Incidental diagnosis of Brugada syndrome in two girls hospitalized for pediatric inflammatory multisystem syndrome related to COVID-19 (PIMS-TS)

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Incidental diagnosis of Brugada syndrome in two girls hospitalized for pediatric inflammatory multisystem syndrome related to COVID-19 (PIMS-TS)

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Short title: Incidental diagnosis of Brugada Syndrome in children during PIMS

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SARS-CoV-2 related pediatric inflammatory multisystem syndrome (PIMS-TS) is a novel syndrome first described in England in May 2020 [1]. It affects children 2–6 weeks after COVID-19 infection whether symptomatic or not. The course of the disease varies from mild to requiring Intensive Care Unit support with a 2% mortality rate reported [2]. An inherent symptom of PIMS-TS is high fever. Cardiac evaluation is crucial in PIMS-TS patients as the involvement of the cardiovascular system determines the course of disease and prognosis.

We present 2 cases of girls aged 5 years (Patient 1) and 7 years (Patient 2). They were both admitted to a hospital due to high unremitting fever, weakness, and mucocutaneous and gastrointestinal symptoms. Patient 2, presented with severe dyspnea, and had a history of upper respiratory tract infection with a highly possible COVID-19 etiology 3 weeks before.

On admission, the girls presented with fever, polymorphic rash, and conjunctival redness. Patient 2 had hepatomegaly, pleural effusion, and cardiac enlargement in chest X-ray. Laboratory test results in both patients were typical for PIMS-TS with significantly elevated inflammatory parameters, cardiac biomarkers, lymphopenia, hyponatremia and increased D-dimers. Following PIMS-TS diagnostic and treatment protocol, electrocardiography (ECG) and
echocardiography were performed [3]. ECG revealed abnormal repolarization with coved ST-segment elevation $\geq$2 mm, followed by negative T wave in V1 and V2 precordial leads. Such abnormalities are not characteristic of PIMS-TS.

The patients were treated for PIMS-TS in accordance with Polish Pediatric Society recommendations [3]. Immunomodulating therapy (immunoglobulins and glucocorticosteroids) and acetylsalicylic acid were administered. Patient 2 developed symptoms of cardiogenic shock with severely reduced left ventricle ejection fraction and required inotropic medication. They both recovered without complications. Their ECGs have normalized once the fever resolved.

With the aforementioned ECG recordings both patients met the criteria of the Brugada type 1 ECG pattern [4]. Brugada syndrome phenocopies were excluded [5]. Patients and their first-degree relatives underwent a thorough cardiological screening according to the Heart Rhythm Society/European Heart Rhythm Association/Asia Pacific Heart Rhythm Society (HRS/EHRA/APHRS) expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes [4].

Patient’s 1 family’s cascade screening led to the patient’s father being subjected to an ajmaline challenge, which revealed a typical Brugada syndrome pattern. Moreover, the index case paternal grandfather suffered a sudden cardiac death. The girl’s genetic test with Next Generations Sequencing (NGS) testing revealed a variant of unknown significance in SCN5A gene. No other family members share the disease. No significant events were reported in the patient’s 2 family. They are currently being investigated to exclude the disease in first-degree relatives of the patient. NGS testing is in progress. Both patients were then given lifestyle changes recommendations, according to HRS/EHRA/APHRS expert consensus statement [4], however both families were reassured about low risk of life threatening arrhythmias in asymptomatic and incidentally diagnosed patients.

The two cases presented a typical but severe course of PIMS-TS. High and long-lasting fever occurring in PIMS-TS exposed abnormal electrocardiographic repolarization pattern which led to diagnosis of Brugada Syndrome. In children, ECG in fever is rarely performed, however, as stated, it may lead to a correct diagnosis.

REFERENCES


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**Figure 1.**

A. Patient 1 with fever. ECG with Brugada pattern (black arrow) seen in precordial leads.

B. Patient 1 with fever. ECG showing no abnormalities in limb leads.

C. Patient 1. Normal ECG recording after defervescence, precordial leads.

D. Patient 2 with fever. ECG demonstrating Brugada pattern (black arrow) in precordial leads.

E. Patient 2 with fever. ECG with abnormal repolarization, typical for PIMS-TS, seen in limb leads.

F. Patient 2. Normal ECG recording after defervescence, precordial leads.

Abbreviations: ECG, electrocardiogram; PIMS-TS, pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2

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