis

The diagnosis of ALCAPA is based on clinical symptoms and requires a high index of suspicion. Due to its potential catastrophic presentation, an increased awareness in health care providers during pre-athletic screening is needed. The electrocardiogram (ECG) of patients with ALCAPA usually shows typical signs of anterolateral myocardial infarction in 55-80% while ECG changes in infancy have also been delineated [10,11].

In the past, ALCAPA was diagnosed exclusively by cardiac catheterization and angiography. However, recent technical advances in echocardiographic imaging and multidetector computed tomography (MDCT) coronary angiography provide better visualization of the anomalous coronary artery origin and associated findings [12]. Transthoracic echocardiographic signs of ALCAPA include abundant septal color flow doppler signals, dilation of the right coronary artery (RCA) and, according to Ghaderi et al., a unique finding of pulsed-wave doppler at the right coronary ostium showing systolic coronary flow predominance [13]. MDCT is used as a confirmatory test with direct visualization of the coronary arteries origins including a dilated tortuous right coronary artery and intercoronary collateral vessels [14]. Conventional angiography has limitations due to its projectional vascular overlap, inability to correctly identify an interarterial course and invasive nature [15]. Other techniques recently developed include transesophageal echocardiography and magnetic resonance angiography (MRCA). In numerous studies comparing the reliability and diagnostic accuracy of MRCA with coronary angiography, MRCA unambiguously visualized the proximal coronary artery, even in cases where coronary angiography was equivocal [16-19].

Treatment

Prior to treatment, assessment of myocardial ischemia is identified by exercise treadmill, 201TI SPECT, or dobutamine stress echocardiography [20]. Moreover, echocardiographic post-systolic shortening and altered longitudinal strain have recently been described as a potential useful marker of subclinical ischemic dysfunction in a patient with ALCAPA [21].

The definitive treatment of this anomaly is surgical intervention. Zheng et al, described the treatment of 23 patients aged 2.5 months to 65 years utilizing four surgical interventions; left coronary artery (LCA) ligation, LCA ligation plus coronary artery bypass grafting, the Takeuchi procedure or LCA reimplantation [22].

Six patients in this series died in hospital after surgery and the remainder of the cohort had no overt symptoms during a 6 to 166 months follow-up period. Survivors' abnormal Q waves gradually regressed or disappeared, and left ventricle (LV) ejection fraction (EF) and LV size returned to normal range with alleviation of mitral regurgitation [22].

In a report of 151 adult patients with ALCAPA by Yau et al. 21% of the patients underwent LCA ligation, whereas 79% had establishment of dual coronary perfusion [23]. The overall surgical mortality was 1-4% with postoperative stress ECG and imaging studies demonstrating improvement in ischemia in 90% and 93% of cases respectively [23]. Of the 7 medically treated patients with reported follow-up, 5 were described as stable 5 years after diagnosis.

Establishment of a dual coronary system with coronary transfer is preferred even if an interposition graft is required in adults [24].

Discussion

Heart disease persists as the leading cause of death in the US as per the Centers for Disease Control and World Health Organization [1]. Each year in the United States, 180,000 to 250,000 people die of unexpected SCD in an emergency department (ED) or before reaching a hospital [25].

The majority of cases of SCD in women are currently undetermined, whereas in men coronary artery disease is the leading cause [25]. ALCAPA is one of the coronary artery anomalies that can potentially produce SCD in an adult. The symptoms of this disease, ranging from silent to exercise intolerance, chest pain, syncope, dyspnea or SCD can mimic an otherwise common coronary ischemic equivalent. The ECG in patients with ALCAPA similarly shows non-classic findings of ischemia. As such, novel imaging modalities have been described as superior in identifying the anomalous origin of the coronary artery as seen in ALCAPA more so than invasive coronary angiography. Literature has described common doppler echocardiographic changes of this disease and expanded use of this imaging technique could potentially serve as screening tool in pre-athletic evaluation particularly in individuals with a clinical index of suspicion. The use of more advanced imaging modalities like MDCT and cardiac MRI should then be used as a confirmation. The importance of the early identification of ALCAPA in this clinically challenging diagnosis is based on the favourable clinical outcome in surgically treated patients.

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