

An Autopsied Case of a Neonate with the Anomalous Origin of Total Coronary Arteries from the Pulmonary Artery

Case Report

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Abstract

Coronary artery anomaly is not a very common disease. A neonate with the origin of total coronary arteries from the pulmonary artery (TCAPA) is extremely rare and fetal, which has not been yet reported in China. Here an autopsied case of TCAPA is reported to provide reference for the medical colleagues. A female infant died 9 hours after born when the hypoxia symptoms presented, although the rescue measures had already done. The cause of death was identified as TCAPA by forensic pathologists. In this case, TCAPA was ignored by the doctors because of non specifically clinical presentations. If it had been diagnosed, surgery should have been done immediately to be able to supply enough blood for the heart.

Keywords: Congenital Heart Disease; Coronary Artery Anomaly; Total Coronary Arteries Origin From Pulmonary Artery (TCAPA); Coronary Angiography.

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Introduction

Coronary artery anomaly (CAA), which includes the origin, course and structure, exists in 0.9 -1.4% of patients in pediatric population [1-2]. The anomalous origin of one coronary artery (left or right coronary artery, ALCAPA or ARCAPA) is commonly reported [2-4]. Total origin of the coronary arteries from pulmonary artery (TCAPA), however, has been rarely presented [5,6]. This paper introduced a case of an infant with TCAPA, who suffered from severe hypoxia. The pathophysiological, clinical presentations, diagnosis, and treatment were briefly discussed as well.

Case history

The victim was a female infant, who was born to her 29-year-old mother at 39 weeks of gestation, and died 9 hours after elective

caesarean section B- scan ultrasonography showed that the fetal heart rate was 144 beats per minute, and umbilical cord entangled around the fetal neck 1 coil. Apgar score was 3, 0, 3 at 1, 5, 10 min respectively. The umbilical cord entanglement around neck was 3 coils, but no sign of constrictions was found. The physical examination showed heart beat at 60 beats per minute and respiration of 16 breaths per minute; the pulses were absent in the extremities, which were pale and cold. The diagnosis of “severe asphyxia or congenital heart disease” was in consideration. Heart rate and breath returned following emergency intubation, peak airway, and transtracheal naloxone and adrenaline injection. Then the baby was transferred to another hospital for further therapy. The blood analysis showed acidosis and oxygen saturation of 18%. The final diagnosis was inferred as “neonatal asphyxia, pneumonia, multiple organ dysfunction syndrome, hypoxic ischemic encephalopathy, or intracranial hemorrhage”. She finally collapsed despite resuscitation efforts. Forensic autopsy was carried out in 2 days after death. Microscopically, left main and right coronary arteries (LMCA and RCA) originated from two separate ostiums of pulmonary artery (TCAPA). No other specific changes were found.

Discussion

Normally, the coronary arteries originate from the sinus of valsalva. Origin of one or both coronary arteries from the pulmonary artery is a fetal malformation, which happens in approximately 1 of 300,000 live births [9]. The incidence of ALCAPA (also called Bland–White–Garland syndrome) was ranged 1:50,000 to 1:300,000 [10,11], while the incidence of the congenial heart disease and the ARCAPA was 0.25-0.50 % [10,12], 0.003% respectively [12], but the TCAPA incidence remained unknown.

Previous studies discovered [5,13-14] that in utero, the umbilical vein transfers arterial blood from placenta to the body of fetus. Due to the presence of foramen ovale, arterial catheter, com-

pressed fetal lungs, the pressure of pulmonary artery is very close to aorta. Therefore, the coronary arteries origin anomaly has no effect on the fetal. During the process of childbirth, umbilical cord blocked and expansion of alveolar causes the reduction of the pressure of pulmonary circulation, an increase of the flow of the pulmonary venous blood in the meanwhile. TCAPA cannot provide enough oxygen and blood for myocardium, leading to myocardial ischemia and hypoxia [5]. What is different from TCAPA patient is that patients of LCAPA, RCAPA can survive for long time caused by the change of pulmonary artery pressure and the establishment of an extensive intertrial collateral circulation [13,14]. The clinical features of the fetus suffering from TCAPA before birth can be asymptomatic, but after birth it may show symptoms such as severe myocardial ischemia and hypoxia [5].

In this report, because of short duration of death, histological examinations didn't provide proof for the ischemia of heart. The baby was in good conditions before birth. But after the baby was born, the symptoms of severe asphyxia immediately occurred, which was consistent with the above findings. Ultimately the infant died of respiratory insufficiency caused by TCAPA. She was suspected of congenital heart disease by the first doctor, but he did not take effective methods for further diagnosis. As a result, the baby was misdiagnosed as "pneumonia, encephalopathy" in the second hospital. Therefore doctors should pay much more attention to identify the TCAPA and other congenital heart diseases, in order to save the baby's life and avoid medical disputes.

Because of no signs of the umbilical cord constrictions, the cause of death by it could be excluded. Clinically, cardiac auscultation, computerized tomography scan, magnetic resonance imaging and electrocardiograms are essential for the diagnosis of TCAPA, especially coronary angiography is the "gold standard" of the diagnosis [15]. Once the disease is identified, surgery should be promptly performed to guarantee the blood supply for two coronary arteries, so as to ensure the normal function of the heart.

In conclusion, we would like to stress the importance of careful examination of the cardiovascular system in the neonate cases of sudden death.

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