MANIFESTATION OF LYME DISEASE IN CHILDREN

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SUMMARY

Lyme disease (D) is a multisystem disorder caused by the spirocheta *Borrelia burgdorferi (Bb),* transmitted by Ixodes ticks. ECM is the main localized and pathognomonic symptom of the early stage of LD. In children the erythema is most frequently situated in the face. Lymphocytoma (LC) is another early skin manifestation of LD. In children the most frequently involved site is the face, especially the earlobes. There are a few cases reported of acrodermatitis chronica atrophicans (ACA) in children. The dominant joint symptom is an acute gonarthritis. Unlike in adults, neuroborreliosis in children manifests itself predominantly in acute meningeal symptoms or in acute peripheral facial palsy. Cardiac symptoms of LD have been rarely described in children. The author's own observations in 120 children with suspect LD are reported.

KEY WORDS

Lyme disease, children, skin symptoms, manifestations in other system.

Lyme disease (LD) is a tick-borne infectious disease transmitted by the spirochete *Borrelia burgdorferi (Bb).*

It is a multisystemic disease rather common in childhood: outdoor behaviour and playing habits of children make them to be more likely bitten by ticks than adults.

Early manifestations, including erythema chronicum migrans (ECM), meningitis and carditis, begin weeks after the tick bite, and are usually characterized by self-limited inflammatory changes. Late manifestations include arthritis, encephalomyelitis and acrodermatitis chronica atrophicans (ACA).

Despite the fact that LD frequently affects children, published studies concerning exclusively this particular group of patients are rather rare.

Cutaneous manifestations

ECM is the main localized and pathognomonic symptom of the early stage of LD. After a latent period of one to three weeks, erythema develops at the site of the tick bite from where it spreads centrifugally. The typical ECM lesion shows a central discoloration and a red annular edge, often with a circular shape.

Although it is the most frequent skin lesion, ECM does not appear in all the patients: in a study comprising 36 children, only 13 developed the ECM (1). The location of the skin lesion differs in adults and in children. While in adults the erythema is situated most frequently on the extremities, and to a lesser extent on the trunk in children the face is frequently involved (2,3,4).

ECM is often accompanied by systemic symptoms such as fever, headache, malaise, fatigue and arthralgia. Meningeal symptoms may occur simultaneously, or shortly after the appearance of ECM.

Lymphocytoma (LC) is another skin manifestation typical of the early stage of LD, but it is less frequent than ECM (5,6). LC is characterized by reddening and infiltration of certain areas of the skin and it forms nodules or plaques of livid color. The lesion is due to lymphocytic proliferation. The most frequently involved site is the face, especially the earlobes in children, followed by the extremities and by the nipple. (7,8,9). LC may be the only skin manifestation of the early stage of LD and it appears weeks or months after the tick bite. It may follow ECM after a latent period, or rarely it may occur at the same time as ECM. The first description of LC in connection with ECM in a 4-year-old girl was by Strandberg in 1920 (10). In contrast to ECM, the LC often persist for months. The incidence of LC is difficult to assess, but according to certain authors about 10 % of affected persons are children (11,12).

Acrodermatitis chronica atrophicans (ACA) is a characteristic skin manifestation which occurs in the late stage of disease (8,13,14). Painless red nodules or plaques appear mostly on the lower extremities (15), but also on the upper extremities and on the rest of the body. After months or years the peripheral extension may continue, while in the center of the lesion an atrophy of the epidermis, dermis and subcutis develops. In such a way, two phases can be distinguished: an acute phase, characterized by swelling, and a chronic phase characterized by atrophy and sclerosis. In contrast to ECM, in ACA there is no spontaneous remission so it can persist for years or even decades. It is not frequently reported today, probably due to antibiotic treatment performed in the early stage. However children account for approximately 10% of the reported cases (14,16,17).

Various types of lesions were described: fibrotic nodules, sclerotic areas, anetoderma-like lesions and ulcerations (especially on lower extremities) (14,18). In 1902 Herxheimer reported this manifestation in 12 children. An interesting case was reported by Trevisan (19), who described a 6-year old girl with ECM, and a concurrent atrophy with sclerosis of the left breast.

Joint manifestations

Pain and swelling is a characteristic manifestation in early LD. Usually one joint is affected or there may be a few. The most frequent sites in decreasing order are: knee, ankle, elbow, shoulder, wrist, hip, foot and hand (20). Intermittent migratory arthralgias involving tendons and muscles were also reported (21). Frank arthritis is unusual in the first several weeks of illness. Steere, who first described Lyme arthritis in children, observed that the most frequently involved joint was the knee (21). Other studies also revealed that gonarthritis was the typical joint manifestation of LD. In a study involving 43 children, the knees were affected in 41 patients; of these eight also had arthritis in the elbow joint. A tick bite was ascertained in 19 out of 20 children with ECM preceding the joint manifestations. (22,23). Bone erosions seem to be rare in children, and the arthritis may disappear without treatment (24,25).

In Europe arthritis does not seem to be a frequent manifestation of LD in children. According to Kryger only two out of 48 Danish children with arthritis presented elevated antibody titers against Bb (26). In a Swedish study the arthritis was caused by Bb in 10 out of 300 patients with arthritis (27). Swiss authors reported a somewhat higher percentage of affected joints (28). In contrast to this, in the USA the involvement of joints in children has been frequently mentioned by Steere (21) and by Williams (20) with 48% and 64% respectively.

Neurological manifestations

In the early stage, neurological manifestations of LD can be divided into meningeal and peripheral symptoms. Meningeal symptoms include fatigue, fever, headache, malaise, vomiting, and neck stiffness. These symptoms are accompanied by abnormalities in cerebrospinal fluid (CSF) such as pleiocytosis,

elevated proteins and glucose levels, and the presence of specific antibodies against Bb. The involvement of peripheral nervous system includes facial nerve paralysis, which is rather frequent in children, and radicular pain which on the contrary is rare (29,30). Paralysises of other cranial nerves have also been reported (30,31).

Manifestations of the late stage are: meningitis, myelitis, cranial and peripheral neuritis and encephalitis (31,32). This topic has been reviewed in depth (33,34). In his analysis of 208 children with LD, Christen detected 169 neuroborreliosis cases by detection of specific antibodies to Bb in CSF. In over more than 60% of cases acute peripheral facial palsy or an aseptic meningitis were detected (35).

As a principle, the whole spectrum of symptoms that have been described in adults can also be observed in children. However, differences concerning the relative frequency and course of these manifestations occur.

Bannwarth in 1941 described four children with acute peripheral facial palsy and evidence of pleicytosis in the CSF. He mentioned that, in contrast to adults, the typical radicular pain was absent while facial palsy, frequently with bilateral manifestations, was a major problem. Such observation was confirmed later (36). From an historical point of view, it is also interesting that in 1946 Zellweger coined the term "chronic allergic meningitis" for this set of symptoms (37). Meningitis appearing after ECM in a child was described by Schirduan in 1950 (34).

Progressive encephalomyelitis, as a late manifestation of LD was also described in children (32, 38, 39). This manifestation may be responsible for spastic paresis, bladder dysfunction, ataxia, involvement of cranial nerves, depression, cognitive dysfunction, memory loss, somnolence and irritability.

Cardiac manifestations

Atrioventricular block of varying degrees is the most common manifestation of cardiac involvement in LD, although more serious complications such as syncopes, dizziness, palpitations, myocarditis and even pericardial effusion may occur. Cardiac symptoms of LD have been rarely described in children.

Lyme carditis manifests itself in varying degrees of atrioventricular block. Some cases with both myocarditis and pericardial effusion were also described (40, 41). Cardiac dysrythmia was reported in a child with syncope who previously had ECM, requiring the implantation of a temporary pacemaker (41).

Author's experience

During the last eight years 1422 patients, 120 (8%) children, complaining of symptoms compatible with Bb infection were referred to us by dermatologists, neurologists, general practitioners and pediatricians, or they were seen at our clinic for the evaluation of musculo-skeletal symptoms (42). LD was confirmed in 195 (14%) of the patients, 22 (11%) were children.

In 20/22 (91%) children we detected antibodies to Bb, in 2 children the diagnosis LD was confirmed by the presence of ECM only.

Eighteen of these 22 children lived in an endemic area for LB. ECM was present in five cases, in one case with a simultaneous monoarthritis of the knee. Neurological involvement was present in 7 children: facial palsy (2), optic neuritis (1), radiculitis (2), encephalitis (1) and pseudotumor cerebri (1) were the clinical manifestations observed in these patients. Eight children suffered from articular involvement of LD: 7 had frank arthritis, 4 with oligoarthritis and 3 with monoarthritis. Knee was the most frequently involved joint (5/7), followed by wrists (1/7) and the hip (1/7). Arthralgia only was complained by one child. The majority of patients (21/22) presented only one manifestation of the disease; two manifestations were present in one child only.

In our patients there is a higher prevalence of articular manifestations, followed by neurological and cutaneous involvement. However the prevalence of arthritis is lower than that reported in the USA, and higher than reported elsewhere in Europe.

The prevalence of neurologic manifestations in our children is not different from USA and other European countries. The dermatologic involvement is lower than in USA and in Europe. Cardiac manifestations were not observed in any of the children we studied.

CONCLUSION

Clinical manifestations of LD are somehow different in adults and children. The site of ECM differs in adults and children; in the children the face is the area most often affected, in adults ECM is most often found on the limbs. Lymphocytoma manifests itself in distinct changes of the skin. The sites most often affected are the earlobes in children and the nipples in adults. The ACA may develop in children very rarely, and the most common sites are the distal parts of the extremities as in adults. Acute Lyme arthritis, frequently a mono or oligoarthritis

with an intermittent pattern most typically affects the knees. Articular erosions seem to be rare in children.

Unlike in adults, neuroborreliosis in children manifests itself predominantly with acute meningeal symptoms, and with an acute peripheral facial palsy. Cardiac symptoms in LD have been rarely described in children, but no studies based a systematic cardiac examination have been done.

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