

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 also affect** 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, polyarteritis nodosa (**PAN**) was diagnosed, with an appropriate administration of intensive treatment.

Conclusion: The case prove the significance of invasive cardiovascular diagnostics (cardiac catheterization) in every unclear clinical course.

Keywords: cardiovascular imaging, emergency, **interventional cardiology**

1. INTRODUCTION

An isolated coronary artery disease in children is uncommon and its essential reason is usually congenital or infectious. As well as there is lack of 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-threatening condition. Initial sequelae of various forms of coronary insufficiency and severe heart dysfunction usually accompany different disorders that easily lead to cardiorespiratory insufficiency in small babies.

In critical condition routinely performed noninvasive emergency diagnostics remains unclear, especially in patients without previous cardiovascular problems and comorbidities. The differential diagnostics in children suffering from coronary circulation 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.

34 2. PRESENTATION OF THE CASE

35 We present the case of 4-month-old girl admitted to pediatric intensive care after the incident
36 of cardiac arrest and ventricular tachycardia.

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

53

54 3. DISCUSSION:

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

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

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

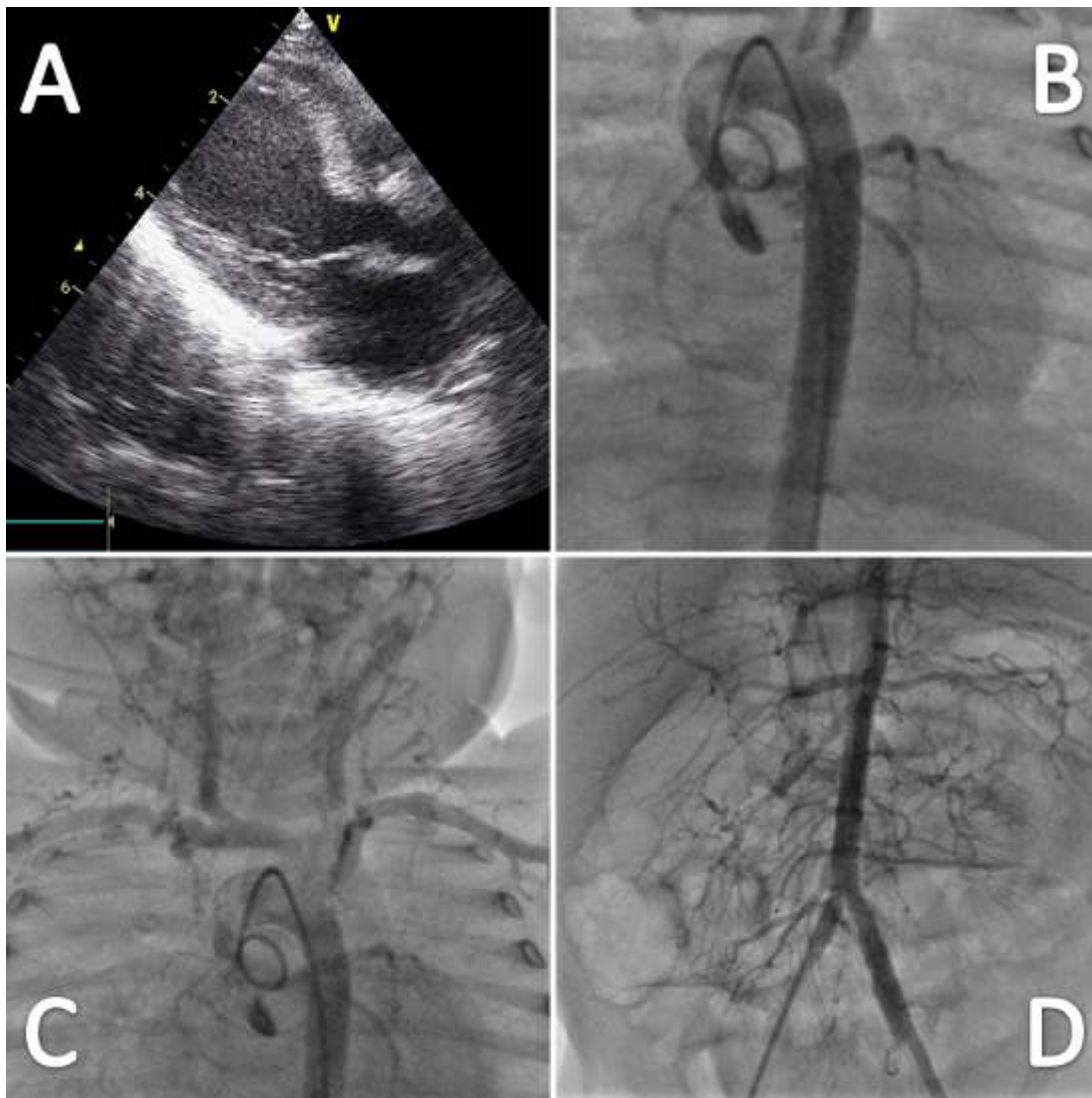
76 We finally diagnosed three abovementioned CHCC vasculitis types in the presented infant
77 girl suffering from PAN, nevertheless in differential diagnostics KD was considered as well.
78 In our infant peripheral vessels affected in the pathology were relatively smaller in caliber

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

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

87 4. CONCLUSIONS:

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



92 Figure 1:

93 1 A – Dilated, sphere shaped left ventricle.

94 1 B – Dilatation of left main and circumflex coronary arteries.

95 1 C - Right subclavian artery aneurysm.

96 1 D – Left common iliac artery aneurysm.

97

98 **CONSENT**

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

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