CHARACTERISTICS OF LYME CARDITIS

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SUMMARY

A short review of the characteristics and present knowledge of Lyme carditis is provided, referring to recent literature. First, the incidence and clinical features are discussed, then the possible diagnostic tools and finally the therapy, course and prognosis of Lyme carditis. Lyme carditis is nowadays a generally known disease with many possible different features, but with a favourable course in most cases.

KEY WORDS

Lyme carditis, features, diagnosis, therapy, course, prognosis

INTRODUCTION

Lyme carditis (= cardiac manifestations of Lyme borreliosis) was first characterized by Steere et al (1) in 1980. In this article, atrioventricular block, perimyocarditis and mild heart failure were described as the features of Lyme carditis. The first specific articles on Lyme carditis in Europe were published in 1984 (2, 3), although already in 1973 a French patient was described with typical features of Lyme carditis (erythema migrans, arthralgias, Wenckebach atrioventricular (AV) block and perimyocarditis). However, this diagnosis could only be confirmed retrospectively in 1985, when the immunofluorescence assay became available for *B. burgdorferi* and the preserved serum was found positive in this patient (4). Other authors had related cardiac complaints and abnormalities to other manifestations of Lyme borreliosis (5, 6). Lately, the number of publications on Lyme carditis has increased rapidly, extending the pattern of Lyme carditis with other cardiac manifestations and showing the diversity of the possible expressions of this disease (7-13).

Nevertheless, it took several years before this manifestation of Lyme borreliosis was generally known in everyday clinical practice. Several International Conferences on Lyme borreliosis and many local symposia contributed significantly to the awareness of Lyme carditis as a treatable heart disease.

In this short article, the clinical features, diagnostic tools and therapy of Lyme carditis are summarized, including recent data on the subject.

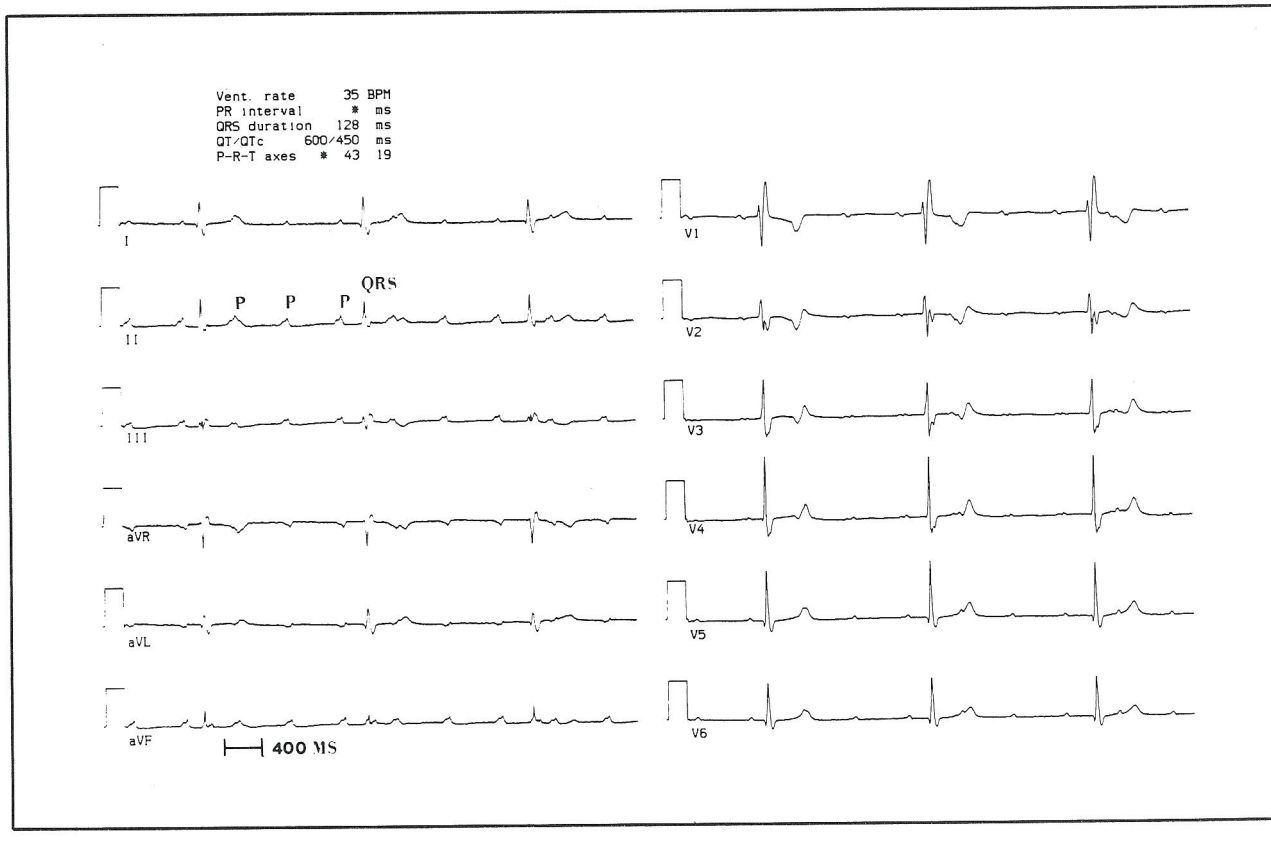

Fig. 1. Electrocardiogram of a 40-year old patient with complete AV block due to (biopsy proven) Lyme-carditis. The duration and configuration of the escape complexes suggest that they originate in the left bundle branch. The duration and configuration of the P waves indicate intra-atrial conduction delay. Sinus frequency 100/min, escape frequency 35/min, QRS duration 130 ms, and P wave duration 120 ms.

INCIDENCE

In the North American literature, the incidence of Lyme carditis was estimated as occurring in 8% of patients with Lyme borreliosis (14). In a recent prospective study of 61 patients with Lyme borreliosis however, the incidence of Lyme carditis was found to be only 1.6% (15). In the European literature, only retrospective studies on the incidence of Lyme carditis are available and a frequency of 1.6% in 817 German patients (16) and 3.3% in 272 French patients (17) with Lyme borreliosis were reported. Difficulties in confirming Lyme carditis, unfamiliarity with the clinical pattern and accidental cardiac abnormalities in cases with Lyme borreliosis without causal relation to this disease may lead to under- or overestimation of the frequency of cardiac manifestations in Lyme borreliosis (18). Lyme borreliosis and thus Lyme carditis can be found on every continent and in every country (1,13,19-26) although endemic areas are known.

Lyme borreliosis and -carditis may occur in either

sex, although Lyme carditis was found to be more frequent in men (male-female ratio 3:1) (9). It may occur at any age, even in children (23,24,27).

CLINICAL FEATURES

Lyme carditis may be the only manifestation of Lyme borreliosis. Lyme carditis may present as:

- Conduction disturbances (disturbances of impulse formation and conduction)
- Rhythm disturbances (extrasystoles, brady- and tachyarrhythmias)
- Pericarditis and/or pericardial effusion
- Myocarditis and/or recent onset heart failure
- Dilating cardiomyopathy as a late manifestation

Conduction disturbances:

Intra-atrial, AV, and intraventricular conduction disturbances may all occur. AV-block and especially a fluctuating degree of AV block is, however, the most frequently observed sign of Lyme carditis. Rapid changes in degree of AV block may occur. In several reports unstable escape foci have been described. The block may be restricted to the supra-hisian (i.e., in the AV node) or infra-hisian areas (i.e., in the common His bundle and bundle branches) or the conduction disturbances may be diffuse, in which case the AV node, common His bundle, bundle branches and the Purkinye system may all be affected, with a possible concomitant delay of intra-atrial conduction (Fig. 1) (7, 28).

Rhythm disturbances:

Rhythm disturbances in Lyme carditis seem to be less frequent than conduction disturbances, but may include atrial fibrillation or flutter, paroxysmal atrial tachycardia, premature supraventricular and ventricular beats and (sinus) bradycardia (7,9). In 1991 a single case was reported with Lyme carditis-related periods of nonsustained and sustained monomorphic ventricular tachycardia with global hypokinesis of a dilated left ventricle and normal coronary arteries (10).

Pericarditis, pericardial effusion, myocarditis and heart failure:

Pericarditis and pericardial effusion in Lyme carditis are frequently observed. Pericarditis can be found in up to 15% of patients with Lyme carditis and in half of these patients echocardiographically demonstrated pericardial effusion may be expected. In some cases this was complicated by right-sided heart failure. Up to 8% of the patients with Lyme carditis may experience myopericarditis (pericarditis associated with enzyme changes, but with no evidence of myocardial infarction), in some cases with right sided heart failure (9,28-32). Overt clinical left heart failure seems to be an infrequent manifestation of Lyme carditis, although it has been described in several reports (9,10,13).

Dilating cardiomyopathy:

Until recently, late manifestations of Lyme carditis were unknown. Stanek and coworkers (8) demonstrated that in a group of patients with dilating cardiomyopathy of unknown cause, some patients may have suffered Lyme carditis in earlier years, resulting in this late manifestation. Another case report on this subject was published in the former East Germany (13).

DIAGNOSTIC TOOLS.

In short, the diagnostic procedures include a complete history and physical examination with respect to Lyme borreliosis and more specific Lyme carditis. General laboratory tests and specific tests on *B. burgdorferi* are routinely performed. With respect to Lyme carditis, serial surface electrocardiograms (ECG's), chest X-rays, an echocardiogram and Holterscans have to be performed depending on the clinical situation. In case of first degree AV block with PR > 300 msec, high degree AV block, rapidly changing AV block, unstable escape foci or hemodynamically significant rhythm disturbances, hospital admission for continuous monitoring is necessary. The same applies for clinical heart failure. Multigated bloodpool scintigraphy can be done on specific indication. In case of diagnostic difficulties, a gallium-67 scintigraphy or an indium-111 scan can be performed to distinguish active myocarditis from primary cardiomyopathies and other non-inflammatory myocardial disorders (21,33-37). A normal gallium-67 or indium-111 scan however, does not exclude Lyme carditis (7,21,33). Endomyocardial biopsies are not routinely performed in Lyme carditis, but can differentiate acute endomyocarditis from a more chronic inflammation (33,38). The presence of spirochetes in endomyocardial tissue may be demonstrated by special staining methods (39,40). In case of diagnostic difficulties, endomyocardial biopsies may be indicated, possibly guided by a gallium-67 or indium 111 scan. Electrophysiological studies can be useful in Lyme carditis, although they are not fundamental for diagnosis and treatment. Electrophysiological studies in Lyme carditis may be restricted

to patients in whom it is already necessary to introduce a temporary pacemaker, in those patients where myocardial biopsies have to be performed and for research purposes in patients who have given special informed consent. In combination with the surface ECG, location and extension of the conduction disturbances (intra-atrial, supra- or infra-his) can be determined (7,41,42).

THERAPY

Treatment of Lyme carditis can be divided in two parts. First, the treatment of the manifestations and symptoms of Lyme carditis. In case of asymptomatic and low grade AV block or other mild rhythm or conduction disturbances, an expectative attitude with rhythm monitoring is sufficient. When symptomatic or hemodynamically compromising high degree heart block is present, a temporary pacemaker must be inserted. Retrospective analysis of 105 patients with Lyme carditis showed about 35 % use of temporary pacemakers worldwide (12). Patience with respect to permanent pacemaker implantation is important. Tachyarrhythmias must be monitored and, if necessary, treated with antiarrhythmic drugs (caution with negative inotropic drugs). Pericarditis and myocarditis, without right or left ventricular heart failure can be treated with a few days of complete rest. If heart failure is present, additional drugs like diuretics and vasodilators must be applied. Duration and intensity of these therapies depend on the clinical pattern. Secondly, antibiotic therapy should be given as soon as the suspicion of Lyme carditis is likely enough to justify treatment. Usually it is not possible to wait for the definite proof of Lyme carditis. The present-day recommended antibiotic choice is ceftriaxone (1x2 gram intravenously for 10-14 days), cefotaxime (2x2 gram intravenously for 10-14 days) or penicillin G (4x5 million U intravenously for 14 days) in case of clinically significant Lyme carditis. In asymptomatic first degree AV block (PR < 300 msec), tetracycline (3-4 x 500 mg orally for 10-30 days) or doxycycline (2x100 mg orally for 10-30) days, may be sufficient (43). The application of steroids is controversial. These recommendations are partly based on empirical data and partly on data from the results of treatment of other manifestations of Lyme borreliosis (44, 45).

COURSE AND PROGNOSIS

In general, Lyme carditis has a favourable course. More than 90% of the documented patients with Lyme carditis had a complete recovery within 2-6 weeks, with or without antibiotic therapy (9,12). In less than 10% minor conduction disturbances remain and in about 2% a permanent pacemaker had to be implanted (7,21). Despite these reassuring data, there have been at least 3 patients described with fatal Lyme carditis (20,46,47). Furthermore, recent reports have demonstrated the possibility of late manifestations of Lyme carditis like dilating cardiomyopathy (8,13).

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