ISCHEMIC CARDIOGENIC SHOCK IN THE INFANT
BY ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY
FROM THE PULMONARY ARTERY

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ISCHEMIC CARDIOGENIC SHOCK IN THE INFANT BY ANOMALOUS ORIGIN OF THE
LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (Abstract): Anomalous
origin of the left coronary artery (LCA) in the pulmonary artery (PA) is a commonly asympto-
matic malformation at birth until the early postpartum PA when pressure drops to a critical level,
causing congestive heart failure or myocardial infarction with cardiogenic ischemic shock. The
authors present the case of an infant aged one month, which is admitted for clinical signs of con-
gestive heart failure associating hypotension and prolonged recolorating time with dilated cardio-
myopathy diagnosed by bidimensional transthoracic echocardiography and myocardial infarction
in the electrocardiogram. Under treatment the evolution was unfavorable and the macroscopic
anatomopathologic examination confirmed the abnormal origin of LCA from PA, dilated cardio-
myopathy, subendocardial necrosis and secondary fibroelastosis. Key words: CONGENITAL
ANOMALY OF THE CORONARY ARTERIES, CONGESTIVE HEART FAILURE, DILATED
CARDIOMYOPATHY, SECONDARY FIBROELASTOSIS

INTRODUCTION

Coronary artery anomalies represents a mi-
nor group of congenital heart diseases that can
evolve on their own or in combination with
other congenital cardiac anomaly. In the ab-
sence of structural heart disease, coronary ar-
tery anomalies can cause dilated cardiomyopa-
thy, and cardiogenic shock (1).

During fetal life, the origin of the LCA from
the pulmonary trunk is well tolerated because
the pressure and saturation in pulmonary and
systemic circulations are equal. Therefore, the
flow in the LCA is antegrade with relatively
saturated blood (2). At birth, the infant is asympto-
matic but during the early months of life the
pulmonary pressure falls below systemic pres-
sure, which results in left-to-right shunt from
the higher pressure left coronary arterial system
to the lower pressure pulmonary arterial sys-
tem. This is known as the coronary steal phe-
nomenon (2). Circulatory insufficiency, myo-
cardial infarction, and life-threatening cardiac
dysrhythmias are the most common clinical
presentations during infancy (2, 3). Older pa-
tients carry the risk of sudden death due to
myocardial infarction, left ventricular dysfunc-
tion and mitral insufficiency, or silent myocar-
dial ischemia.

Case presentation

A one-month-old girl presented with a two-
week history of loss of appetite, hipercreme
urine and the etiological investigation of a lat-
erovesicate tumor straight detected during an
abdominal ultrasound performed as an outpa-
tient and is hospitalized in the Department of
Nephrology of the Clinical Emergency Hospital
for Children „Saint Mary”. At about 3 hours
of hospitalization overall condition worsens,
appears shortness of breath with polipnee,
moaning, thoraco-abdominal balance, cyanosis
with SpO2 below 90% in the atmosphere, the
patient becomes addicted to additional source of oxygen reasons and is transferred to the Department of ICU.

On physical examination the baby was eu­
­trophic (IP = 0.97), with severe overall condi­
­tion, conscious, with flabby turgor, dry mucous membranes, pale, dry, marbled skin fold persistently depressed anterior fontanelle, tachyp­
­neic (74 breaths/ minute), general intercostal circulation, thoraco-abdominal balance, expira­tory moan without crackles, tachycardia (170 b/min) without murmurs, but with low BP (76/53 mm Hg) and prolonged recolored capil­
­lary time (> 5 seconds), along with pale, mot­tled and cold extremities. Hepatomegaly (liver 4 cm below the costal margin) and neurological sufference with messy Moro reflex draws attention to a congestive heart failure. Emergency performed echocardiography describes dilated cardiomyopathy, diastolic dysfunction of the left ventricle, severe mitral insufficiency, severe tricuspid insufficiency and left to right perme­
able foramen ovale. Repeated echocardiogra­phy could not prove the anomalous origin of LCA from PA (fig. 1, fig. 2).

Thoraco-abdominal radiography reveals ple­
­thoric lungs, cardiomegaly and hepatomegaly.

Abdominal ultrasound confirms hepatome­
galy, denies the presence of a laterovezicale tumor, and describes kidney with bilateral hydro­
­nephrosis I degree, free liquid between kidney, live, bilateral subfrenic and pelvic, which tends to aglutinates.

Electrocardiogram (ECG) describes sinus tachycardia with left ventricular overload and diffuse secondary changes of repolarization. Serial ECG highlights the elevation of ST seg­
­ment (2-3 mm) in the right precordial leads (fig. 3).

The biological exploration shows normo­
cytic normochromic anemia (Hb = 7.3 g/dl, and 8.2 g/dl), metabolic acidosis (RA = 14.2 mmol/L), cytolytic liver syndrome, cytolitic enzyme syndrome (CPK = 2621 U/L, CK-MB wasn’t available). IgM, IgG anti-CMV antibod­
­ies and anti-rubella virus were positive. Urinalysis, stool and blood cultures were negative.

Under treatment with IV infusion, dopa­
­mine, dobutamine, broad-spectrum antibiotics, furosemide, spironolactone, oxygen, the red blood cell transfusion, the general condition remains serious, it’s oxgeno-dependent heart failure worsens, making it impossible to transfer baby to a specialized and more experienced center for further investigation in determining with certainty the origin of LCA and dies.

The pathological exam describes cardio­
megaly with left ventricular dilatation (fig. 4), abnormal origin of LCA based on PA, endocar­
dial fibroelastosis (fig. 5). Microscopic exami­nation reveals marked myocardial myocytes suffering recent outbreaks of necrosis and marked areas of vacuolar degeneration (fig. 6) and areas calcification of subendocardial necrosis.

**DISCUSSION**

Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital coronary abnormality. It has an estimated incidence of about 1 in every 300,000 live births and comprises between 0.24% and 0.46% of all congenital cardiac diseases (1, 2, 3). It is an isolated defect in about 95% of the cases;
Aniela Rugină et al.

Fig. 3. ECG: The elevation of ST segment (2-3 mm) in the right precordial leads

Fig. 4. Cardiomegaly

Fig. 5. Ventricular endocardial fibroelastosis, right coronary artery originating from the aorta

Fig. 6. Microscopic examination: outbreaks of recent necrosis and marked vacuolar degeneration

however, in 5% of cases it may be associated with other cardiac anomalies such as coarctation of the aorta, arterial septal defect, and ventricular septal defect (2). ALCAPA has a mortality rate up to 90% within the first year of life if left untreated. However, there are several cases reported in adolescents and adults (4). This is attributed to the development of collaterals between the right and left coronary arteries (5).

Anomalous origin of LCA from PA was first described in 1866 by Brooks HSJ (1), and in 1933 was published for the first time the relationship between the clinical and pathological changes induced by this congenital malformation by Bland EF (5), the anomaly is known as Bland-White-Garland syndrome or syndrome ALCAPA (Anomalous origin of the Left Coronary Artery from the Pulmonary Artery). During fetal life, the origin of the LCA from the pulmonary trunk is well tolerated because the pressure and saturation in pulmonary and systemic circulations are equal. Therefore, the flow in the LCA is antegrade with relatively saturated blood (6). At birth, the infant is asymptomatic but during the early months of life the pulmonary pressure falls below systemic pressure, which results in left-to-right shunt from the higher pressure left coronary arterial system to the lower pressure pulmonary arterial system manifested in relation to the effort of crying or eating, causing denial of food (as in this case). If the collateral circulation does not develop,
Ischemic Cardiogenic Shock in the Infant by Anomalous Origin of the Left Coronary Artery

the anterolateral infarction appears with left ventricular heart failure, functional mitral insufficiency, ischemic cardiogenic shock and death (7).

New experimental data show that the normal development of the coronary system involves many growth factors, adhesion molecules and chemotactic factors coordinating cell migration and transformation of the coronary vessels. The presence of congenital anomalies of the coronary arteries suggests pathogenic abnormalities in cellular signals or alteration of these pathways with the involvement of local factors in the development of coronary vessels (4).

The case presented clinical symptoms starting at 1 month of age with clinical signs of congestive heart failure and myocardial infarction that will be confirmed by the ECG and increased myocardial enzymes.

ECG resembles the pattern described in lateral wall myocardial infarction in adult. The left ventricular leads also show abnormal Q waves, deep T-wave inversion and ST segment changes along with left ventricular and atrial overload.

Echocardiography confirmed dilated cardiomyopathy, severe mitral insufficiency, severe tricuspid insufficiency and doesn’t highlights the origin of LCA. The macroscopic pathologic exam confirms the LCA anomaly home base in PA, dilated cardiomyopathy, and fibroelastosis with secondary subendocardial necrosis.

Usually it is possible to identify LCA originating from PA, but in difficult cases a high parasternal position in parasagittal long axis view of main PA is often useful. Absence of aortic origin of LCA in a minimum of three views was proposed as a diagnostic criterion by some authors (8). But sometimes, spurious aortic origin (thus missing the diagnosis) can be ‘created’ due to presence of fluid filled transverse sinus of pericardium which can artificially mimic the origin of the left coronary artery from aorta (2). Color Doppler showing the flow in coronary artery towards the aorta (blue) is abnormal and confirms the diagnosis of ALCAPA. Yet again, this is not always present either because of angulation of the transducer, or due to the presence of pulmonary artery hypertension (1).

Most children with this rare anomaly die before one year of age, often after the age of 1 month with non treatable congestive heart failure. If collateral circulation develops clinical symptoms like chest pain on exercise, syncope or even sudden death in athletes may occur at older age (9).

In the presented case we suspected the malformation, but for accurate diagnosis we required cardiac catheterization and angiography or multislice computed tomography (10), which have not been available in our center, and the poor state of the patient did not allow his transfer to another medical center.

There are several surgical options available. These include: a) a bypass graft combined by ligation of the anomalous artery; b) the Takeuchi-procedure in which a tunnel is created between the aortopulmonary window and the coronary artery; or c) reimplantation of the anomalous artery into the aortic sinus (11, 12). Postoperative systolic function and heart failure significantly improved, the severity of mitral insufficiency diminishes and the ECG ischemic signs may disappear (13). Surgical intervention is the definitive treatment for ALCAPA and the severe state of the patient made it impossible in this case. Recent studies have shown that the estimated long-term survival at 20 years is 94.8% (14).

CONCLUSION

The aim of this case report is to increase awareness of this condition and to highlight that the need for early diagnosis of ALCAPA using echocardiography with color flow mapping is crucial in infants presenting with clinical symptoms suggestive of heart failure or ischemia.

CONFLICT OF INTEREST

Authors declares no conflicts of interest.

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