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CASE REPORT

Left Ventricular Noncompaction Cardiomyopathy and Myocardial Bridging Association: A Coincidence Or a Usual Association?

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Abstract

Left ventricular non compaction (LVNC) is a rare congenital disease. It occurs due to an arrest of the myocardial fibers compaction during embryogenesis. Myocardial bridge (MB) is a coronary anomaly in which the myocardium.

covers segments of the coronary arteries. We report a rare case of 62-year-old women who was diagnosed with the association of LVNC and MB revealed by chest pain and dyspnea. Some similar cases were reported in the last two decades suggesting that we may be in front of a usual yet underdiagnosed association. To our knowledge, this is the first case described in the Arab World.

Keywords: Left ventricular non compaction, Myocardial bridging, Heart failure, Congenital, Genetics

1. Case report

A 62 -year-old woman presented to our cardiology department for progressive dyspnea and chest pain evolving for fourteen years. Given the persistent symptoms, she was admitted for extensive cardiological evaluation in March 2021. No cardiovascular risk factors were found. Her past medical history was unremarkable, Physical examination found blood pressure at 130/80 mmHg, heart rate at 80 beats per minute oxygen saturation by pulse oximetry was 99% on room air. An electrocardiogram showed a sinus rhythm at 83 bpm with a complete left bundle branch block and QRS duration of 0.16 s.

An immediate echocardiography was performed revealing diffuse hypokinesis with left ventricular function impairment measured at 40% using the biplane Simpson method, Left-sided filling pressures were increased. Apical view and parasternal short axis view showed increased trabeculations below the level of papillary muscle of the left ventricle, and deep intertrabecular recesses in the endocardial apex and the left free wall (LV). The non compaction-to-compaction ratio was evaluated at 2.5 in the end diastole time (Fig. 1).

Ischemic heart disease was suspected first; A coronary angiography was carried out showing an intramyocardial bridge (MB) at the middle portion of the left anterior descending artery (LAD). The rest of the coronary artery network was normal (Fig. 2).

To confirm the diagnosis of LVNC, a magnetic resonance imaging was performed revealing a non-dilated left ventricle with global hypokinesia and an apical cryptic hypertrophy and trabecular aspect (Fig. 3).

The diagnosis of LVNC with MB has been made, Diuretics were administered to the patient: Furosemide 120 mg per day.

After the improvement of her clinical status under medical treatment. The patient was discharged on conventional heart failure therapy including: bisoprolol 5 mg per day, ramipril 5 mg per day, furosemide 80 mg per day, Dapagliflozin 10 mg per day,

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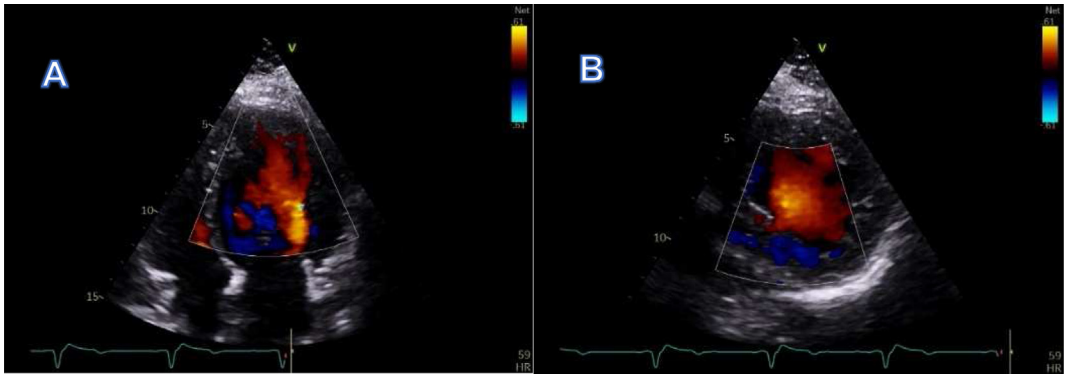

Fig. 1. Transthoracic echocardiogram in Apical view (A) and parasternal short axis view (B) demonstrating deep intertrabeculations recesses in the endocardial wall of left ventricular particularly in the apex free wall.

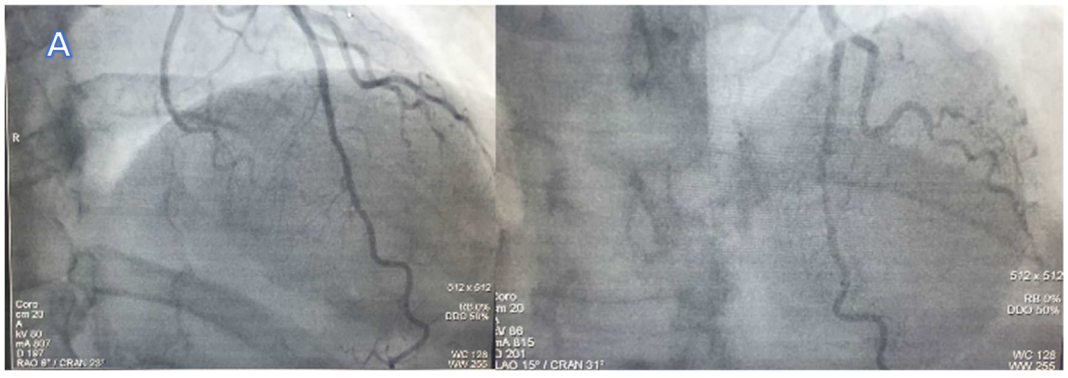

Fig. 2. Coronary angiogram in the cranial view showing normal left ascending coronary artery during diastole (A) and typical milking effect in the left ascending coronary artery during systole suggesting Myocardial bridging (MB) (B).

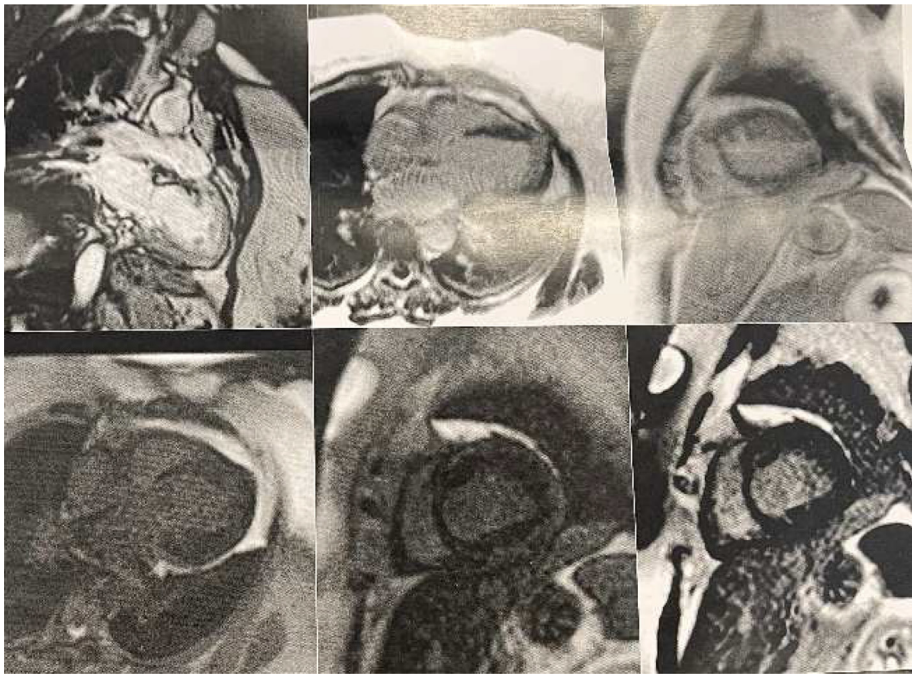

Fig. 3. Heart Magnetic resonance imaging (MRI) showing diffuse deep trabeculations in the left ventricle that communicate with the ventricular cavity.

and Rivaroxaban 20 mg per day, due to the important thrombo-embolic risk.

2. Discussion

LVNC is an unclassified cardiomyopathy with proven genetic-associated mutations. It is known to be familiar in 20–40% of cases. Many genes have been implicated and an autosomal dominant inheritance is mostly involved [1]. LVNC aspect result from an alternation of non-compacted and compacted muscle, which leads to deep trabeculations and, thus, a spongy-like aspect myocardium [2].

Myocardial bridging (MB) is a congenital coronary anomaly characterized by an intramyocardial course of an epicardial coronary artery, also described as a tunneled artery, and causing its compression and therefore myocardial ischemia [3].

Presently, the current approach for diagnosing non-compaction cardiomyopathy (NVM) using an echocardiograph involves two main sets of criteria: the Jenni criteria and the Chin criteria [4]. The former is used widely. It consists of identifying typically two layers (inner loose vs. outer tight $\geqq 2$) at end-systole, the inner layer should Communicate with the intertrabecular space, and coexisting cardiac abnormalities should be absent [5].

LVNC and MB are not benign conditions as each of them increases the risk of heart failure and cardiovascular events. It is already known that LVNC causes heart failure, arrhythmia, and embolic events [6,7].

Recently some patients had presented ischemic symptoms and ECG abnormality which led to confusion among the physicians since there is no clear explanation of these events.

The pathophysiology of LVNC is worth speaking of briefly here: Left ventricular noncompaction is normal in the first weeks of embryogenesis. Blood is supplied to the myocardium through the intertrabecular spaces. It is actually during weeks 5–8 of human life that gradual compaction of the spongy myocardium starts. This compaction progresses from the base to the apex of the heart. This interestingly matches with the coronary artery formation and therefore the time when MB may arise [8].

Therefore, LVNC is a morphogenetic defect that occurs at the site and time of embryologic development of the coronary arteries probably increasing the risk of coronary artery anomaly. Chiara Rovera reported a possible involvement of Ino80 chromatin remodeler mutation. He explained that It prevents ventricular compaction and that it results in a defect in coronary vessel formation which may cause LVNC and MB association [9]. Recently some published case reports from Turkey, China, and the USA argues for this hypothesis [9–11] this association is extremely rare, and only case report have been reported as stated before. To our knowledge, this is the first case described in Morocco and the Arab World.

3. Follow up

A close follow-up was scheduled after 1 week, 1 month, 3 months, and 6 months respectively. The patient remained free of symptoms and reported a significant improvement in her quality of life. LVEF remained the same as the initial echocardiography.

4. Conclusion

This is a case where LVNC and MB diagnosis was delayed due to a lack of knowledge of these diseases, this association may be usual and may not be a coincidence. Therefore physicians should screen for MB in LVNC with ischemic symptoms. This report opens the gates for more clinical and genetic testing to identify a possible association much more frequently than we think.

Learning objectives

This case report gives us many crucial clinical messages.

1 Ischemic symptoms in patients with LVNC should raise suspicion of coronary artery disease or MB and treatment (especially beta blockers and statin) should be started rapidly.
2 Always screen for the combination of LVNC and MB.
3 LVNC is often underdiagnosed always screen for it in the parasternal short–axis view in echocardiography using the color Doppler.

Consent statement

Written informed consent was obtained from the patient.

Author contribution

Conception, Data collection and/or processing, Visualization: AC; SO. Literature review: AC; SO; AMB; RH. Methodology: AC; RH. Software, Analysis and/or interpretation, Project administration: AC. Investigation, Resources: AC; SO; AMB Writer-original draft: AC; AMB. Writing- review & editing: AC; AMB; RH. Supervision: RH.

Conflicts of interest

The Author declares that there is no conflict of interest.

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