

Unclassified cardiomyopathy or Lyme carditis? A three year follow-up

Nieklasyfikowalna kardiomiopatia czy zapalenie mięśnia sercowego w przebiegu boreliozy? Trzyletnia obserwacja

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Abstract

Lyme carditis can be a clinical manifestation of the early disseminated stage of Lyme disease caused by the tick-transmitted pathogen *Borrelia burgdorferi*. We present the case of a 41 year-old Caucasian woman referred to our hospital with symptoms of fatigue, progressive exertional dyspnoea, supraventricular cardiac arrhythmia, and an enlarged heart revealed on chest radiography. Following an untypical result of transthoracic echocardiography, cardiac magnetic resonance was performed. This showed structural cardiac changes and focus of late gadolinium enhancement in the midwall of the apex region. Further diagnostic processes, including endomyocardial biopsy and serology tests, made it possible to diagnose Lyme carditis. Clinical observation was followed-up for three years.

Key words: magnetic resonance imaging, cardiomyopathy, myocarditis

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A 41 year-old woman with a four month history of progressive fatigue, exertional dyspnoea and atrial fibrillation (AF) was referred to our hospital. Physical examination showed an elevated blood pressure of 160/100 mm Hg, a normal heart rate and a correct body temperature. The patient's family history revealed the sudden death of her 22 year-old sister (cause unknown). Chest X-ray showed cardiomegaly with pulmonary congestion. Laboratory data presented elevated but stable cardiac enzymes: troponin I 0.31; 0.4; 0.36 (0.0–0.1 ng/L, BNP 2878 (0.0–125 pg/mL), leucocytosis 13,200/ μ L with normal value of hs-CRP 2.9 (0.0–3.0 mg/L).

The ECG demonstrated regular sinus rhythm of 90 bpm (spontaneous conversion from AF was observed) and incomplete left bundle branch block.

Echocardiography showed an enlargement and deformation of the left ventricle (LV), as well as not-homogenous

myocardial hypertrophy more intense in the apical region with thinning in the basal segment of the intraventricular septum. Mild systolic dysfunction with an ejection fraction of 52% was observed. Doppler echocardiography showed a restrictive pattern of mitral flow and increased filling pressure of the LV.

A 24-hour Holter ECG presented sinus rhythm with short episodes of AF, numerous supraventricular and ventricular contractions, and one episode of non-sustained monomorphic ventricular tachycardia (nsVT).

Due to an atypical morphology and heart structural changes, a cardiac magnetic resonance (CMR) was performed. It demonstrated midwall focus of late gadolinium enhancement in the apical region (Fig. 1).

Coronarography did not reveal significant coronary artery lesion. Simultaneously, an LV endomyocardial biopsy (EMB) was performed with three samples obtained from different

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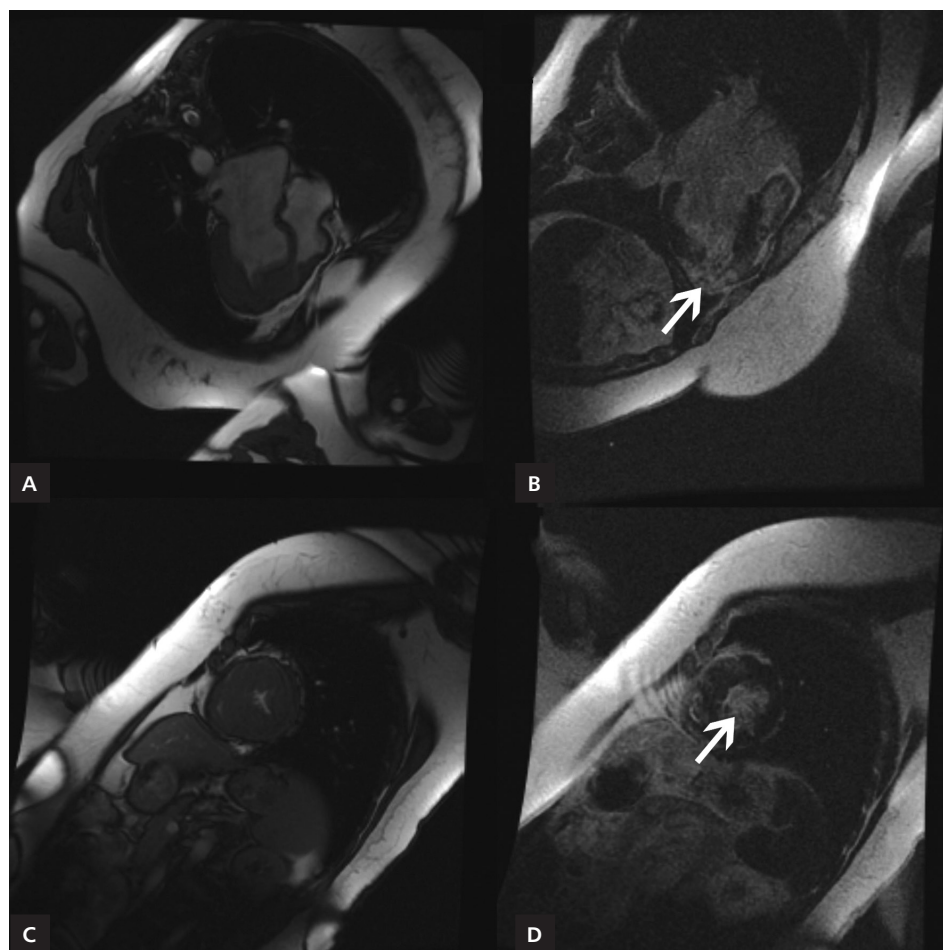


Figure 1. Cardiac magnetic resonance imaging, long (A, B) and short axis view (B, C). Steady-state free precession imaging showing structural changes in the left ventricle (A, C). Intramyocardial late gadolinium enhancement in the apical region (B, D)

regions. Light microscope examination and immunohistochemistry did not confirm a typical image of myocarditis, nor did it clearly indicate any other known disorder. Immunohistochemistry evaluation for the presence of CD34 antigen showed subendocardial cavities covered with endothelium of unclear aetiology (Fig. 2A).

Due to the occurrence of numerous abnormalities whose coincidence with clinical manifestation was still difficult to determine, other aetiological factors were taken into account. In one arm of the diagnostic process, the biopsy specimens were exposed to Warthin-Starry Silver Technique. A structure resembling a spirochaete was identified (Fig. 2B).

Serological tests were positive for antibodies of *Borrelia burgdorferi*. ELISA test showed elevated level of IgM antibodies – 13.6 μmL (negative < 9 μmL) and was negative for IgG antibodies. The confirmatory Western blot test identified IgM antibodies against antigens of *Borrelia burgdorferi*: 41 (+); VLse (+); OspA (++) . Other serological tests and clinical evaluation allowed us to exclude an infection or co-infection with *Treponema pallidum* and spirochaetes of the genus *Leptospira*.

Following the diagnosis, a four-week antibiotic therapy with ceftriaxon was introduced. Eight weeks after discontinuation of antibiotic treatment, an ELISA serological test and a Western blot test were performed and showed negative levels of serum antibodies IgM and IgG against antigens of *Borrelia burgdorferi*. A 12 month follow-up CMR showed features of past inflammation without any progression. Arrhythmias were still present, including AF and nsVT episodes with poor reaction to pharmacological treatment. The patient underwent implantation of an implantable cardioverter-defibrillator. Actually, we have followed her up for three years and she has been in good health without serious arrhythmic episodes.

Lyme boreliosis (LB) is the commonest tick-borne infection in Europe and North America. The whole of Poland is regarded as endemic to the prevalence of *Borrelia burgdorferi* with approximately 8,000-9,000 new cases reported annually. LB is a multisystemic infectious disease caused by several different *B. burgdorferi* genospecies. In about 5–10% of cases of LB untreated at an early stage, Lyme carditis (LC) can be recognised [1]. Typical and most frequent manifesta-

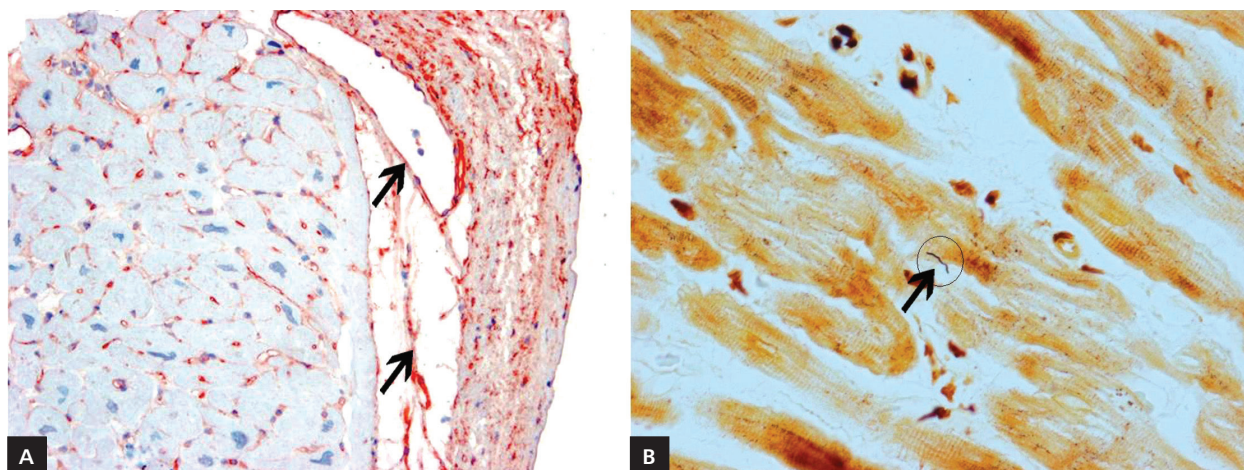


Figure 2. Light microscopy; **A.** Subendocardial cavities lined with endothelium cells (CD34 antigen expression). Magn. $\times 100$; **B.** Warthin-Starry silver impregnation. Spirochaete-like intramyocardial structure. Magn. $\times 400$

tion of cardiac involvement is transient atrioventricular block. Intraventricular blocks or supraventricular and ventricular arrhythmias occur much less frequently. There have been a few cases of dilated cardiomyopathy or acute coronary syndrome, which have been considered to be caused by *Borrelia* [1, 2]. The typical diagnostic process is based on tick bite history, occurrence of a characteristic rash known as erythema migrans, and serological tests. However, approximately 50% of LB cases are asymptomatic, and a large number of patients don't recollect a tick bite or erythema. Serodiagnostic tests can also be difficult due to weak antibody response or its lack at early stages of the disease and false negative results [3].

CMR and gadolinium enhancement is useful in diagnosing various cardiac diseases including myocarditis, fibrosis or post infarct remodelling. A coincidence of CMR with LC has been reported only in a few cases. Localisation of late gadolinium enhancement has been observed in the basal region of antero-septal region, which could correspond with the most frequent clinical manifestation of LC including atrioventricular block [4, 5].

This study presents the case of a young woman with LC causing diagnostic problems. Our patient did not report a tick bite or erythema, hence it was difficult to determine the duration of the disease. Clinical manifestation, including symptoms of heart failure and different cardiac arrhythmias without atrioventricular block, was not typical for LC but coherent with other rare cases of the disease reported in

literature. Localisation of late enhancement in CMR was different from those known from literature.

However, positive serodiagnostic tests as well as findings from EMB made it possible to diagnose LC and introduce proper antibiotic treatment. To the best of our knowledge, this is the first case showing a direct correlation between CMR image with presence of spirochaetes in myocardium. It is not clear whether other findings including subendocardial cavities and serious structural and functional heart changes observed in the presented case belong to the image of LC. The question concerning aggravating familial history also remains unanswered. Perhaps what we have seen here are two independent pathological processes: LC superimposed on earlier existing unclassified cardiomyopathy.

Conflict of interest: none declared

References

1. Steere AC. Lyme disease. *N Engl J Med*, 2001; 345: 115–125.
2. Nagi KS, Joshi R, Thakur RK. Cardiac manifestation of Lyme disease: a review. *Can J Cardiol*, 1996; 12: 503–506
3. Bratton RL, Whiteside JW, Hovan MJ, Engle RL, Edwards FD. Diagnosis and treatment of Lyme disease. *Mayo Clin Proc*, 2008; 83: 566–571.
4. Munk PS, Orn S, Larsen AI. Lyme carditis: persistent local delayed enhancement by cardiac magnetic resonance imaging. *Int J Cardiol*, 2007; 115: e108–e110.
5. Naik M, Kim D, O'Brien F et al. Lyme carditis. *Circulation*, 2008; 118: 1881–1884.