Case Study

Polyarteritis nodosa in infancy – generalized vascular problem diagnosed after initial cardiac manifestation in emergency cardiovascular catheterization.

ABSTRACT

Aim: Coronary artery diseases in children are uncommon, but in selected cases general vascular problems affect also the heart.
Case: We present the case of 4-month old girl admitted to Pediatric Intensive Care Unit after the incident of cardiac arrest and ventricular tachycardia, who underwent diagnostic catheterization due to cardiac ischemia. Coronary angiography showed changes in coronary arteries in the form of disseminated aneurysms, mixed with severely stenosed segments. General angiography discovered also changes in the number of peripheral arteries, with the most significant right subclavian and left iliac artery aneurysms.
Result: Upon angiographic images and a history of untreated infection, the PN was diagnosed, with an appropriate administration of intensive treatment.
Conclusion: The case prove the significance of invasive cardiovascular diagnostics in every unclear clinical course.

Keywords: cardiovascular imaging, emergency, interventional cardiologu

1. INTRODUCTION

An isolated coronary artery disease in children is uncommon, its underlying reason is usually congenital or infectious, without typical atherosclerotic injury. In selected cases generalized vascular non-cardiac problems in childhood affect also the heart. The diagnosis of general vascular disorder with cardiac manifestation is more difficult if the symptoms appear in a small child in life-threating emergency. Initial sequele of various forms of coronary insufficiency and severe heart dysfunction usually accompany different disorders that easily lead to cardiorespiratory insufficiency in small babies.

In dramatic circumstances routinely performed noninvasive emergency diagnostics remains unclear, especially in individuals free of previous cardiovascular problems and comorbidities. The differential diagnostics in children suffering from cardiovascular emergencies should consider anomalous origin of left coronary artery from pulmonary artery (ALCAPA), Kawasaki disease (KD), polyarteritis nodosa (PAN) and nonspecific organ vascular reactions [1]. An adequate diagnosis is important because of the need to prevent small babies from the most dangerous severe arrhythmias that can cause sudden cardiopulmonary arrest.

In unclear clinical course of severely ill babies nothing but an aggressive diagnostics path including coronary and peripheral angiography could provide an appropriate diagnosis, and become respective treatment on immediately. Here we present the case of an infant after the incident of cardiac arrest and ventricular tachycardia, who underwent an emergency diagnostic catheterization due to cardiac ischemia.

2. PRESENTATION OF THE CASE

We present the case of 4-month-old girl admitted to pediatric intensive care after the incident of cardiac arrest and ventricular tachycardia, who underwent diagnostic catheterization due to cardiac ischemia.

The indications for emergency angiography were established after meticulous analysis of risk-to-benefit ratio, and finally - despite the risk of invasive catheterization in decompensated child, the baby was referred to cardiac catheterization. Her initial transthoracic echocardiography (TTE) performed on admission showed significant akinesia of the apex, lateral wall of the left ventricle (LV) and interventricular septum, with dilated LV (Fig. 1D). There were no alarming patterns of coronary or pulmonary fistula, the heart muscle showed no signs of non-compaction. Coronary angiography showed changes in coronary arteries in the form of disseminated aneurysms, mixed with severely stenosed segments (Fig. 1A). Simultaneously performed general angiography discovered also the injuries in the number of peripheral arteries, with the most significant right subclavian and left iliac artery aneurysms. The aorta showed no angiographic thickening or stenosis, while there were findings of vasculitis-related changes in aortic arch branches, significantly present in cervical, subclavian and brain arteries (Fig. 1B). Thoracic descending and abdominal aorta were affected with visceral, renal and and the most evident left common iliac aneurysms (Fig. 1C). Finally upon her angiographic images and a history of untreated infection, the PAN was diagnosed, with an immediate administration of intensive treatment.

3. DISCUSSION:

Although polyarteritis nodosa still remains rare disease in childhood, the diagnosis of this disease should be kept in mind, especially in infancy. There is no single pattern of clinical presentation of PAN, but abdominal pain, central or peripheral nervous system disease, arthritis, myalgia and skin lesions occur during the course of the illness. The symptoms listed above remain unspecific, thus an advanced diagnostics is necessary to establish the correct diagnosis, and an appropriate treatment.

Polyarteritis nodosa in its angiographic view mainly affects the bifurcation areas of medium-sized muscular arteries and intravisceral small arteries of the testis, spermatic cord, nerves, muscles, skin, kidneys, gastrointestinal tract and joints [2]. Disseminated vascular disease cause severe clinical manifestations with life-threating conditions in groups of every age, including infants [1]. The clinical course of PAN with its initial critical cardiac and respiratory insufficiency is uncommon. Nevertheless in small babies, like in presented case, life-threating cardiopulmonary emergencies could be caused by various reasons, including PAN.

The diagnostics of PAN is based on central and peripheral cardiovascular catheterization. Following the 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides (CHCC) classifies vasculitides into three types primarily according to the predominant type of vessel involved: large vessel vasculitis, medium vessel vasculitis, and small vessel vasculitis [3]. Large vessels are defined as the aorta and its major branches. Medium vessels include major extravisceral arteries and their branches. Small vessels include intravisceral arteries, arterioles, capillaries, venules and veins [2].

We finally diagnosed three abovementioned CHCC vasculitis types in the presented infant girl suffering from PAN, nevertheless in differential diagnostics KD was considered as well. In our infant peripheral vessels affected in the pathology were relatively smaller in caliber

than those usually observed in KD, although we keep in mind that these two conditions can overlap [4, 5].

Following our patient's history we highly appreciate contemporary technical advances in emergency cardiovascular imaging, what enabled complete diagnosis of utmost importance for infant child admitted in her real critical condition. In summary we carefully considered well-defined risk of infant emergency cardiac catheterization procedures, although taking int consideration the potential to establish life-saving final diagnosis we became convinced to follow our strategy in forthcoming cases.

4. CONCLUSIONS:

The images obtained while emergency catheterization of an infant after cardiac arrest enabled an appropriate diagnosis and effective treatment with no wasted time, thus the case prove the significance of invasive cardiovascular diagnostics in every unclear clinical course.

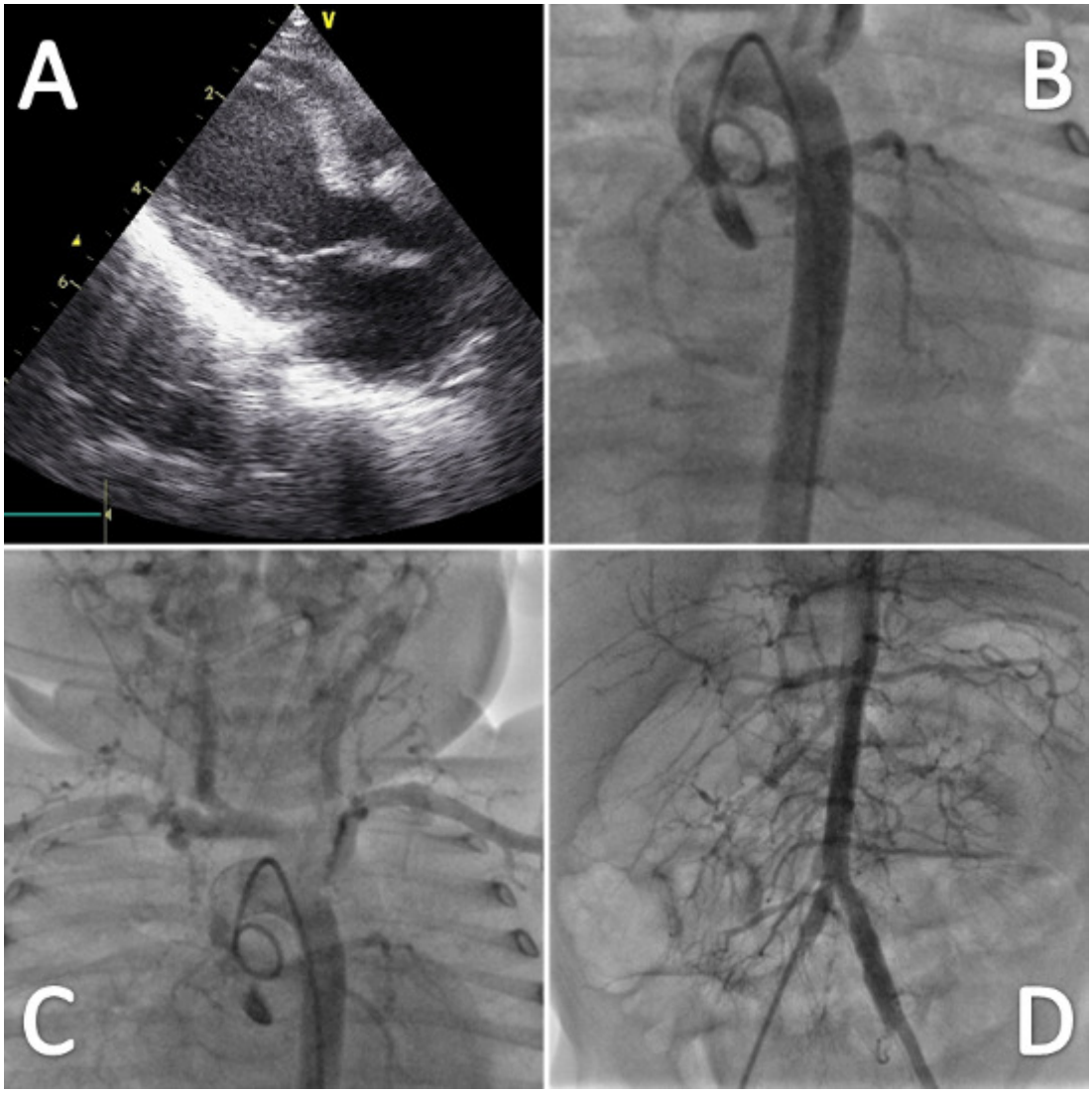

Figure 1:

1 A – Dilated, sphere shaped left ventricle.

1 B – Dilatation of left main and circumflex coronary arteries.

1 C - Right subclavian artery aneurysm.

1 D – Left common iliac artery aneurysm.

CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office.

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