Atypical dermatological manifestations of Lyme borreliosis

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SUMMARY

Lyme borreliosis (LB) is a multisystemic infectious disease involving the skin, joints, nervous system, heart, and eyes. Today at least three subtypes pathogenic for humans have been identified: *Borrelia burgdorferi* sensu stricto, *Borrelia garini, Borrelia afzelii.* Different genospecies strains of Borrelia have been associated with different clinical manifestations. LB is classically described as having three clinical stages or, similarly to syphilis, an early phase and a late one. The early infection corresponds to the first stage, the late infection includes the second and the third stages. LB skin manifestations could be divided into five classes. Erythema migrans, lymphadenosis benigna cutis, and acrodermatitis chronica atrophicans are proven skin manifestations of LB. Lichen sclerosus et athrophicus, morphea, scleroderma, scleredema Buschke, atrophodermia of Pierini and Pasini, Parry-Romberg progressive facial hemiatrophy, and Shulman fasciitis are controversial LB manifestations. Granuloma annulare, atypical persistent pityriasis rosea, and pityriasis lichenoides are skin lesions occasionally related to LB. Urticaria, erythema nodosum, and papular acrodermatitis (Giannotti Crosti disease) are reactive LB skin manifestations. Nodular panniculitis (Pfeifer-Weber-Christian), B-cell cutaneous lymphoma, and juvenile chronic myeloid leukemia are exceptional skin manifestations of LB.

KEY WORDS

Lyme borreliosis skin, atypical manifestations,

In the last years, there have been numerous and important advances of many aspects of Lyme borreliosis (LB). However, it appears that many questions concerning this disease remain unanswered. It is not clear, how spirochetes behave when they enter into the human body. They can produce pathognomonic lesions, skin manifestations mimicking other diseases, or clinical pictures that can be induced also by other etiologic agents.

We became aware of the complexity of this disease since genetic studies can identify different species of *Borrelia burgdorferi* (Bb) sensu lato responsible for human infections: *Bb* sensu stricto, *B. garinii* and *B. afzelii.* In Japan a new species was recently described, *B. japonica*, which does not appear to be a human pathogen. In future, additional species of Borrelia will probably be identified. The recurrent fever also is caused by several species of Borrelia, and the vectors are different ticks (*B. recurrentis* transmitted by *Pediculus sp.* in epidemic relapsing fever, *B. caucasica, B.crocidurae, B. duttonii, B.hermisii, B. hispanica, B. mazzottii, B.*

parkeri, B. persica, B. turicatae, B. venezuelensis transmitted by *Ornithodoros sp.* in endemic relapsing fever). Diagnosis of LB is certain, when the infection is transmitted by a hard tick of genus Ixodes, and erythema chronicum migrans (ECM) appears.

Different *Bb* species and strains can have different organ tropisms and can induce different clinical manifestations. *B. afzelii* has been found in patients with acrodermatitis chronica atrophicans (ACA), while *Bb* sensu stricto in patients with arthritis and *B. garinii* in patients with neuroborreliosis. A possible *Bb* follicular hair tropism can explain the ECM with hair loss.

Atypical LB skin manifestations could be ordered in the following classes:

1.- Controversial LB skin manifestations
2.- Skin manifestations occasionally related to LB
3.- Reactive LB skin manifestations
4.- Exceptional skin manifestations during LB.

Skin lesions that appear immediately after tick-bite should also be mentioned: they can be mild and transient reactions, or, sometimes, edematous papular dermatitis of some centimeters of diameter, exceptionally with tissue necrosis.

Bb infection requires certain preconditions:

- Infected tick
- Attachment of the tick to the skin
- Appearance of enlarging erythema after an incubation period of at least 4-5 days

During the visit the tick can be observed on the skin, or sometimes the patient shows a detached tick in a little box. In such cases the tick bite is certain, while in all other cases the tick bite may be only supposed.

1.Controversial manifestations of LB

Essentially the following atrophic and sclerosing (sclerodermatous) disorders can be included:
- Lichen sclerosus et atrophicus (LSA)
- Morphea
- Scleroderma with generalized plaque lesions
- Linear scleroderma
- Atrophoderma profundum (Pierini-Pasini)
- Parry-Romberg progressive facial hemiatrophy
- Shulman's syndrome (Eosinophilic fasciitis)
- Buschke disease

Lichen sclerosus et atrophicus (LSA) has been related to Borrelia infection by Åsbrink who noticed the frequent association between LSA and ACA. Aberer demonstrated the presence of Borrelia in LSA. It is characterized by sclerotic atrophic patches, sometimes confluent, and often located on genitals. In 3 children affected by LSA living in endemic areas, borrelial DNA was found in the involved skin, by PCR (2). No specific DNA was found in 4 cases affected by LSA living in non endemic area. The same authors confirmed the relation of morphea to Borrelia infection, while others maintain that there is no such correlation. Probably morphea can be caused sometimes by *Borrelia afzelii* (1).

The possible relationship between LB and LSA or morphea is suggested by the following evidence:

- Clinical and histological similarities between morphea, LSA and ACA.
- The presence of antibodies against *Borrelia burgdorferi* in some patients with LSA and/or morphea.
- Identification of borrelial organisms in histological sections.
- The coexistence of ACA, LSA and/or morphea in the same patient.
- A response to antimicrobial therapy in many cases of LSA and morphea.

2. Skin manifestations occasionally related to LB

Granuloma annulare (GA) has been described in association with LB. In some cases the author was able to find positive serological tests or spirochetal bodies in the affected skin by silver stain. In his experience the GA is very seldom related to LB. In 3 patients he detected Borrelia in the affected skin by PCR, but in these cases clinical evolution has been unusual and the treatment by nimesulide hasn't been effective.

Atypical persistent pityriasis rosea, lasting longer than 4-5 months, could be suspected of being related to LB.

The author is studying some children who developed a papular dermatitis with perifolliculitis, mimicking *pityriasis lichenoides* (2). In one case he was able to isolate Borrelia sp. in BSK from the involved skin. Further studies are necessary to confirm the relationship with Lyme disease (3).

3. Reactive skin manifestations of LB

Such manifestations can be observed also in other infectious diseases:

- urticaria,
- erythema nodosum and

- papular acrodermatitis

Two varieties of urticaria can be distinguished:
- diffuse
- localized

The first form appears usually in early LB, whereas the localized form is more frequent in late LB. The localized form often involves the skin adjacent to the affected joints.

Erythema nodosum has also been observed during active LB. Recently, the author has reported two children who have developed *papular acrodermatitis (Gianotti-Crosti disease)* after borrelial infection.

4. Exceptional skin manifestations described during LB

Hassler (4) has reported an association between LB and involvement of subcutaneous tissue (*Pfeifer-Weber Christian disease*) and he has been able to demonstrate the presence of Borrelia in the affected skin even after several antibiotic treatments. There is also the problem of a possible correlation between LB and cutaneous B-cell (or T-cell) lymphomas. Evolution of borrelial lymphadenosis benigna cutis (LABC) towards malignancy has been supposed, but this hypothesis needs further investigations.

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