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Fulminant myocarditis with complete heart block associated with multisystem inflammatory syndrome in children: a case report

Piorunujące zapalenie mięśnia sercowego z blokiem całkowitym serca związane z wieloukładowym zespołem zapalnym u dzieci: opis przypadku

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Abstract

Multisystem inflammatory syndrome in children, or paediatric inflammatory multisystem syndrome, is a rare disorder characterised by a hyperinflammatory response to a previous severe acute respiratory syndrome coronavirus 2 infection. The course of the illness is usually relatively mild; nevertheless, severe cases have also been reported. We present a rare case of a 3-year-old girl with fulminant myocarditis with complete heart block associated with multisystem inflammatory syndrome in children. The condition was life-threatening and required intensive treatment of bradycardia with isoprenaline and the percutaneous implantation of an electrode for temporary pacing. Additionally, anti-inflammatory treatment with intravenous immunoglobulins and glucocorticosteroids was administered, leading to a rapid recovery with complete resolution of heart conduction abnormalities. In cases of fulminant myocarditis with complete heart block, prompt and intensive bradycardia treatment, combined with appropriate anti-inflammatory treatment, may be crucial for the patient's prognosis.

Keywords: atrioventricular block, myocarditis, multisystem inflammatory syndrome in children

Streszczenie

Wieloukładowy zespół zapalny u dzieci lub dziecięcy wieloukładowy zespół zapalny to rzadka jednostka chorobowa charakteryzująca się nadmierną odpowiedzią zapalną na uprzednią infekcję drugim koronawirusem ciężkiego ostrego zespołu oddechowego. Chociaż przebieg choroby jest zwykle relatywnie łagodny, to opisywano również ciężkie przypadki. W pracy przedstawiono rzadki przypadek 3-letniej dziewczynki z piorunującym zapaleniem mięśnia sercowego i całkowitym blokiem serca powiązanym z wieloukładowym zespołem zapalnym u dzieci. Choroba zagrażała życiu i wymagała intensywnego leczenia bradykardii izoprenaliną i przezskórnej implantacji elektrody do stymulacji czasowej. Dodatkowo zastosowano terapię przeciwzapalną dożylnymi immunoglobulinami i glikokortykosteroidami. Doprowadziło to do szybkiego zdrowienia z całkowitym ustąpieniem zaburzeń przewodnictwa przedsionkowo-komorowego. W przypadku piorunującego zapalenia mięśnia sercowego z całkowitym blokiem serca natychmiastowe i intensywne leczenie bradykardii w połączeniu z odpowiednią terapią przeciwzapalną mogą mieć fundamentalne znaczenie dla rokowania.

Słowa kluczowe: blok przedsionkowo-komorowy, zapalenie mięśnia sercowego, wieloukładowy zespół zapalny u dzieci

INTRODUCTION

Multisystem inflammatory syndrome in children (MIS-C), or paediatric inflammatory multisystem syndrome (PIMS), is a rare disorder characterised by a hyperinflammatory response to a previous severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection[1]. The cardiovascular system is frequently involved, and patients may present with shock. However, most patients recover completely and the mortality rate is low, approximately 2%[1,2]. We report a case of 3-year-old girl with an uncommon, severe course of the illness, presenting with myocarditis and complete heart block, which required prompt and intensive medical treatment.

CASE REPORT

A 3-year-old girl was transferred to the Paediatric Cardiology Clinic from the Regional Hospital Emergency Department due to symptomatic bradycardia. The child was previously healthy, with no significant past medical history. The parents reported that two weeks before hospitalisation, the girl had experienced laryngitis, which had been treated with intramuscular and nebulised glucocorticosteroids. In addition, for three days prior to admission, she had a low-grade or mild fever. From the morning of admission, the child was week and apathetic, with nausea and vomiting, prompting her parents to call an ambulance.

On admission to the Paediatric Cardiology Clinic the patient was conscious, weak, with pale skin, visible dyspnoea, an irregular heart rate of around 40 beats per minute, a weak peripheral pulse, cold distal parts of limbs, and prolonged capillary refill. The electrocardiogram (ECG) showed complete atrioventricular (AV) block with an atrial rhythm of 150 beats per minute and an irregular ventricular rate of 35–50 beats per minute (Fig. 1).

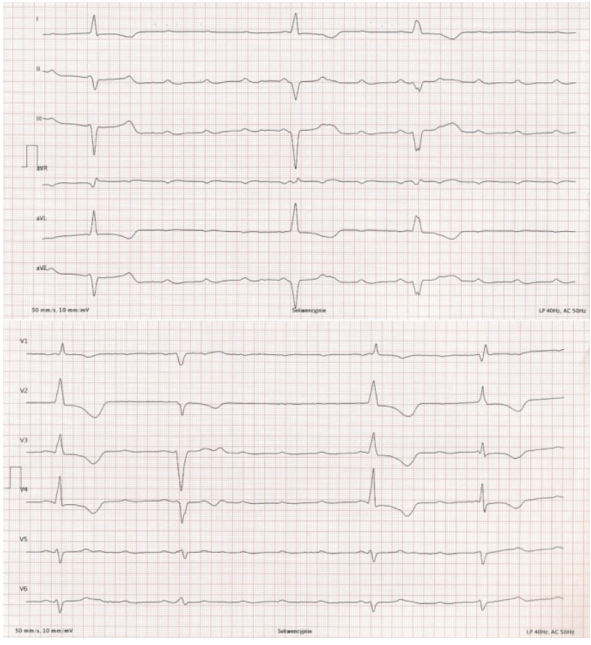

Fig. 1. ECG on admission. Complete atrioventricular block with irregular ventricular escape rhythm – polymorphic ventricular contractions, rate ~35–55/min

Laboratory tests showed metabolic acidosis with elevated lactate levels, as well as increased inflammatory markers and cardiac troponin T (Tab. 1).

Isoprenaline infusion and intravenous fluids were used for the initial treatment. With isoprenaline, the heart rhythm evolved to sinus with first-degree AV block at a rate of 95 beats per minute and repolarisation abnormalities (Fig. 2). The patient's condition started to improve. Initial echocardiography showed no structural cardiac defects, a left ventricular ejection fraction (LVEF) of 57–62%, and hypokinesis of the basal septal segments. No significant changes in coronary arteries were observed, except for a slightly hyperechogenic right coronary artery wall.

After a few hours of stabilisation, the heart block progressed. The patient, who was still on isoprenaline, experienced several episodes of second- and third-degree AV block and a few pauses lasting around 10 seconds. After the isoprenaline dose was increased, these episodes were intermitted by episodes of tachycardia with a wide QRS complex. The decision was made to implant an electrode for temporary pacing. During the induction of general anaesthesia, the patient had an episode of asystole with successful resuscitation. The electrode was introduced through the right jugular vein and positioned in the right ventricle.

Laboratory parameter	Reference value	On admission	At discharge
WBC [10^3/µL]	5.5–15.5	17.3	8.6
RBC [10^6/µL]	3.7–5.7	4.16	3.94
HGB [g/dL]	10.7–14.7	12	11.3
HCT [%]	31–43	35	31.8
PLT [10^3/µL]	140–553	148	424
LYM [10^3/µL]	1.8–2.9	4	7
NEUT [10^3/µL]	1.7–7	12.7	3.42
LYM %	40–60	23.4	60.7
NEUT %	25–55	72.8	29.7
CRP [mg/L]	0–10	39.8	0.5
PCT [ng/mL]	0–0.5	1.95	0.15
Ferritin [ng/mL]	4–67	3,080	–
ALT [U/L]	10–35	325	49
AST [U/L]	15–40	775	59
D-dimers [ng/mL]	0–500	33,571	571
pH	7.35–7.45	7.2	7.42
Lac [mmol/L]	0.5–1.6	15	2
Troponin T [µg/mL]	0–14	3,183	31
NT-proBNP [µg/mL]	0–125	1,232	40

ALT – alanine transaminase; **AST** – aspartate transaminase; **CRP** – C-reactive protein; **HCT** – haematocrit; **HGB** – haemoglobin; **Lac** – lactates; **LYM** – lymphocytes; **NEUT** – neutrophils; **NT-proBNP** – N-terminal prohormone of brain natriuretic peptide; **PCT** – procalcitonin; **PLT** – platelets; **RBC** – red blood cell count; **WBC** – white blood cell count.

Tab. 1. Laboratory parameters on admission and at discharge

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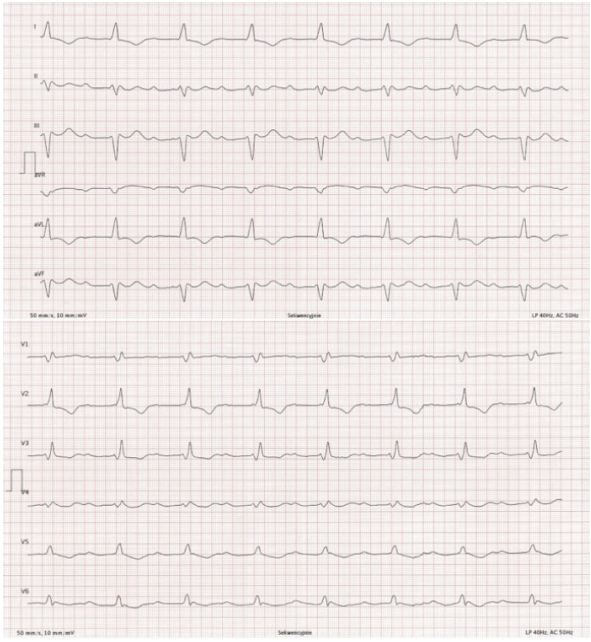

Fig. 2. ECG after initial treatment with isoprenaline infusion. Sinus rhythm ~95/min, first-degree AV block, repolarisation abnormalities

The stimulation threshold was set to 80 beats per minute in VVI mode. The child was then transferred to the Intensive Care Unit (ICU).

As the most probable cause of the block was fulminant myocarditis, the patient received intravenous immunoglobulins and glucocorticosteroids, leading to rapid and systematic improvement of the patient's state. During the first few days of hospitalisation, the girl had recurring episodes of fever. Since the second day, the ECG showed a sinus rhythm with no conduction disturbances, and the repolarisation abnormalities slowly normalised. After five days, the electrode was removed, and the patient was transferred back to the Paediatric Cardiology Clinic.

Cardiac magnetic resonance imaging (MRI) performed two weeks after the admission to the hospital showed mildly decreased left ventricular systolic function (LVEF: 47%), but no evident signs of inflammation. Follow-up laboratory tests showed normalisation of inflammatory parameters and troponin levels (Tab. 1). No direct infectious cause of myocarditis was found in laboratory assays (including antigen tests for flu, SARS-CoV-2 and RSV, antibody tests for Coxsackie A, Coxsackie B, CMV, EBV, DNA/RNA panels for parvovirus B19, adenovirus, HHV-6, Coxsackie). Lyme disease was also excluded, and blood cultures taken at admission were negative. However, the test for anti-SARS-CoV-2 immunoglobulin G was positive (level of 230.52 BAU/mL, positive >7.1 BAU/mL), and the child had not been vaccinated. This raised a suspicion that the myocarditis was related to MIS-C or PIMS. Due to lack of fever >38.5°C for at least three days, not all criteria necessary for a PIMS diagnosis were met. However, the case fulfilled the criteria for MIS-C according to the Centers for Disease Control and Prevention (CDC). Consequently, following the gradual withdrawal of glucocorticosteroids, the patient was prescribed aspirin, as recommended for MIS-C/PIMS.

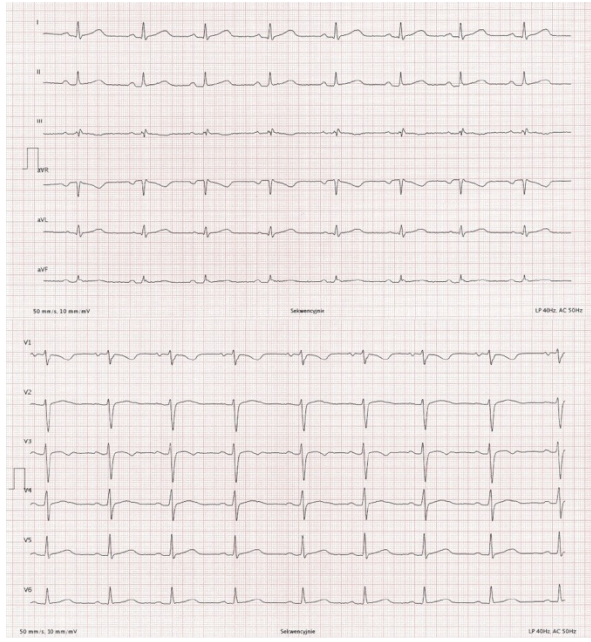

Fig. 3. ECG at discharge. Normal axis. Sinus rhythm ~100/min

After 20 days of hospitalisation, the girl was discharged in good clinical condition on oral aspirin and lisinopril, which has been initiated earlier due to slightly decreased LVEF. The ECG was normal (Fig. 3). On the follow-up evaluation after six weeks, the patient had a normal sinus rhythm in ECG, normal systolic function of the left ventricle, and no changes in the coronary arteries on echocardiography. As a result, both aspirin and lisinopril were discontinued.

DISCUSSION

Cardiovascular symptoms are among the most common presentations of MIS-C[1]. Decreased left ventricular ejection fraction, valvulitis, pericardial effusion, and coronary artery dilation are often present, and the majority of patients exhibit various ECG changes[1,3,4]. These symptoms may result from myocarditis which, in the context of MIS-C, is an outcome of a hyperinflammatory response to a prior SARS-CoV-2 infection with elevated levels of proinflammatory cytokines[1,5–7].

Electrophysiological changes most frequently reported in patients with MIS-C include low QRS amplitude, transient T-wave inversions, and other repolarisation abnormalities, while significant arrhythmia or conduction disorders are rarely observed[4,8]. As shown by a large observational study, the presence of AV block, especially high-degree AV block (second-degree Mobitz II AV block and complete AV block), is associated with longer hospital stays and higher mortality in patients with myocarditis. The odds ratio for mortality in patients with high-degree AV block compared to those with myocarditis assessed in the cited study was 1.58 (95% confidence interval, 95% CI: 1.03–2.49)[9].

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Studies from Boston and New York observed first-degree AV block in around 20% of patients with MIS-C[10,11]. Dionne et al. reported progression to second- or third-degree AV block in up to 80% of patients with first-degree AV block. These patients were admitted to the ICU; however, they did not require intervention for the block, which resolved within 1 to 6 days[11].

Severe cases of high-degree AV block in patients with MIS-C are rare. Rahmadhany et al. reported a case of a 2-year-old with complete heart block[12]. Similarly to our case, the child was initially treated with a temporary pacemaker; however, in that case, the block did not resolve and required implantation of a permanent dual-chamber pacemaker. One of the possible explanations for this difference is the anti-inflammatory treatment – our patient received both intravenous immunoglobulins and glucocorticosteroids, while the latter was treated only with glucocorticosteroids. This highlights the significance of timely and adequate anti-inflammatory treatment delivered alongside symptomatic management, whenever MIS-C is suspected.

LIMITATIONS

A limitation out of our report is that we cannot fully exclude causes of myocarditis other than MIS-C. Many of the most common viral infections were excluded. Nevertheless, the number of pathogens that could potentially cause myocarditis is vast, making it virtually impossible to exclude all of them. However, the combination of fever, cardiac involvement with cardiogenic shock, gastrointestinal symptoms prior to admission, elevated inflammatory parameters, and positive anti-SARS-CoV-2 antibodies, along with a positive and rapid response to immunoglobulin, supports the diagnosis of MIS-C. Another factor that might support MIS-C diagnosis over classic viral myocarditis is the fact that there were no fibrotic or inflammatory changes on the cardiac MRI performed after two weeks. It has been shown that MIS-C the inflammatory changes are transient, while in myocarditis such changes usually persist beyond the acute phase[13]. In addition, in comparison to classic viral myocarditis, patients with MIS-C myocarditis usually experience a rapid recovery of cardiac function and are less likely to have persistent ventricular disfunction, even when their state was critical at presentation[14]. Such rapid recovery was observed in the presented case.

Nevertheless, with clinical diagnoses like MIS-C, some level of uncertainty is always present.

CONCLUSIONS

The presented case demonstrates a rare and dangerous form of MIS-C with myocarditis and complete heart block. This situation requires prompt treatment of bradycardia, accompanied by adequate anti-inflammatory treatment, which may be crucial for the patient's prognosis.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organisations which might negatively affect the content of this publication and/or claim authorship rights to this publication.

Author contribution

Original concept of study; collection, recording and/or compilation of data: KK, LZ. Analysis and interpretation of data; writing of manuscript: KK. Critical review of manuscript; final approval of manuscript: JK.

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