CASE REPORT

Folia Cardiologica 2023
vol. 18, no. 3, pages 141–144
DOI: 10.5603/FC.2023.0014

ISSN 2353–7752
e-ISSN 2353–7760

Spongy cardiomyopathy with the first clinical manifestation at the age of 90 years

Aneta Kucharczyk-Foltyn, Dagmara Bijak

Department of Internal Disease, District Hospital, Chmielnik, Poland

Abstract

The present paper reports a case of a 90-year-old woman, admitted to the hospital because of decompensated heart failure *de novo*. Echocardiography examination revealed characteristic spongy cardiomyopathy: excessive trabeculation and occurrence of deep intertrabecular recesses. Standard treatment for heart failure was initiated, achieving clinical improvement. The case indicates, that spongy cardiomiopathy diagnosis can be made at a very advanced age and does not always mean a bad prognosis.

Key words: spongy cardiomyopathy, heart failure, echocardiography

Folia Cardiologica 2023; 18, 3: 141–144

Introduction

Spongiform cardiomyopathy also known as left ventricular noncompaction (LVNC) is considered a rare form of cardiomyopathy with a genetic basis [1]. The hallmark is abnormal left ventricular (LV) trabeculation with deep intertrabecular recesses communicating with the LV cavity. The lesions are mainly located in the LV apex, LV inferior and lateral wall, below the papillary muscles, less commonly affecting the interventricular septum, and may also involve the right ventricular (RV) muscle [2–4]. The clinical manifestation can range from completely asymptomatic cases diagnosed incidentally on echocardiography to severe heart failure (HF). The following triad of symptoms is typical of spongiform cardiomyopathy: HF, supraventricular and ventricular arrhythmias, including sudden cardiac death and thromboembolic events [4].

The diagnosis can be made based on transthoracic echocardiography. The most commonly used criteria are those of Jenni et al. [3]. These include the finding of two layers of myocardium: a thin epicardial compacted (C) layer and a much thicker endocardial noncompacted (NC) layer; NC to C ratio should be above 2:1 in the end-systolic phase.

Until recently, this form of cardiomyopathy was mainly diagnosed in children. It is estimated to represent 9% of all cardiomyopathies in young patients [4]. In contrast, over the past decade or so, there has been an increasing number of cases reported in adults, including elderly patients [5–7].

The prognosis of patients with this form of cardiomyopathy is poor, with a risk of sudden death exceeding 50% in a symptomatic patient population [8].

Case report

A 90-year-old female patient, treated psychiatrically for many years for personality disorders with signs of dementia and for hypertension, taking the following medications permanently: doxepin 3 × 25 mg, promethazine 2 × 25 mg, quetiapine 3 × 25 mg , perindopril 1 × 5 mg, amlodipine 1 × 5 mg, was admitted for the first time in her life to the

Address for correspondence: Aneta Kucharczyk-Foltyn MD, Oddział Chorób Wewnętrznych Szpitala Powiatowego w Chmielniku, ul. Bohaterów Warszawy 65D, 28–100 Busko-Zdrój, Poland, e-mail: kucharczykfoltynaneta@gmail.com

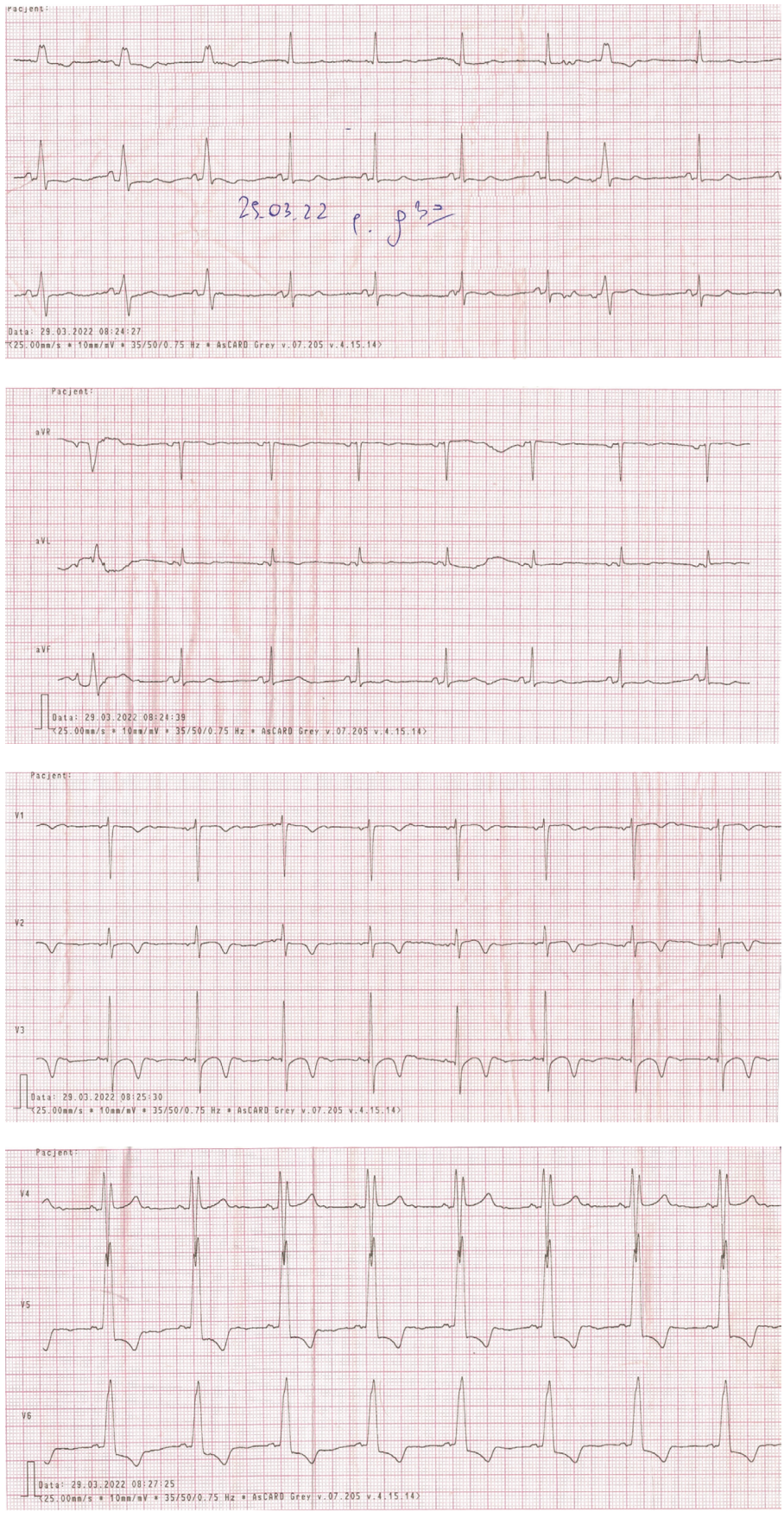

Figure 1. Electrocardiography recording during hospital admission. Intermittent left bundle branch block

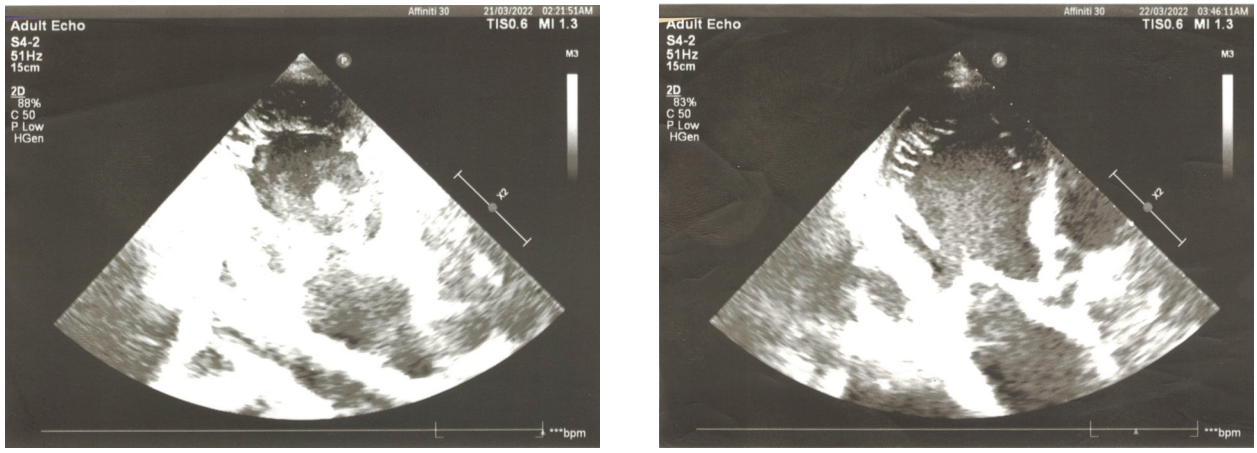

Figure 2. Transthoracic echocardiography. The presence of increased left ventricle trabeculation with deep recesses; noncompacted to compacted ratio greater than 2:1

Internal Medicine Ward in February 2022 due to dyspnoea at rest. Physical examination revealed BP 120/70 mm Hg, regular heart function, muffled heart sounds, muted vesicular breath sounds at the base of the lungs, slight periosteal oedema.

Laboratory tests showed the following abnormalities: moderate anaemia (Hb 10.6 g/dL), reduced creatinine clearance (Cockcroft-Gault GFR: 30 ml/min) and moderate hyponatraemia (Na 133 mmol/l).

The electrocardiography (ECG) recording (Figure 1) revealed intermediate heart axis, sinus rhythm 60/min, disturbed by single atrial premature beats with aberration, intermittent left bundle branch block (LBBB), signs of left atrial hypertrophy/enlargement, possible LV hypertrophy. Chest X-ray revealed the following abnormalities: opacity at the base of both lungs, possibly representing pleural fluid, with a greater amount on the right side, and fibrostreaky opacities in the lower fields of both lungs. Echocardiography was performed, which showed moderate enlargement of the left heart cavities and RV cavities, generalized LV hypokinesia with severely reduced ejection fraction (approximately 20–22%), high-grade LV diastolic dysfunction (restrictive mitral inflow pattern, E/E'sept = 22.5), impaired RV longitudinal systolic function (TAPSE 15 mm), low mitral, aortic and tricuspid regurgitation, high probability of pulmonary hypertension (RVSP: 51 mm Hg), low amount of pericardial fluid (up to 8 mm near the right atrium). In the region of the LV apex there was evidence of excessive trabeculation with deep recesses communicating with the LV cavity, passing into the middle segments of the lateral, anterior, and former posterior wall (Figure 2).

The psychiatric treatment, as recommended by the specialist, was modified: doxepin, which can promote the occurrence of hyponatraemia, was discontinued and the dose of quetiapine was increased. The following medications were added to the therapy: torasemide 1 × 10 mg, eplerenone 1 × 25 mg, bisoprolol 1 × 1.25 mg and a non-vitamin-K antagonist oral anticoagulant (eliquis 2 × 2.5 mg).

The treatment resulted in a significant clinical improvement — resolution of dyspnoea at rest, resolution of periosteal oedema. A follow-up ultrasound examination revealed regression of the amount of pericardial fluid and a reduction in the amount of pleural fluid. The patient refused to have a Holter monitor fitted but intermittent LBBB and single premature supraventricular beats were observed in the 12-lead ECG recordings performed and repeated several times during her stay on the ward.

The patient was discharged home with a recommendation for follow-up visits in the Cardiology Outpatient Clinic. The patient's family was informed of the genetic basis of the disease and immediate family members were advised to report to the Cardiology Outpatient Clinic.

Conclusions

The case presented in this paper, in which LVNC was detected at a very advanced age, proves that the disease does not always have to mean a poor prognosis. In the patient, the first clinical manifestation was the appearance of signs of HF. There was no history of embolic complications or loss of consciousness. The literature reports cases of LVNC patients in whom life-threatening arrhythmias or HF already appeared in childhood [4, 8]. There are also reports of good long-term prognosis in patients with this form of cardiomyopathy [9], which can be confirmed by the case presented in this paper. One year and 7 days have passed since the patient was discharged from hospital and we know that she is alive.

Conflict of interest

None declared.

References

1. Lubiszewska B, Hoffman P, Rużyłło W. Niescalony mięsień sercowy. Opis przypadku z przeglądem literatury. Kardiol Pol. 2001; 55: 447–449.
2. Włodarska EK, Woźniak O, Konka M, et al. Isolated ventricular concompaction mimicking arrhytmogenic right ventricular cardiomyopathy. Int J Cardiol. 2010; 145: 107–111.
3. Jenni R, Oechslin E, Schneider J, et al. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart. 2001; 86(6): 666–671, doi: 10.1136/heart.86.6.666, indexed in Pubmed: 11711464.
4. Paszkowska A, Kowalczyk-Domagała M, Turska-Kmieć A, et al. Obraz kliniczny kardiomiopatii pod postacią niescalenia mięśnia lewej komory u dzieci — doświadczenia własne. Postępy Nauk Medycznych. 2017; 30(9): 471–477, doi: 10.25121/pnm.2017.30.9.471.
5. Borys M, Snopek G, Sadowski K, et al. Kardiomiopatia gąbczasta — nie tylko u dzieci. Kardiol Pol. 2009; 67: 513–515.
6. Jamro M, Wiśniowski M, Curzytek A, et al. Niewydolność serca jako manifestacja izolowanego niescalonego mięśnia prawej i lewej komory. Folia Cardiologica. 2016; 11(4): 334–337, doi: 10.5603/fc.2016.0054.
7. Curzytek A, Filip M, Lubas W, et al. Dwa rzadkie przypadki izolowanego niescalonego mięśnia lewej komory zdiagnozowane u pacjentów w podeszłym wieku. Kardiol Pol. 2010; 68(6): 712–715.
8. Aleszewicz-Baranowska J, Kwiatkowska J, Potaż P, et al. Izolowany niescalony mięsień lewej komory : różne oblicza kliniczne. Kardiol Pol. 2009; 67: 1037–1039.
9. Murphy RT, Thaman R, Blanes JG, et al. Natural history and familiar chracteristics of isolated left ventricular non-compaction. Eur Heart J. 2005; 26: 187–192.