Lyme Disease and Seventh Nerve Paralysis in Children

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Purpose: This study was undertaken to determine the frequency of Lyme disease (LD) as a cause of transient facial nerve palsy (FNP) in children. Acute onset FNP in children has been primarily associated with acute otitis media (AOM). Recently, LD has emerged in regions where the deer-tick vector is present and has been associated with multiple cranial neuropathies.

Patients and Methods: Fifty children with transient FNP were evaluated and treated at our institution over a 5.5-year period.

Results: The rank of etiologies confirmed LD to now be the most common (50%), followed by AOM (12%), varicella (6%), Herpes zoster (4%), and coxsackievirus (2%). Thirteen children (26%) had idiopathic FNP consistent with Bell's palsy.

Conclusion: We conclude that transient FNP in children is most commonly caused by LD for regions with endemic infections caused by *Borrelia burgdorferi.*

The acute onset of facial nerve palsy (FNP) in children may often have an infectious etiology.[1,2] In the past, acute otitis media (AOM) was felt to be the most common infectious bacterial cause of FNP. The use of antibiotics has changed this pattern.[1-4] Viruses have emerged as the most frequent infectious cause of FNP.[1,3,5] Idiopathic FNP (Bell's palsy) remains a diagnosis of exclusion and accounts for most cases of acute onset FNP when an underlying infection is not detected.[1,6,7]

Lyme disease (LD) is caused by the spirochete *Borellia burgdorferi.*[8,9] Recent reports by Christen et al[10] suggest that within Western Europe, LD is the leading cause of acute FNP in children. We therefore undertook a retrospective study of all cases of FNP in children seen at our children's hospital (the mid-Atlantic region of the United States) to determine the frequency of LD as a cause of transient FNP in children.

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PATIENTS AND METHODS

All medical charts at The Alfred I. duPont Institute children's hospital over a 5.5-year period (January 1, 1990 to June 30, 1995) were reviewed for the diagnosis of FNP. All children under the age of 19 years were eligible. Referrals came from southeastern Pennsylvania, Delaware, and southern New Jersey, a geographic area of endemic LD. Children were evaluated by specialists in pediatric infectious disease or rheumatology in a clinic with special focus on consultative LD.

The diagnosis of LD was based on either the presence of the pathognomic rash of erythema migrans or the characteristic systemic signs such as arthritis/arthralgias, cranial neuropathy, and aseptic meningitits. Serum-positive Lyme enzyme-linked immunosorbent assay (ELISA) was an important diagnostic tool and was combined with immunoblot reactivity when equivocal.

RESULTS

The medical records of 67 children with FNP were identified and reviewed. Seventeen children were excluded because the cause was determined secondary to trauma or tumor, and/or the duration of the FNP could not be determined as a result of loss of follow-up. Therefore, 50 children were identified and compared. Twenty-five children had FNP secondary to LD, six from AOM, and six from viral causes (three from Varicella zoster, two Herpes zoster, and one from coxsackievirus).

Thirteen cases were considered to be idiopathic FNP consistent with Bell's palsy (Table 1).

The 25 children with FNP and LD (Table 2) included 18 boys and seven girls. The mean age was 10 years and all were white. The side of FNP was almost equally distributed between right and left (14:11). There were no bilateral cases identified. The mean duration of FNP was 5.1 weeks. Over 80% (21/25) of the children had systemic symptoms (eg, headache, arthralgia, fevers) at the time of the facial neuropathy. The arthralgias were located in the head and neck region 1.6 times more frequently than in the upper and lower extremities (10:6) when present. One child in the LD group reported altered taste, but none of the other children in this group reported or were noted to have any other associated cranial nerve signs or symptoms. All children in the FNP + LD group received antibiotic treatment consisting of amoxicillin, doxycycline, or ceftriaxone.

The duration of the FNP in the LD group was compared with the 12 children with FNP of other identifiable causes. Again, this group showed a slight male predominance (7:5). The duration of FNP appears longer in the non-LD group (11.4 *v* 5.1 weeks).

DISCUSSION

Lyme disease, the most frequent vector-borne infection in the United States, is caused by the spirochete *B burgdorferi* and is spread by the bite of Ixodes genus ticks.[8,11] Lyme disease may manifest itself in three overlapping stages. Initially, a classic annular rash termed erythema migrans (EM) at the site of the tick bite may be seen within the first weeks of infection.[8,12] In children, the bite and rash are more frequently located on the head or neck region than in adults.[8,10] Fever, headache, arthralgias, myalgias, neck stiffness, and generalized malaise are also noted at this stage.[8,12,13] Neurologic manifestations can subsequently occur either within this first stage or later in the disease, with FNP being the most common peripheral neuropathy.[8,11,13-17] In children, the FNP may be unilateral, occurring on the same side as the EM, or bilateral.[10,16] Other central and peripheral neurologic manifestations of Lyme borreliosis may be seen, particularly as the disease stages advance.

TABLE 1. Diagnosis of 50 Children With Facial Paralysis

Diagnosis	No.	%
Lyme	25	50
Otitis media	6	12
Varicella zoster	3	6
Herpes zoster	2	4
Coxsackievirus	1	2
Idiopathic	13	26
Total	50	100

In the secondary stages, lymphocytic meningitis, painful radiculoneuritis, and mild encephalitis can be noted. Cardiac conduction defects may also be apparent at this time. If untreated, progression to stage III disease, which is marked by arthritis and chronic central nervous system disease, may occur.[8,12]

The acute onset of FNP in children can present a diagnostic dilemma that has significant impact on disease outcome. In endemic regions, the physician must maintain a high index of awareness of LD as a possible etiology. Failure to diagnose LD places the patient at risk of developing stage II or III disease with the attendant sequelae. While various clinical factors, such as EM or concomitant aseptic meningitis, may provide a worthwhile guide to diagnosis, they may not always be present or they may not be specific for LD.[18] Laboratory tests help, but early in the course of LD, there is a lack of immunoglobulin G (IgG) antibodies.[19]

Gerber and Shapiro[14] have indicated that only 30% of patients can document a tick bite as a cause for LD. Although the classic annular rash of EM provides strong clinical evidence of *B burgdorferi* infection, perhaps 50% to 60% of patients may fail to demonstrate it and there are other cutaneous diseases that may act as false positives.[8,16] Because of the tick vector, peak incidence of LD is reported to occur in the summer to fall months.[10,12] In our experience, most cranial neuritis occurs in the late spring to early summer.

The constitutional "flu-like" symptoms that frequently accompany early stage LD may also act as a guide to the etiology of FNP. Fever and headache are the most frequent symptoms associated with LD.[13,20] Facial nerve palsy will typically manifest itself early in LD and has been noted to arise during or even after treatment has been initiated.[16] In children, EM frequently occurs on the head and neck re-

TABLE 2. Characteristics of 25 Children With Lyme Disease

Age (yrs)	Sex	CR VII	Duration (wk)	Headache	Rash	Arthralgia
15	male	left	3	pos	neg	TMJ
10	male	right	10	pos	pos	knee
18	male	right	4	pos	pos	neck
10	male	left	8	pos	neg	neck
9	female	right	2	neg	pos	neg
11	female	left	4	neg	pos	elbow
3	male	right	2	neg	pos	neg
14	male	right	12	neg	neg	neg
6	male	left	4	neg	neg	neck
12	male	left	3	neg	neg	neg
7	male	left	4	pos	pos	knee
8	male	right	20	pos	pos	knee
5	female	left	7	neg	pos	neg
11	male	right	3	pos	pos	neck
12	male	right	4	pos	neg	neck
8	female	right	2	pos	pos	neg
7	female	left	2	pos	neg	neg
8	male	left	2	pos	neg	neck
7	male	right	4	pos	neg	neg
9	male	right	4	neg	pos	TMJ
11	male	right	2	neg	pos	knee
5	female	right	2	pos	pos	neck
11	female	left	9	neg	neg	neck
17	male	left	2	pos	pos	ankle
13	male	right	12	neg	neg	neg
Average 9.9	18 M:7 F	14 right:11 left	5.1 weeks	15 pos:10 neg	14 pos:11 neg	10 HN:6 extremities

Abbreviations: CR, cranial nerve VII; HN, head and neck arthralgia; TMJ, temporomandibular joint.

gion. This is in contradistinction to adults who typically develop EM on the lower extremities. This likely results from a child's shorter stature. That subsequent FNP arises on the ipsilateral side suggests direct neural invasion, although there is no histopathologic evidence for this.[10] Additionally, FNP is one of the more common neurologic manifestations of LD, especially in children.[10,13,16,17] Children also manifest bilateral FNP more frequently than adults. Bilateral FNP is more frequently caused by LD than other causes.

Early CNS invasion can occur in LD, and this has significant impact on antibiotic therapy.[21] The question arises as to whether FNP indicates CNS disease. It may be necessary to consider a lumbar puncture and intravenous antibiotics in LD patients with FNP associated with fever or systemic symptoms.

In general, the duration of FNP with LD is reported to be shorter than with other causes of peripheral FNP.[11,15] Resolution may even begin before beginning treatment, and overall prognosis is typically good.[15,16] We did note several patients who had FNP lasting many months even after intravenous ceftriaxone.

The laboratory diagnosis of LD can be difficult. Erythrocyte sedimentation rates, while elevated, are nonspecific. Cerebral spinal fluid (CSF) lymphocytosis and elevated CSF proteins can be seen in viral meningitis as well as LD. Although specific antibody tests may be beneficial, in early LD, the IgM may not be positive for 3 to 4 weeks and the IgG may not be positive for 6 to 8 weeks.[14]

ELISA testing is efficient and sensitive but is nonspecific because it can cross-react with other spirochetal infections, autoimmune diseases, and certain viruses. To validate a positive ELISA or when the results of the ELISA are equivocal, immunoblot testing can be helpful.[14]

Medical imaging may also help in the diagnosis of LD. Magnetic resonance imaging of children with neurologic manifestations of LD can document abnormal, hyperintense signals on T2-weighted images of the brain.[22] Additionally, ultrasonography of the tail of the parotid gland may show enlargement of the lymph nodes near the stylomastoid foramen in LD patients.[9]

LD may be overdiagnosed or misdiag-

nosed.[18,23] Obviously, diagnosing the specific cause of FNP is critical for definitive care. Although treatment with antibiotics is effective even in late stage disease, early appropriate intervention is the therapeutic goal. In the past, AOM was felt to be the most common cause of FNP. The availability of antibiotic treatment has reduced the frequency of this entity as a cause of FNP. Acute otitis media and chronic otitis media with cholesteatoma have distinct clinical pictures that should prevent them from being confused with LD.

Varicella zoster has a rash and clinical pattern that differs from LD. Herpes zoster presents with a painful vesicular rash that should be distinct from the painless or pruritic annular rash of LD. Other viral syndromes, however, may mimic the "flu-like" constitutional symptoms of early LD. An understanding of the value and limitations of laboratory tests combined with the clinical course of LD can help to differentiate these entities.

Treatment of an isolated FNP in LD is doxycycline, 100 mg twice daily in adults and children over 8 years of age. Amoxicillin 50 mg to 60 mg/kg/d is suggested in younger children. The duration of therapy is usually 4 weeks. In disseminated infection, either with or without positive CNS findings, ceftriaxone should be considered.[17,21]

CONCLUSION

Lyme disease has emerged as the leading cause of FNP in children. A keen awareness of the clinical and laboratory findings with LD will help in the differential diagnosis, particularly for physicians who practice in regions of endemic LD.

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