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Neurological Findings of Lyme Disease

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Neurologic involvement of Lyme disease typically consists of meningitis, cranial neuropathy, and radiculoneuritis, alone or in combination, lasting for months. From 1976 to 1983, we studied 38 patients with Lyme meningitis. Headache and mild neck stiffness, which fluctuated in intensity, and lymphocytic pleocytosis were the common findings. Half of the patients also had facial palsies, which were unilateral in 12 and bilateral in seven. In addition, 12 patients had motor and/or sensory radiculoneuropathies; asymmetric weakness of extremities was the most common finding. Although incomplete presentations of neurologic involvement of Lyme disease may be confused with other entities, the typical constellation of neurologic symptoms represents a unique clinical picture.

Three major types of lesions comprise the neurological manifestations of Lyme disease: meningitis, cranial neuropathy, and radiculoneuritis [1]. These three may occur alone or in combination (Fig. 1). In this report, we describe thirty-eight patients who had meningitis sometimes accompanied by cranial neuropathy and/or peripheral radiculoneuropathy, as reported in depth elsewhere [2]. We believe that this constellation of symptoms is unique among neurological diseases.

The thirty-eight patients were studied at Yale from 1976 through 1983. The ages of the patients ranged from 7 to 64 with a median of 28. All but three patients experienced pre-neurologic symptoms (stage one) consisting of erythema chronicum migrans (ECM), fever, arthralgias, myalgias, malaise, and fatigue [3]. The usual time between the first stage and subsequent meningitis was one month.

The predominant symptom of meningitis was headache, which was present in all but three patients. Its location was usually frontal or occipital, and it characteristically fluctuated in intensity over a period of weeks. On examination, the patients had mild neck stiffness but only on extreme flexion; signs often present in purulent meningitis were absent. Most patients experienced a number of other symptoms, including nausea, vomiting, malaise, fatigue, and irritability. However, physical findings that were common during the first stage (i.e., fever, lymphadenopathy, and ECM) were absent in the majority of patients by the time neurological symptoms were present. The duration of neurological abnormalities varied according to treatment. In the 24 prednisone-treated patients, the neurological stage of their illness lasted for a mean of thirty weeks, with symptoms fluctuating during this period. In the 14 patients treated with high-dose intravenous penicillin most symptoms resolved within ten days [4]. It should be stressed that meningitis may be the first manifestation of Lyme disease. The first sign of illness in

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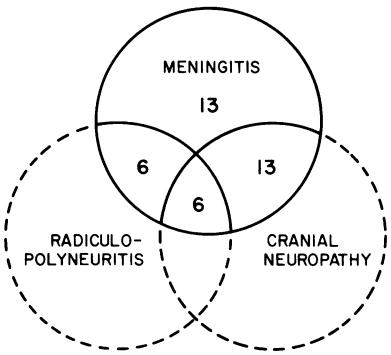

FIG. 1. This Venn diagram illustrates the distribution of findings in Lyme neurological disease. Patients without meningitis, i.e., CSF pleiocytosis, were not included in this study.

one of our early patients was meningitis which recurred three times. The diagnosis became clear only when he developed arthritis.

In addition to meningitis, 11 patients experienced mild encephalitic symptoms—lethargy, difficulty in concentrating, fatigue, emotional lability, irritability, and poor memory. These symptoms also varied from day to day but did not necessarily correlate with the severity of headache and stiff neck. Although a few patients made occasional mistakes in their mental status testing, none had a definite organic brain syndrome, obtundation, or coma. Neither the CT scan nor the EEG was helpful in supporting the diagnosis of encephalitis. The CT scan was normal in all eight patients studied. Nine of the 11 patients with encephalitic symptoms had abnormal electroencephalograms, which showed mild generalized slowing or some sharp activity. However, four patients without such symptoms also had these EEG findings. Although some of the initial patients were thought to have significant encephalitis [1], we now believe that these subtle encephalitic symptoms may have been due to subarachnoid inflammation and not to parenchymal CNS disease.

Except for one patient, other signs of parenchymal CNS disease were absent. This patient had transient bowel and bladder dysfunction and an intermittently positive Babinski on the right. Although these findings suggest the possibility of myelitis, they were not documented well enough for us to be certain whether myelitis is a feature of Lyme disease.

Lumbar puncture was performed on all patients and consistently revealed a lymphocytic pleocytosis with a normal opening pressure. The median CSF white count was 166 with a range of 5–700. The median protein was 79 with a range of 8–400. The CSF glucose was usually normal.

Facial palsies were present in half of the patients; twelve had unilateral involvement and seven, bilateral. The weakness was often preceded or accompanied by a sensation of numbness or tingling on the weak side of the face, but a clear sensory abnormality could not be demonstrated. The facial palsy characteristically occurred at or very near the same time as the headache and meningismus. The weakness was rarely a complete paralysis and usually began improving within weeks of its onset. Residual weakness was usually minimal to none at all. It should be stressed that unilateral or bilateral facial palsy often occurs alone in Lyme disease, without other neurological abnormalities.

Involvement of other cranial nerves was unusual. A few patients had intermittent diplopia without clinically evident extraocular movement palsy. However, one patient had a VI nerve palsy along with an ipsilateral VII.

Twelve patients had radiculoneuritis. Nine patients had symptoms in their extremities, and three had only thoracic sensory radiculopathy. Thoracic radiculopathy appeared concurrently with other neurological symptoms and was experienced as intense pain or pressure within the distribution of a few dermatomes, usually between T8 and T12. On examination, hypoesthesia was present over affected areas in two of the patients and hyperesthesia in one.

The radiculoneuritis in the extremities involved more than one extremity in six of the nine patients. Radicular pain, dysesthesias, and weakness were the most common symptoms. On examination, most patients had focal weakness or loss of reflexes but lacked sensory findings. The duration of symptoms was related to the severity of the lesion. Patients with little or no weakness recovered within days to weeks, but the two patients with severe weakness and atrophy required months to recover. In the two patients with mononeuritis multiplex, different sites became affected days to weeks apart.

In Lyme disease, it seems that peripheral nerve lesions may occur at the root, plexus, or distal nerve. Although this distinction was often difficult to make clinically, electromyography and nerve conduction studies (EMG/NCV) sometimes helped. Of two patients with a clinical picture of mononeuritis multiplex, one had marked slowing of nerve conduction velocity in a number of nerves. In three other patients with clinically diagnosed radiculitis, nerve conduction velocities were relatively spared, but neuropathic abnormalities were noted electromyographically in muscles innervated by one or more roots. A patient with scapular winging had denervation changes in the infraspinatus muscle. One patient was thought to have brachial plexitis and another radiculitis, but their EMG/NCV were normal. These tests were not done in the final patient.

Neurological involvement of Lyme disease may be confused with a number of other disorders. The most common misdiagnosis is viral meningitis, which rarely runs the protracted and relapsing course of Lyme meningitis. Unlike another spirochetal infection, meningovascular syphilis, Lyme disease lacks dramatic parenchymal involvement or basilar meningitis. In some cases of Lyme radiculoneuritis, the Guillain-Barré syndrome has been considered, but in Lyme disease, the nerve involvement is rarely ascending or symmetrical. Finally, other relapsing-remitting diseases like multiple sclerosis sometimes appear in the differential diagnosis, but we have not seen dramatic CNS involvement in Lyme disease.

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