

## Case Report

# Borrelial Lymphocytoma in children: don't miss this skin marker of Lyme Disease

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### Keywords

Lyme Disease, lymphocytoma, *Borrelia burgdorferi*

### Abstract

Borrelial lymphocytoma is a rare but pathognomonic cutaneous manifestation of Lyme disease. It mainly occurs in the early disseminated stage (secondary stage). Here we report two cases of borrelial lymphocytoma in children. Furthermore, the steps to the diagnosis as well as the treatment are discussed.

### Introduction

Lyme disease or Lyme borreliosis is a systemic disease caused by the spirochete *Borrelia burgdorferi* sensu lato (*B. afzelii*, *B. garinii* et *B. burgdorferi* sensu stricto in Europe (1)). This spirochete is transmitted to humans by ticks, most frequently by *Ixodes ricinus* in Europe (1). The clinical manifestations can vary and are divided into three stages: the first stage with early localized manifestations that appear a few days to a few weeks after the tick bite, the second stage with early disseminated manifestations that appear several weeks to several months after the bite, and the third stage with late disseminated manifestations appearing months or even years after the contamination (2).

The incidence of Lyme disease in Belgium is difficult to assess due to the multiple clinical presentations. In 2018, 90.3 per 100,000 people consulted their general practitioner with erythema migrans, a clinical manifestation of the early localized stage of Lyme borreliosis for which serological testing is not necessary to confirm the diagnosis (3). Furthermore, a study by Lernout et al. on blood samples from July 2013 to January 2015, shows a seroprevalence for *B. Burgdorferi* sensu lato of 1.06% (4). The number of hospitalizations for Lyme disease in Belgium was around 300 patients per year from 2010 to 2014 (2).

Borrelial lymphocytoma is a cutaneous manifestation that essentially appears in the second stage of Lyme disease, from the 10th day to 6 months after the tick bite according to sources (1,5-6). It is a 1-5 cm bluish, erythematous or purplish nodule usually localized on the earlobe in children and in the nipple area in adults (7). It can also be found in other areas of the body. This symptom can be found in 0.9-5% of patients with Lyme disease (6,8). It is more frequent in children than in adults occurring in 7% of the cases (5,9). Borrelial lymphocytoma is a clinical manifestation of Lyme disease caused by *B. afzelii* or *B. garinii* (1,5). These species are almost exclusively present in Europe, making this symptom endemic in Europe (1,5).

We here report two cases of borrelial lymphocytoma in children.

### Case reports

A 7-year-old girl, with no particular medical history, presented with a 5-month recurrent erythema and swelling of the left earlobe, sometimes associated with itching (Figure 1). A transient erythema on the left cheek gradually

Figure 1 : borrelial lymphocytoma of the left earlobe.



spread over the entire left side of the face at the same time. About 4 months later, the patient developed fatigue and daily left-parietal headaches. The ENT examination, ophthalmologic check-up and the cerebral MRI were unremarkable. Two blood serologies for *Borrelia burgdorferi* sensu lato performed 15 days apart, proved positive for IgG and negative for IgM. No personal history of tick bite was reported. With these typical clinical manifestations and the serological results, the diagnosis of borrelial lymphocytoma was made. A lumbar puncture was performed, revealing an elevated white blood cell count as well as an intrathecal secretion of anti-Borrelia IgG. This revealed an associated neuroborreliosis. Treatment by intravenous cefotaxime (200mg/kg/day) was given followed by oral doxycycline (4mg/kg/day) for a total period of 28 days. Fifteen days after starting the antibiotherapy, the earlobe nodule and the headaches disappeared, and the erythema of the face appeared less frequently.

A previously healthy 8-year-old boy sought medical advice as he presented with a one-month non-pruritic, painless purple nodule, located on the left breast areola (Figure 2). An erythema surrounding the nodule and an annular erythema on the left side of the patient's back appeared about 15 days later. There was no history of a tick bite but the patient had attended a summer scout camp a few months earlier. On biological testing, anti-Borrelia titers were positive for IgG and negative for IgM. The diagnosis of borrelial lymphocytoma was made and a treatment by amoxicillin (500 mg 4 times a day) was started for a total of 3 weeks. After 15 days of antibiotherapy, the erythema had disappeared and the size of the nodule diminished. The good response to the treatment confirmed the diagnosis.

## Discussion

The diagnosis of borrelial lymphocytoma is mainly based on clinical examination, combined with a history of tick bite(s) or of visiting places where there is a risk of being exposed. Serological testing helps to confirm the diagnosis. A two-tier approach is recommended with an enzyme immunoassay confirmed by immunoblotting (6,8-10). However, it can initially be negative, turning positive afterwards. It is sometimes necessary to perform a skin biopsy for differential diagnosis if the history is not typical, for example if there is no response on antibiotics, and serologies are not helpful. Indeed, this type of lesion can also be seen in lymphomas, cutaneous lupus, sarcoidosis and other dermatological disorders (8). Histological analysis shows lymphocytic inflammatory infiltration of the dermis with plasmocytes and lymphoid germinative centres (1,5,8). Search for the spirochete via PCR or culture can also be done on the biopsy but the sensitivity is variable (1,6,8).

The duration of the therapy should be 2 weeks if the borrelial lymphocytoma is the only sign of presentation (10). The choice of the antibiotic depends on the age of the patient and other criteria listed in the table below (Table 1).

**Table 1:** antibiotic type and dosage in the treatment of borrelial lymphocytoma (1). Duration of treatment is of 14 days (10).

Population	Antibiotic	Dosage	Maximum dosage
7-8 years or older	Doxycycline	4 mg/kg/day	200 mg/day
- Younger than 7-8 years	Cefuroxime	30 mg/kg/day	1000 mg/day
- Allergy to doxycycline	Amoxicillin	50 mg/kg/day	1500 mg /day
- Pregnancy or breast-feeding			

With appropriate antibiotics, the lymphocytoma disappears within 2 weeks to 2 months with a mean of one month, as in our two cases (6,10).

If other signs or symptoms are associated, the duration of the treatment as well as the route of administration may vary, which was the case for our first patient who had concomitant neuroborreliosis.

**Figure 2 :** borrelial lymphocytoma of the nipple.



It is important to ensure follow-up after the initiation of antibiotherapy to make sure of efficacy. If treatment doesn't work, it is necessary to revise the differential diagnosis.

## Conclusion

Borrelial lymphocytoma is a rare, easily misdiagnosed but pathognomonic cutaneous manifestation of Lyme disease in Europe. It is important to make appropriate diagnosis to start treatment as soon as possible and to avoid progression towards a later stage of the disease, for example neuroborreliosis. A thorough anamnesis, the features and localisation of the nodule, serological testing and follow-up ensuring a good response to the treatment are so many keys for diagnosis and can avoid unnecessary biopsy.

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