

Left ventricular noncompaction cardiomyopathy

Editor,

We appreciate the effort of (Finsterer *et al.*) for writing a letter to editor entitled, “Hyperthyroidism and noncompaction” on our published article “Left ventricular noncompaction diagnosed following Graves’ disease.” On behalf of all authors, we would hereby like to bring forward the following comments.

Isolated left ventricular noncompaction (LVNC) is a rare genetic cardiomyopathy. Clinical manifestations are variable; patients may present with heart failure symptoms, arrhythmias, and systemic thromboembolism. However, it can also be asymptomatic. When asymptomatic, LVNC can manifest later in life after the onset of another unrelated condition.

We do not agree with what the author’s mentioned about excluding LVNC as a cause of heart failure in our article. As we clearly mentioned that LVNC was diagnosed following the exacerbation in contractile dysfunction triggered by Graves’ disease,^[1] in patients with hyperthyroidism, heart failure may occur in the absence of underlying heart disease. Most patients with hyperthyroidism have a high cardiac output state without heart failure symptoms. However, reduced LV contractile reserve due to LVNC may impair the ability to increase the cardiac output to match the increase of peripheral demand. In our case, we think that Graves’ disease-induced hyperthyroidism led to a remarkable increase in preload and heart rate resulting in precipitation of heart failure.^[2] Literature review reveals that many patients with LVNC are minimally symptomatic or totally asymptomatic with many patients diagnosed with LVNC as an incidental finding on echocardiogram.^[3]

In addition, although we agree that autoimmune disease is a multisystem disease usually, many autoimmune diseases manifest as a single organ disease and we clearly

mentioned that the patient did not have any history or other manifestation of autoimmune disease besides Graves’ disease.

LVNC has familial occurrence rates of 18%–50% which have been reported in different case series. It is classified as an inherited genetic cardiomyopathy in 50% of the cases and can be sporadic in other cases.^[4] Therefore, our patient received family and genetic counseling, and she was advised that her first-degree family relatives need to be screened for LVNC.

However, there are reports on association of LVNC with other noncardiac abnormalities including neurological, facial, and hematological. In addition, dermatological and skeletal anomalies have also been described in isolated case reports.^[3-6] However, due to the wide range of noncardiac abnormalities associated with LVNC and the rarities of these conditions, there are no recommendations to screen for these associated abnormalities other than history and physical examination. Therefore, it is not appropriate or necessary to perform an extensive diagnostic workup in the absence of signs and symptoms of other organ involvement. Our patient did not have any neurological or musculoskeletal symptoms, and her neurological examination was normal. Hence, it was deemed unnecessarily to perform more diagnostic workup or have her evaluated by a neurologist.

LVNC is a rare cardiomyopathy that can rarely be asymptomatic and manifest later in life following other unrelated conditions that lead to exacerbation in contractile dysfunction such as Graves’ disease. Therefore, increased awareness of this rare but clinically significant cardiomyopathy is imperative for preventative and targeted therapy as well as for identification of potentially affected family members.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Amer Hawatmeh, Habib Habib¹, Fayez Shamoon

Department of Cardiology, Saint Michael's Medical Center, New York Medical College, ¹Department of Cardiology, Saint Joseph Regional Medical Center, New York Medical College, New Jersey, USA

Address for correspondence:

Dr. Amer Hawatmeh,
111 Central Avenue, Newark, New Jersey 07102, USA.
E-mail: amer_hawatmeh@hotmail.com

REFERENCES

1. Habib H, Hawatmeh A, Rampal U, Shamoon F. Left ventricular noncompaction diagnosed following Graves' disease. *Avicenna J Med* 2016;6:113-6.
2. Fazio S, Palmieri EA, Lombardi G, Biondi B. Effects of thyroid hormone on the cardiovascular system. *Recent Prog Horm Res* 2004;59:31-50.
3. Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: A distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol* 2000;36:493-500.
4. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, *et al.* Contemporary definitions and classification of

the cardiomyopathies: An American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation* 2006;113:1807-16.

5. Gati S, Chandra N, Bennett RL, Reed M, Kervio G, Panoulas VF, *et al.* Increased left ventricular trabeculation in highly trained athletes: Do we need more stringent criteria for the diagnosis of left ventricular non-compaction in athletes? *Heart* 2013;99:401-8.
6. Stöllberger C, Finsterer J, Blazek G. Left ventricular hypertrabeculation/noncompaction and association with additional cardiac abnormalities and neuromuscular disorders. *Am J Cardiol* 2002;90:899-902.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

Access this article online

Quick Response Code:



Website: www.avicennajmed.com

DOI: 10.4103/ajm.AJM_27_17

Cite this article as: Hawatmeh A, Habib H, Shamoon F. Left ventricular noncompaction cardiomyopathy. *Avicenna J Med* 2017;7:79-80.

© 2017 Avicenna Journal of Medicine | Published by Wolters Kluwer - Medknow