Combination of Left Ventricular Noncompaction and Partial Atrioventricular Canal Defect in a 21-Year-Old Male: A Case Report

Malick Bodian¹, Modou Jobe¹, Mohamed Lèye², Mouhamadou Bamba Ndiaye¹, Adama Kane¹, Simon Antoine Sarr¹, Alassane Mbaye³, Maboury Diao¹, Fatimata Gatta Ba¹, Aliou Alassane Ngaïdé¹, Sarah Mouna Coly¹, Anna Thiam¹, Moustapha Sarr¹ and Serigne Abdou Bâ¹

¹Department of Cardiology, Aristide Le Dantec Teaching Hospital, Dakar, Senegal. ²Department of Cardiology, FANN Teaching Hospital, Dakar, Senegal. ³Department of Cardiology, Grand Yoff General Hospital, Dakar, Senegal. Corresponding author email: modoujobe@gmail.com

Abstract

Introduction: Left ventricular noncompaction (LVNC) is classified as a genetic cardiomyopathy characterized by a progressive systolic dysfunction. It may occur alone or in association with congenital cardiac anomalies. The combination of left ventricular noncompaction with partial atrioventricular canal defect is rare and has not, to our knowledge, been described previously.

Case presentation: A 21-year-old male who traveled to our center from a neighboring country presented with signs of heart failure. Transthoracic echocardiography showed prominent trabeculations in the left ventricle predominantly in the left ventricle involving the apical lateral and mid anterolateral segments associated with a partial atrioventricular canal defect. There was a biventricular systolic dysfunction. There was good response to medical treatment.

Conclusion: This case stresses the importance of maintaining a high degree of suspicion for this rare cardiomyopathy and the need to systematically look for other associated anomalies in order to institute proper short- and long-term managements.

Keywords: left ventricular, noncompaction, atrioventricular canal defect

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Introduction

Left ventricular noncompaction (LVNC) is a rare genetic cardiomyopathy characterized by a progressive left ventricular systolic dysfunction possibly due to arrest of compaction of myocardial fibers and meshwork during intrauterine life. It may occur alone or in association with other congenital cardiac or noncardiac anomalies. The average time from onset of symptoms to diagnosis of isolated left ventricular noncompaction according to a study by Ritter et al was 3.5 ± 5.7 years. In this study, the most common presentations for this cardiomyopathy were heart failure, ventricular arrhythmias, and thromboembolic events.

LVNC is commonly associated with other congenital anomalies; however, a combination with PAVD to our knowledge has not been previously described.

Case Presentation

A 21 year old young man with a childhood history of repeated sore throats and polyarthritis and with no known family history of cardiomyopathy presented at our center with dyspnea (NYHA functional class IV), bilateral leg edema, and dry nocturnal coughs. Clinical examination found a borderline systolic-diastolic hypertension (blood pressure = 140/90 mmHg), regular heart sounds with a 3–4/6 mesocardiac systolic murmur radiating to areas all of the precordium, signs of decompensated biventricular heart failure, a splenomegaly, and finger clubbing.

Laboratory analysis showed a slight normochromic normocytic anemia of 12.1 g/dL and a SaO₂ of 88%.

Electrocardiogram (ECG) inscribed a regular sinus rhythm with a heart rate of 94 cycles per minute, right QRS axis deviation at +130°, bi-atrial hypertrophy, a double heart block (first degree AV block and an incomplete right bundle branch block [RBBB]). There was also an inferior and a lateral subepicardial ischemia (Fig. 1). Chest X-ray showed a cardiomegaly with the apex above the diaphragm, a rectitude left middle heart border, bulged right lower heart border, and prominent hilar vessels (Fig. 2). Transthoracic echocardiography showed characteristics consistent with a hypokinetic dilated cardiomyopathy due to a left ventricular noncompaction characterized by the presence of prominent trabeculations predominantly in the left ventricle involving the apical lateral and mid anterolateral segments (Fig. 3) using the American Heart Association standardized segmentation of the left ventricle. There was a severe left ventricular systolic dysfunction (LVEF of 28% using biplane Simpson’s

Figure 1. ECG on admission demonstrating bi-atrial hypertrophy, first degree AV block and an incomplete RBBB.
an assessment of the clinical (symptoms and signs) evolution. The patient is at present seen on an outpatient basis to assess his International Normalized Ratio (INR), cardiac status (using clinical, electrocardiographic, and echocardiographic parameters), as well as to check on his intake of oral drugs.

Medical treatment involved both drug and nondrug therapies. The former involved salt and fluid restriction during the acute phase of treatment, and the patient was also encouraged to self-administer oral drugs. The drug therapy, on the other hand, involved intravenous furosemide (Lasilix) (which was changed to oral on day 7 of admission), oral spironolactone (Aldactone), and oral captopril. Acenocoumarol (Sintrom) and subcutaneous enoxaparine (lovenox) were given to prevent thromboembolic events. The latter was stopped after a good INR (of 2.14) was obtained with 2 mg of acenocoumarol.

The outcomes measured were favorable with the patient adhering to treatment given and with remarkable clinical improvement noted at the time of discharge and during follow-up care. Transthoracic echocardiographies were performed again on the day of discharge and at two months of follow-up care during which LVEFs of 32% and 48% respectively (using biplane Simpson’s method) were obtained. However, the patient has not yet undergone surgical intervention for the PA VD.

Discussion
LVNC is classified by the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases as an unclassified cardiomyopathy, while the American Heart Association classifies LVNC as a primary genetic cardiomyopathy. It is characterized

Figure 2. Chest X-ray of patient at admission.

Figure 3. Apical two chamber view shows zones of noncompaction and compaction by 2D echocardiography.

Figure 4. Apical four chamber view shows PA VD by 2D echocardiography.
by the presence of an extensive noncompacted myocardial layer lining the cavity of the left ventricle. Echocardiography is the most widely used imaging modality for the diagnosis of LVNC. However, its use is sometimes challenging. The apical region is poorly visualized by echocardiography and can lead to underestimation of the degree of the left ventricular non-compaction. Poor acoustic windows could mislead even experienced echocardiographers the diagnosis of this disease, resulting in an erroneous label such as dilated or hypertrophic cardiomyopathy. Cardiovascular magnetic resonance (CMR) has become the method of choice to confirm or rule out LVNC and may outperform echocardiography in defining the morphology and extension of myocardial noncompaction. This was demonstrated in a study by Yousef et al. CMR can also give valuable diagnostic and prognostic information about the disease by depicting fibrosis on delayed contrast-enhanced images.

Both isolated forms of LVNC or forms associated with another congenital anomaly (or other congenital anomalies), cardiac or noncardiac, have been described. Commonly associated anomalies include ventricular septal defect, coarctation of the aorta, transposition of the great vessels, and atrial septal defect. The precise pathophysiologic mechanism of the association of LVNC and other congenital heart diseases (including PAVD) has not been elucidated, and more studies are needed in this area.

Patients at the time of diagnosis may be asymptomatic or may present with complications that include heart failure, arrhythmias, and thromboembolic events. Rhythm disturbances include atrial fibrillation, Wolff-Parkinson-White syndrome (WPW syndrome), and ventricular tachycardia and, hence, the need for electrocardiograms to diagnose and monitor the occurrence of these potentially lethal conditions and hence the indispensability of electrocardiogram in both the short- and long-term management. Thromboembolic events that occur are due either to atrial fibrillation or to thrombus formation within the intratrabecular recesses in the noncompacted myocardium.

In this patient who presented with signs of biventricular heart failure and a relatively low arterial oxygen saturation, there was no arrhythmia; however, there were ECG features of a double block (a first degree AV block and an incomplete RBBB). Also in this case, we saw a very rare combination of LVNC associated with PAVD, of which we did not find a description in the literature.

Our long-term management plan involved monitoring his anticoagulation through periodic INR testing, close watch on the occurrence of arrhythmias by clinical and periodic ECG monitoring, subsequent evaluation for implantation of an implantable cardioverter-defibrillator, and exploration of eventual surgery of the PAVD.

Conclusion
The coexistence of LVNC and PAVD is rare. Echocardiography and, more recently, cardiac magnetic resonance are the main methods of diagnosis. Once diagnosis of LVNC is made, it is important to look for other congenital anomalies that might occur in association with LVNC in order to institute proper management strategies. The importance of other tests such as an ECG should not be overlooked, as this can be useful in identifying potentially life-threatening problems.

Consent
A written informed consent was obtained from the patient after careful consideration for publication of this case report and any accompanying images.

Author Contributions
MB and MJ conducted the literature search, drafted the first manuscript, performed language correction, and participated in article design and coordination. ML and MBN conducted the echocardiography and participated in manuscript draft. AK, SAS, and AM cared for the patient in the ward and during follow-up and contributed to drafting of manuscript. MD, FGB, and AAN critically revised the manuscript for important intellectual content. SMC and AT participated in investigation studies and critically evaluated the article. MS and SAB conceived the case study and participated in its design and coordination. All authors read and approved the final manuscript.

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