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Case Report Open Access

Lown-Ganong-Levine (LGL) Syndrome in a 4 Years Old Girl with Abnormal Tricuspid Valve, Tricuspid Valve Regurgitation and Right Sided Heart Dilatation

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Abstract

LGL syndrome is a rare pre-excitation syndrome proposed to be due to abnormalities related to the signal conduction at the atrioventricular level with only few case reports describing it in a structurally abnormal heart, mainly left side lesions. In this case report this syndrome is reported in association with right side abnormalities which is unique. We conclude that structural cardiac problems should always be considered during evaluation of patients with LGL syndrome.

Keywords: Lown-Ganong-Levine (LGL) syndrome; Supraventricular tachycardia; Tricuspid valve

Introduction

LGL syndrome, includes a short PR interval, normal QRS complex, and paroxysmal tachycardia. The pathophysiology of this syndrome includes an accessory pathway connecting the atria and the atrioventricular (AV) node (James fiber), or between the atria and the His bundle (Brechenmacher fiber) [1]. A short A-V node refractory period and enhanced A-V conduction also has been suggested as a cause of this syndrome [2]. Other possible underlying mechanism is atrial myocarditis with possible causation of sudden death. [3] There are few case reports describing this syndrome in association with structural or functional heart disease [3-6]. Herein, we are reporting a case of LGL syndrome in a four years old girl with structurally and functionally abnormal tricuspid valve; to the best of our knowledge this is the first case in the literature describing such combination.

Case Report

A 4 year old girl was presented to the author's institute (Ibn AL-Atheer pediatrics hospital in Mosul) with sudden onset of breathlessness, chest discomfort and palpitation for the first time. On physical examination she was frightened and irritable; her pulse rate was around 250 beats/minute; her blood pressure was 110/60 mmHg and there was grade 2-3/6 systolic murmur heard over the left lower sternal border on precordial auscultation. An electrocardiogram was obtained and revealed an attack of supraventricular tachycardia (SVT) (Figure 1). After a few trials of intravenous adenosine and beta adrenergic blocking drug, propranolol, the rhythm converted to sinus one but with PR interval shorter than the normal for the child age according to Davignon table (<0.08 second) without delta wave which was consistent with LGL syndrome [7] (Figures 2 and 3). An echocardiography examination was performed to evaluate the cardiac structure and functions and surprisingly we found abnormal structure of tricuspid valve (TV) with a septal leaflet that was tethered directly to the interventricular septum (I.V.S) without any visible septal papillary muscle (Figure 4). The tricuspid valve was not closing perfectly during systolic contraction causing tricuspid valve regurgitation of moderate severity with peak pressure gradient between right ventricle and right atrium of 35 mmHg which in turn caused mild to moderate right side dilatation (Figure 5).

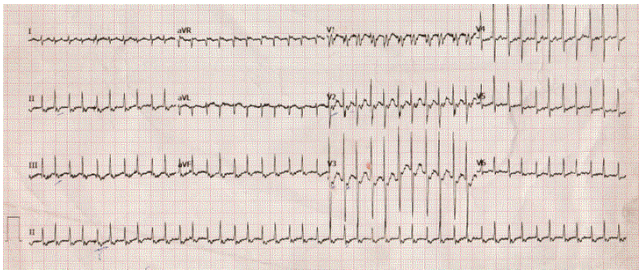

Figure 1: Electrocardiogram showing an attack of supraventricular tachycardia in a 4 year old girl.

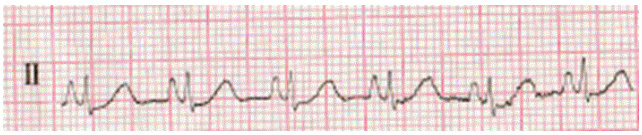

Figure 2: LGL syndrome with P-R interval = or < 0.08 second in lead II.

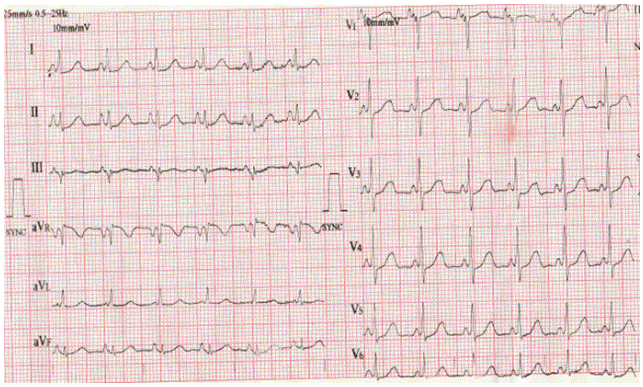

Figure 3: LGL syndrome after conversion to sinus rhythm by adenosine and propranolol.

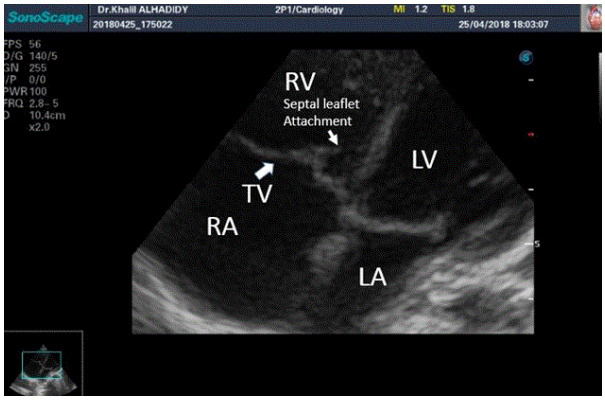

Figure 4: Echocardiographic picture showing short septal leaflet of tricuspid valve tethered to the I.V.S. RA (right atrium), RV (right ventricle), LA (left atrium), LV (left ventricle) and TV (tricuspid valve).

Discussion

The tricuspid valve is the right-sided atrioventricular valve, and it is composed of three leaflets (i.e., anterior, posterior, and septal) attached to a fibrous annulus. In spite of the documented variations in the papillary muscles of this valve; the septal leaflet is the least likely one to be affected by the abnormalities of these papillary muscles and their related chordae tendineae. Echocardiography remains the best method of evaluation of TV structure and function [8,9].

Tricuspid regurgitation is functional and is a satellite of left-sided heart disease and/or elevated pulmonary artery pressure most of the time; and when progressive, it worsens the patient prognosis whatever the underlying etiology [10].

The gross cardiac structure is generally normal in patients with LGL syndrome and the problem is basically related to the structure and /or function of the cardiac conductive system [1,2]; however atrial myocarditis, isolated non compaction of the left ventricle, mitral valve prolapse and Fabry's disease all have been reported in association with LGL syndrome without clear understanding of the role that these disorders may play in this syndrome [3-6].

The interesting point in our case is the finding of right side disease in association with LGL syndrome which has not been reported before; a point that should be considered in the future evaluation of LGL syndrome patients. Probably the main drawback in our case is the absence of electrophysiological study which, if were done, it could explain a new idea about the pathophysiology of this syndrome.

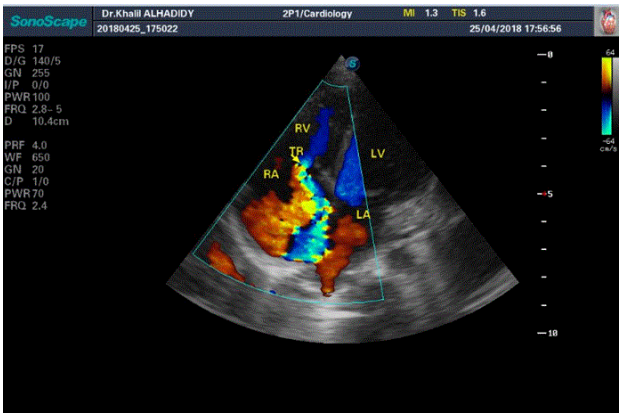

Figure 5: Moderate TV regurgitation causing moderate right atrial dilatation; RA (right atrium), RV (right ventricle), LA (left atrium), LV (left ventricle), TV (tricuspid valve) and TR (tricuspid regurgitation).

Conclusion

We conclude that the abnormalities of signal conduction described in association with LGL syndrome is probably liable to be affected by some cardiac structural disease and interestingly we are reporting right side heart disease in such problem.

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