Editöre Mektup — Letter to the Editor

Late detection of noncompaction of the myocardium in an adult with complete interventricular septal defect

Diagnostic pitfalls in myocardial noncompaction

Dear Editor,

We have read the recent case report by Emini et al.[1] with great interest. The authors reported an interesting case with noncompaction of ventricular myocardium (NVM) in a 27 year-old-man followed-up with a "large ventricular septal defect" ("single ventricle"). Emini et al. discussed the pathophysiology of NVM and highlighted the importance of careful echocardiography in diagnosing NVM. In our opinion, some points of this work are not sufficiently clear.

The authors noted left ventricular trabeculations and deep intertrabecular recesses as the diagnostic criteria of NVM. However, in the presented case, they only showed the hypertrabeculations and deep recesses in the right ventricular portion of the single ventricle (especially in Figure 1b). Isolated left ventricular involvement in NVM is satisfactory for the diagnosis, although the sole right ventricular involvement requires further investigation as the right ventricle is structurally a trabecular ventricle. As in the reported case, accompanying elevated pressure (due to large interventricular septal defect) in the right ventricle leads to hypertrophy and elongation of trabeculas. Accordingly, definition of NVM requires the presence of hypertrabeculations and intertrabecular recesses in the left ventricle by clearly presenting the borders of the noncompacted and compacted layers.[2]

The authors reported pulmonary artery systolic pressure as 95 mmHg in the text, whereas they reported pulmonary artery systolic pressure as >100 mmHg in the figure legend of Figure 2. I am confused about the pulmonary artery systolic pressure of this case. Actually, pulmonary artery systolic pressure should be the same as the systolic systemic arterial pressure (100 mmHg) in the absence of aortic or pulmonary stenosis (which were not noted) as both chambers share common ventricular pressures.

Beyond the above-mentioned points, authors reported admission SaO_2 as 72% and they noted improved SaO_2 of 86% after the therapeutic interventions. Unresponsiveness of cyanosis (and SaO_2) is the clinical hallmark of cyanotic congenital heart diseases differentiating them from pulmonary diseases. Therefore, I wonder about the mechanism of SaO_2 improvement and the treatment modalities improving SaO_2.

We hope that the above-mentioned issues might add to the value of the case report by Emini et al.

Sincerely,

Yusuf Sezen, M.D., Ali Yıldız, M.D.
Harran Üniversitesi Tıp Fakültesi,
Kardiyoloji Anabilim Dalı,
63100 Şanlıurfa, Turkey.
Tel: +90 414 - 314 11 70 / 1143
e-mail: ghcayildiz@yahoo.com

REFERENCES

1. Emini M, Selmani H, Bajraktari G. Late detection of noncompaction of the myocardium in an adult with complete interventricular septal defect. Türk Kardiyol Dern Arş 2009;37:407-9.
2. Aras D, Tüfekçioğlu O, Ergun K, Özeke Ö, Yıldız A, Topaloğlu S, et al. Clinical features of isolated ventricular noncompaction in adults long-term clinical course, echocardiographic properties, and predictors of left ventricular failure. J Card Fail 2006;12:726-33.

Author's reply

Dear Editor,

We thank Mr. Yıldız, whose comments gave us the opportunity to discuss our case report published previously in your journal.

We agree with the author that the presence of intertrabecular recesses in the left ventricle is not very clear. It could be due to poor-quality images and also to difficulties in obtaining two-dimensional echo scans, because the patient was not clinically stable during the echocardiographic examination. However, we were convinced that the left ventricle was also non-compacted. We have sent the editor an echo

video which may further clarify the presence of non-compaction of the myocardium.

Regarding the improvement seen in SaO_2, we think that mild improvement in SaO_2 was associated with the bed-rest of the patient and oxygenotherapy; however, it did not reach the optimal level.

There was a misinterpretation of the systolic pulmonary artery pressure. We measured the tricuspid transvalvular pressure gradient as 94 mmHg, as shown in Fig. 2. We added the supposed pressure of the right atrium of 10 mmHg, and this yielded a value >100 mmHg. We agree that systolic pressure should be the same in the aorta and pulmonary artery in the absence of aortic valve disease and/or pulmonary valve disease.

Sincerely,

On behalf of the authors,

Gani Bajraktari, M.D.

University Clinical Center of Kosova,
Rrethi i Spitalit, P.n. 10000 Prishtina, Kosovo
Tel: ++377 44 355 666
e-mail: ganibajraktari@yahoo.co.uk

Myocardial noncompaction recognized following a transient ischemic attack

Thromboembolism in young patients with noncompaction cardiomyopathy: more than what we thought

Dear Editor,

I read with interest the case report by Karabulut et al.[1] about a transient ischemic event in a young man with noncompaction of the ventricular myocardium (NCVM). Although this complication was well documented by Oechslin et al.,[2] its occurrence in children and young adults is still probably underestimated. The presence of multiple trabeculations with deep intertrabecular recesses leads to blood stagnation and clot formation as was previously reported in three pediatric patients (Figure).[3] It is well documented that the left ventricle function may improve within days of presentation,[3,4] but this event may then provoke embolization, which was reported in children as young as 2 years of age. Serious or even fatal outcome may ensue when the left ventricle thrombus is large, and immediate anticoagulation or thrombolysis should be instituted.[5] Routine use of anticoagulation drugs in NCVM is controversial;[6] however, if there is poor ejection fraction, anti-platelet therapy is mandatory. Awareness about this important life-threatening complication of NCVM in young patients can help prevent/reduce mortality and morbidity.

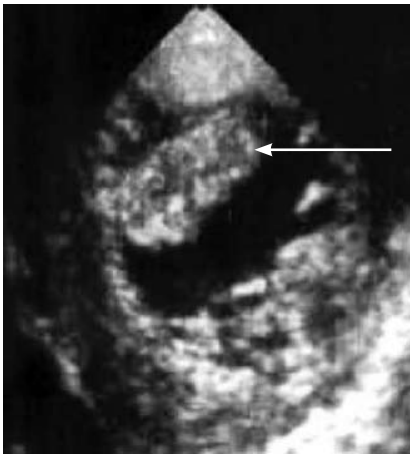

Figure. A short-axis view distal to the papillary muscles showing a thrombus (arrow) with noncompaction of the ventricle.

Sincerely,

Sulafa KM Ali, M.D.

Department of Paediatrics,
Faculty of Medicine,
University of Khartoum,
102 Khartoum, Sudan.
Tel: 00249 918075694
e-mail: sulafakhalid2000@yahoo.com

REFERENCES

1. Karabulut A, Erden I, Erden E, Cakmak M. Myocardial noncompaction recognized following a transient ischemic attack. [Article in Turkish] Türk Kardiyol Dern Arş 2009;37:205-8.
2. 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.
3. Ali SK. Unique features of non-compaction of the ventricular myocardium in Arab and African patients. Cardiovasc J Afr 2008;19:241-5.
4. Ali SK, Godman MJ. The variable clinical presentation of, and outcome for, noncompaction of the ventricular myocardium in infants and children, an under-diagnosed cardiomyopathy. Cardiol Young 2004;14:409-16.