Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

A 27-year-old patient with Left Ventricular Noncompaction (LVNC) and impaired systolic function after acute cardiopulmonary decompensation was admitted to our Center of Rare Cardiovascular Diseases for detailed evaluation. In the medical review he got mild limitation of exercise tolerance. 12–lead ECG showed sinus rhythm, 75 bpm, signs of both atria hypertrophy and left ventricle hypertrophy. The transthoracic echocardiography showed enlarged size of left ventricle, with impaired systolic function with moderate mitral and tricuspid regurgitation and increased trabeculations, spontaneous contrasting of blood with flow within lacunes. The MRI of the heart was planned which revealed impaired LV contractility, enlarged size of left ventricle. There were the ratio of NC/C layer >2,3 (Petersen et al. criteria) - Left Ventricular Noncompaction. Patient had also Holter-ECG which revealed significant ventricular arrhythmias.

DISCUSSION

Left ventricular noncompaction or "spongy myocardium", is a rare congenital cardiomyopathy that can be diagnosed at any age. It is characterised by a thin, compacted epicardial layer and an extensive non-compacted endocardial layer, with prominent trabeculation and deep recesses that communicate with the left ventricular cavity but not with the coronary circulation [1], probably due to an arrest of compaction during intrauterine life. Based on echocardiographic studies, reported prevalence is between 0.014 and 1.3% in the general population. Eventually, this condition can potentially lead to chronic heart failure, life threatening ventricular arrhythmias and systemic embolic events [2].

Although there is no consensus on diagnostic criteria, echocardiography is the main diagnostic tool. Cardiac magnetic resonance imaging has become the method of choice to confirm or rule out left ventricular noncompaction, because echocardiography cannot allow proper visualisation of the apex in some cases. The classical triad of complications - heart failure, ventricular arrhythmias and systemic embolic events - are common in patients with

advanced disease. Oechslin et al [3] reported that in a group of 34 adults with left ventricular noncompaction, the presence of higher final diastolic diameter of left ventricle, low ejection fraction, functional class III-IV (New York Heart Association), persistent or permanent atrial fibrillation and bundle branch block were related with high risk and poor prognosis, calling to consider the possibility of implantation of an automated cardiac defibrillator (ICD) and evaluation for transplant. Mortality was similar in left ventricular noncompaction as with patients with nonischemic dilated cardiomyopathy (3 year survival of 83 to 85 %) [4].

EXPERT'S OPINION

If there is obvious ventricular arrhythmias and impaired cardiac contractility (EF<35%), i.e. ICD implantation seems necessary.

CONCLUSION

In a patient with left ventricular noncompaction, the presence of higher final diastolic diameter of left ventricle, low ejection fraction, ventricular arrhythmias were related with high risk and poor prognosis, calling to consider the possibility of implantation of an automated cardiac defibrillator.

REFERENCES

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