

Lyme disease

David R Snyderman

Abstract

Lyme disease is the most common tick-borne illness in the USA and Europe. Pathogens involved are *Borrelia burgdorferi* in the USA and *B. afzelii* and *B. garinii* in Europe. The characteristic rash of erythema migrans occurs in approximately 75% of patients. Neurological disease, including facial palsy, meningoencephalitis, aseptic meningitis and polyradiculopathy, occurs in 10–15%. Cardiac disease, primarily manifest as heart block, is seen in 1–2%. Arthritis is seen as a late complication in about 30%. Patients with erythema migrans can be treated for 10–14 days with doxycycline 100 mg orally twice a day. For those unable to tolerate doxycycline, amoxicillin 500 mg orally three times per day for 14–21 days will be effective as will cefuroxime axetil 500 mg three times per day. For late manifestations, including central nervous system disease, oral doxycycline has been shown to be equivalent to parenteral ceftriaxone. Arthritis requires 28 days of treatment but can be accomplished with oral therapy. A small proportion of patients have some persistence of symptoms, but such post-Lyme disease symptoms or ‘chronic Lyme’ disease has not been shown to respond to >28 days of antibiotic therapy. Chronic long-term antibiotics are not recommended. Prevention measures currently rely on tick avoidance, tick repellants and careful inspection and removal if present. No vaccine for humans is available.

Keywords *Borrelia afzelii*; *Borrelia burgdorferi*; *Borrelia garinii*; chronic Lyme disease; doxycycline; erythema migrans; Lyme arthritis; Lyme disease; Lyme meningoencephalitis; MRCP; neuroborreliosis

Introduction

Lyme disease is caused by the organism *Borrelia burgdorferi* (genus recently changed to *Borrelia burgdorferi*) and was first recognized in the USA in 1975. It has become the most common tick-transmitted infection in the USA, with 28,000 cases reported annually and as many as 300,000 estimated annually in 2015 by the US Centers for Disease Control (CDC). Although the rash of erythema migrans is quite recognizable, one has to assume that the number of cases not reported is fairly sizeable. In Europe, about 35,000 cases are reported annually, and the disease, in retrospect, had been present for more than a century before the first reports emerged from the USA. In Europe, the species *B. afzelii* and *B. garinii* predominate as the cause of Lyme disease, although cases caused by *B. burgdorferi* and two other rare species, *B. spielmanii* and *B. bavariensis*, have been reported.

In the USA, co-infection with other pathogens, including *Babesia microti* and *Anaplasma phagocytophilum* (human

Key points

- *Borrelia burgdorferi* is the species seen in the USA whereas *Borrelia afzelii* and *Borrelia garinii* are seen in Europe; the clinical manifestations are similar
- Erythema migrans is the typical rash, with arthritis relatively common. Facial nerve palsy, radiculopathy and cardiac involvement are less common
- Oral antibiotics are usually sufficient, with doxycycline or amoxicillin most commonly used
- Post-Lyme syndrome (sometimes called chronic Lyme) does not respond to prolonged antibiotic therapy

granulocytic anaplasma), can occur during attachment and engorgement by the same tick vector, *Ixodes scapularis*. Two recently described species, *B. miyamotoi* and *B. mayonii*, have also been associated with tick transmission. The latter is found in the upper Midwest of the USA and presents like Lyme disease; the former has different clinical manifestations more akin to anaplasmosis.¹

Clinical manifestations

The most common manifestation following tick bite is the rash of erythema migrans. It typically arises in about 75% of patients within 1 or 2 weeks of a tick bite, although this can be as soon as 2 days or as long as a month.

The rash is typically circumscribed, and can be as large as 5 cm or more in diameter (Figure 1). A bull’s eye appearance is seen in 30–40% of those with erythema migrans (Figure 2). In areas with a lot of hair, the rash may not be so obvious (Figure 3) or may look like a large area of ‘sunburn’, so careful inspection can be required. In addition, the rash may not be readily apparent in persons of colour. The rash can sometimes appear vesicular. Multiple erythema migrans lesions can also occur, representing early dissemination (Figure 4). Typical areas where the rash is found include the inguinal region, popliteal fossa, axilla and abdominal wall.

The rash can itch, burn or be hot to the touch, but it is seldom painful. Lymphadenopathy may be present around the area affected. Patients can have a flu-like illness with fever, anorexia, fatigue, muscle aches and arthralgias. The laboratory abnormalities can include leucopenia, or leucocytosis, thrombocytopenia, elevations of the erythrocyte sedimentation rate, and even mild abnormalities of liver function tests.

Frank arthritis has been reported to develop in 30% of untreated cases in US surveillance. The arthritis is monoarticular, and typically involves large joints such as the knees, although it can be pauciarticular. Lyme arthritis is a manifestation of late-stage disease, occurring in patients who were not treated in the early stages.

Other common features (10–15%) of Lyme disease include facial nerve palsy (which can be bilateral and is one of the few causes of bilateral facial nerve palsy), lymphocytic meningitis

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Figure 1 Erythema migrans can be as large as 5 cm or more in diameter. Photograph courtesy of Linda Kaplan MD.

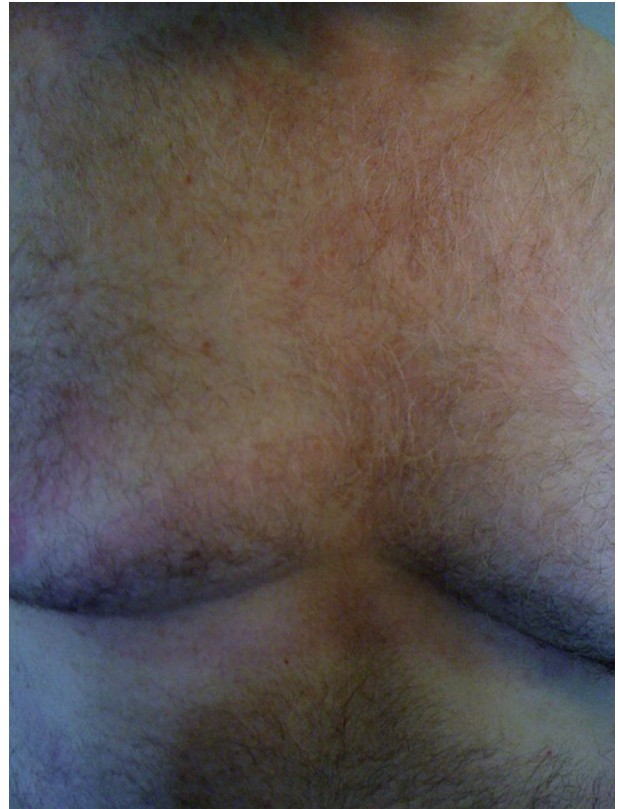


Figure 3 Erythema hidden on the chest where hair is present. Photograph courtesy of Linda Kaplan MD.



Figure 2 Typical bull's eye with central clearing. The border outlined by red marker dots shows the extent of the spreading erythema before treatment was initiated. Photograph courtesy of David R. Snyderman MD.

and radiculopathy, which can be associated with significant pain. Myopericarditis is much less common, occurring in about 1–2% of patients, and is usually manifest by varying degrees of heart block. Sudden cardiac death has been rarely reported. Cardiomyopathy caused by Lyme disease has been reported in Europe. Ocular manifestations including iridocyclitis, keratitis, conjunctivitis, optic neuritis, vasculitis, and uveitis have been reported but are rare. Conjunctivitis can occur in as many as 10% of patients.

Other manifestations of central nervous system disease are rare and include chronic encephalopathy and encephalomyelitis. In both Europe and the USA, encephalopathy has been reported with mild cognitive disturbances, including poor memory and concentration. Spinal radiculopathy or paraesthesias can occur. A form of chronic spastic paraparesis, cognitive impairment and cranial neuropathy seen in Europe is caused by *B. garinii*.

Uncommon skin manifestations of Lyme disease include acrodermatitis chronica atrophicans and morphea-like skin lesions. Both have been described in Europe and are associated with *B. afzelii*. Typically, acrodermatitis chronica atrophicans appears years after infection, and manifests as bluish-red discoloration over the extensor surfaces of the hands or feet. Fibrous nodules over the elbow or patella have been seen. A borrelial lymphocytoma has also been described in Europe, and can be seen in the scrotum or around the ear lobe in children or nipples in adults. It can occur in sites with previous erythema migrans.

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