CASE REPORT

Brugada syndrome in a 4-year-old child with Lemierre syndrome—A case report

Sami Alanazi [b], Hussain Moafa [c], Faleh Al-Qahtani [c], Tahir Hameed [a,b,*]

[a] College of Medicine, King Saud bin Abdulaziz University for Health Sciences, Riyadh
[b] Department of Pediatrics, King Abdullah Specialized Children's Hospital, King Abdulaziz Medical City, Ministry of National Guard – Health Affairs, Riyadh
[c] Division of Pediatric Cardiology, King Abdulaziz Cardiac Centre, King Abdulaziz Medical City, Ministry of National Guard – Health Affairs, Riyadh

[a,b,c] Saudi Arabia

Brugada syndrome is a rare arrhythmogenic disease with characteristic electrocardiogram (ECG) findings. Fever represents an important triggering factor. We report the case of a 4-year-old Saudi boy who was diagnosed with Lemierre syndrome and subsequently developed Brugada syndrome.

Keywords: Brugada syndrome, Channelopathy, Lemierre syndrome

Introduction

Brugada syndrome is an inherited malignant arrhythmogenic disease characterized by electrocardiogram (ECG) patterns as well as arrhythmias that could present with syncope or cardiac arrest [1]. The ECG patterns suggestive of this disease are not always confirmatory, and include coved-type ST-segment elevation $\geq$2 mm in the right precordial leads and T-inversion at a minimum of two precordial leads. This pattern is termed "Type I" Brugada ECG pattern and is one of the requirements to diagnose Brugada syndrome [2]. Other electrocardiographic patterns, such as coved ST-segment elevation of 1 mm, or saddle-shaped ST-segment elevations have also been reported [3]. While fever-induced Brugada syndrome is a recognized clinical entity [4], there have been few pediatric case reports in the literature of fever-induced or other triggers of Brugada syndrome.

Case report

A 4-year-old-boy presented to the emergency room because of a 3-day history of fever, abdominal pain, and neck pain. Two days prior to

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* Corresponding author at: Department of Pediatrics – Mail Code 1940, King Abdullah Specialized Children's Hospital, King Abdulaziz Medical City, King Saud bin Abdulaziz University for Health Sciences, Ministry of National Guard – Health Affairs, Post Office Box 22490, Riyadh 11426, Saudi Arabia.
E-mail address: hameedta@ngha.med.sa (T. Hameed).

P.O. Box 2925 Riyadh – 11461KSA
Tel: +966 1 2520088 ext 40151
Fax: +966 1 2520718
Email: sha@sha.org.sa
URL: www.sha.org.sa

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symptom onset there was a history of trauma to the back of his skull. In the emergency room he was found to be grunting with poor perfusion and severe neck pain on examination. Vital signs were temperature 39 °C, heart rate 200 per minute, blood pressure 111/79 mmHg and respiratory rate 40 per minute. An intravenous line was inserted, two boluses of intravenous fluid were administered, a full septic work-up was done, and broad spectrum antibiotics were started. The child was admitted to the pediatric intensive care unit.

In the pediatric intensive care unit, the patient was noted to have progressive left neck pain with neck swelling. A lateral neck radiograph was taken to rule out a retropharyngeal abscess and it showed slightly thickened prevertebral soft tissue. A computed tomography scan of the neck with contrast revealed minimal fluid collection in the retropharyngeal space with no definite abscess. However, a left jugular vein filling defect was seen suggestive of thrombosis. Neck Doppler ultrasound showed partial thrombosis of the left internal jugular vein. Brain venogram computed tomography also confirmed this thrombosis. Blood culture became positive for Methicillin-sensitive *Staphylococcus aureus* (MSSA), and the repeated culture came back with same pathogen. Other cultures including cerebrospinal fluid culture were negative. With the persistent growth of MSSA, echocardiography was done to exclude infective endocarditis (IE); no vegetation or signs of IE were found. With the findings of jugular vein thrombosis and MSSA bacteremia/sepsis, a diagnosis of Lemierre syndrome was made.

Over the course of the patient's hospital stay, he developed a new left-sided neck swelling and imaging studies confirmed left vertebral artery aneurysm. He subsequently underwent coiling of the aneurysm by neuro-interventional radiology and insertion of a peripherally inserted central catheter (PICC) line. Following PICC line insertion, the patient had episodes of bradycardia. An ECG was done and the findings were consistent with Type I Brugada syndrome pattern (elevated J point, coved ST segment, and inverted T wave in precordial leads; Fig. 1).

Follow up ECGs were repeated many times after the patient was transferred to the general pediatric ward; the abnormal ECG findings completely disappeared as shown in Fig. 2. There was no family history of cardiac diseases or sudden death, and genetic studies for Brugada syndrome (including SCN5A gene) were negative. After discharge from the hospital, ECGs were repeated and they were normal with no findings of Brugada pattern. The child had follow-up with pediatric cardiology and the family was counseled regarding the nature of Brugada syndrome and precautions to take (e.g., to treat any fever immediately, precautions during swimming, and to avoid over the counter medications).

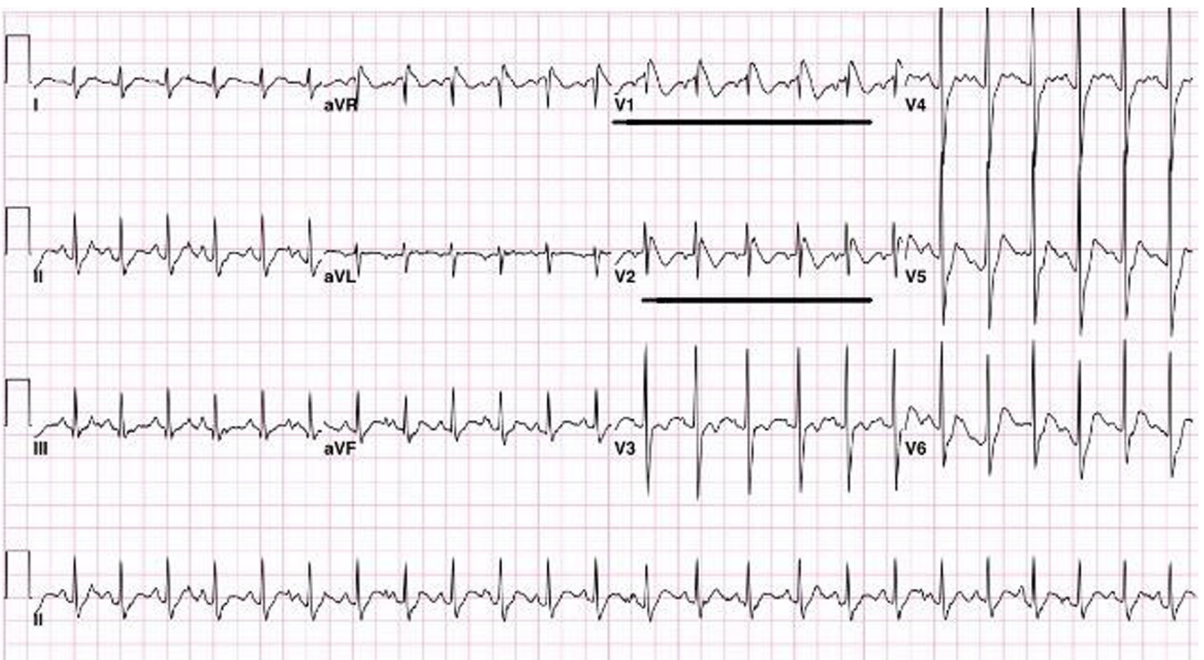

Figure 1. The electrocardiogram findings characteristic of Brugada syndrome.

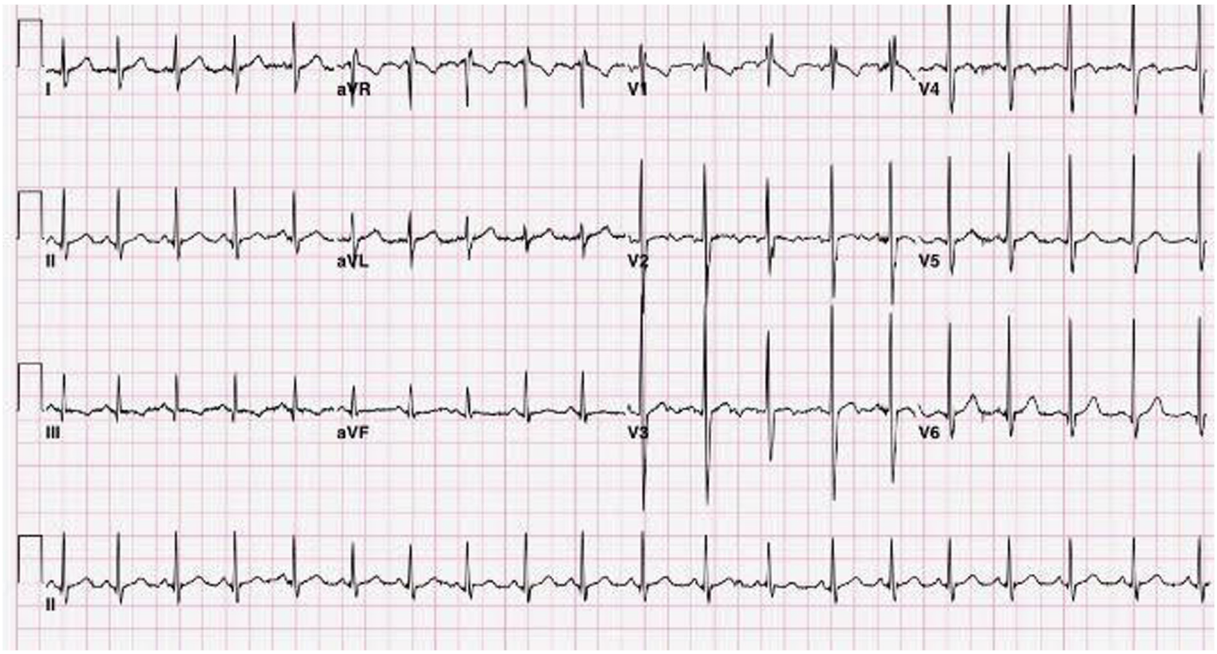

Figure 2. Complete disappearance of the Brugada pattern.

Discussion

To the best of our knowledge, this is the first case report of Brugada syndrome in a Saudi child. Although other pediatric case reports in the literature have described Brugada syndrome unmasked by simple febrile illnesses, our patient was diagnosed with another rare disease—Lemierre syndrome. Lemierre syndrome is an infection induced suppurative thrombophlebitis of the internal jugular vein and is mainly caused by oropharyngeal infections such as tonsillitis, retropharyngeal abscess, and teeth infections [5]. The main causative pathogen is the *Fusobacterium* species, but other organisms could also induce this disease [6].

Fever-induced Brugada is a well-accepted phenomenon as the fever unmasks the Type I Brugada pattern in carriers of this disease[7]. Other researchers have described it as a disturbance of cardiac sodium channels during the fever [8]. As fever is an important trigger of life-threatening arrhythmias in children with cardiac channelopathies, it appears reasonable to perform an ECG on all febrile children with a family history of sudden death [9,10].

Our patient did not have a documented fever at the time of appearance of the Type I Brugada pattern. However, the ECG pattern did occur after insertion of a PICC line. We reviewed the literature and found no case reports of Brugada syndrome unmasked after insertion of central venous catheters. We highlight this case to raise the awareness of Brugada syndrome and to report two rare syndromes that developed in the same child.

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