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Case report

Parsonage–Turner syndrome revealing Lyme borreliosis

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Abstract

Parsonage–Turner syndrome, also known as acute brachial neuritis or neuralgic amyotrophy, can be caused by various infectious agents. We report on four patients who experienced Parsonage–Turner syndrome as the first manifestation of Lyme disease. The clinical picture was typical, with acute shoulder pain followed rapidly by weakness and wasting of the shoulder girdle muscles. Electrophysiological testing showed denervation. A single patient reported erythema chronicum migrans after a tick bite. Examination of the cerebrospinal fluid showed lymphocytosis and protein elevation in 3 patients. Serological tests for Lyme disease were positive in the serum in all 4 patients and in the cerebrospinal fluid in 2 patients. Antibiotic therapy ensured a favorable outcome in all 4 cases. Two patients achieved a full recovery within 6 months. Parsonage–Turner syndrome should be added to the list of manifestations of neuroborreliosis. Serological tests for Lyme disease should be performed routinely in patients with Parsonage–Turner syndrome.

Keywords: Parsonage–Turner syndrome; Lyme disease; Borreliosis

Parsonage–Turner syndrome, also known as acute brachial neuritis or neuralgic amyotrophy, was first described in 1948 [1]. Abrupt onset of severe shoulder pain followed by weakness and wasting of several shoulder muscles is the typical clinical picture. The exact cause is unknown, although risk factors are found in more than half the cases [2]. Infectious agents are among the main suggested culprits. We report four cases of Parsonage–Turner syndrome revealing Lyme disease.

1. Methods

We retrospectively reviewed cases of Parsonage–Turner syndrome documented by electrophysiological testing in patients with recent-onset Lyme disease confirmed by serological testing. We recorded the following data for each patient: age, sex, clinical manifestations of Parsonage–Turner syndrome and nerves involved, whether there was a history of a tick bite and/or of erythema chronicum migrans, results of laboratory tests in serum and cerebrospinal fluid including serological tests for *Borrelia burgdorferi*, findings from electroneurophysiological testing and imaging studies, treatments, and outcome. The cases were identified by searching the PubMed database with the indexing terms [Parsonage–Turner syndrome] and [Lyme disease], [borreliosis].

2. Results

We identified four patients, whose main characteristics are reported in Table 1. Three patients lived in eastern France and one in the Paris area near the Fontainebleau forest. There were three men and one woman, whose ages ranged from 38 to 66 years. All 4 patients were frequent hikers. Pain duration ranged from a few hours to 2 months and time from pain onset

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Table 1
Main characteristics of the four patients with Parsonage–Turner syndrome and Lyme disease.

Case	Age	Sex	Muscles involved side time to weakness	Tick bite ECM	Imaging studies	Blood cell counts CRP	CSF	*Borrelia* serology	EMG	Evolution
1	38	M	Deltoid, supraspinatus, infraspinatus, serratus ant., biceps, and triceps, on the right side, 24 h	No	MRI: normal	N	17 lympho/mm^3, Prot: 0.57 g/L	IgM+, IgG+, Wb+, CSF−	Acute denervation	Seroconversion recovered within 6 months
2	45	F	Deltoid, rhomboid, supraspinatus, infraspinatus, biceps, and triceps, left side, 1 week	Yes	MRI: normal	N	N	IgM+, IgG+, Wb+, CSF−	Denervation	Seroconversion improved after 4 months
3	50	M	Deltoid, biceps, triceps, et triceps, palmar interosseous muscle, right side, 1 month	No	NA	N	27 lympho/mm^3, Prot: 0.67 g/L	IgM+, IgG+, Wb+, CSF+	Denervation on both sides	Seroconversion improved after 4 months
4	66	M	Trapezius, deltoid, supraspinatus, infraspinatus, serratus ant., biceps, brachial ant, both sides, 1 month	No	CT: osteoarthritis	12,000 WBC/mm^3, CRP N	299 lympho/mm^3, Prot: 1.08 g/L	IgM+, IgG+, Wb+, CSF+	Denervation on both sides	Seroconversion recovered within 3 months

NA, not available; ECM, erythema chronicum migrans; MRI, magnetic resonance imaging; CT, computed tomography; CSF, cerebrospinal fluid; Wb, Western blot; EMG, electromyogram.

to muscle weakness ranged from 24 h to 1 month. Both shoulders were affected in 1 patient. Various shoulder–girdle muscles were involved, as well as arm muscles in some of the patients. Wasting developed within 1 month after the painful phase. A single patient reported a tick bite followed by erythema chronicum migrans on the same arm 2 months before the onset of the pain. She was not treated at the time.

Findings were normal from routine laboratory tests, including C-reactive protein. Blood cell counts showed mild lymphocytosis in 1 patient and normal results in the other 3 patients. Electrophysiological testing consistently showed acute denervation of the proximal upper limb muscles, usually in a distribution that correlated with the clinical findings, although 1 patient with unilateral symptoms had bilateral denervation. The cerebrospinal fluid was abnormal in 3 patients, with lymphocytosis (17–299/mm^3) and protein elevation (0.57–1.08 g/L). IgM antibodies to *B. burgdorferi* were detected in sera from all 4 patients. Western blot results confirmed this finding. Changes over time were consistent with recent-onset infection (Table 2). *B. burgdorferi* antibodies were found in the cerebrospinal fluid in 2 patients. Serological tests for the cytomegalovirus, herpes simplex virus, and Epstein Barr virus were performed in 3 patients and were consistently negative. Two patients underwent magnetic resonance imaging (cervical spine and brachial plexus, respectively), which showed no evidence of cervical epidural disease or signal changes from the brachial plexus. MRI of the shoulder muscles was not performed. Computed tomography of the cervical spine was performed in 1 patient and showed degenerative disease with no other abnormalities.

Injectable ceftriaxone was given in a dosage of 2 g/day for 21 days. The antibody titers changed over time in a pattern consistent with recent-onset infection. The pain resolved fully. Two patients recovered full muscle function, within 3 months and 6 months, respectively. Muscle function improved in the other 2 patients over a follow-up period of 1 year. None of the patients experienced recurrences.

3. Discussion

Our four patients had Parsonage–Turner syndrome associated with recent-onset Lyme disease documented by serological tests. Peripheral neurological involvement is a well-documented manifestation of the second phase of Lyme

Table 2
Serological tests for *Borrelia burgdorferi* over time in the four patients.

Case	Serum at diagnosis	CSF at diagnosis	Serum on day 15	Serum on day 40
1	ELISA: ($N \leq 1$, 1), IgM: 130 U/L, IgM: 0.7 U/L, Western blot: positive	Negative	ELISA: IgM: 43, IgG: 54	ELISA: IgM negative
2	ELISA: ($N \leq 160$), IgM: 64 U/L, IgG: 512 U/L, Western blot: positive	Negative	ELISA: IgM: negative, IgG: >180 U/L	
3	Immunochromatography, IgM+, IgG+, Western blot: positive	ELISA: IgM: 0.5, IgG: 51, PCR: negative	ELISA: IgG: 151 U/L, IgM: 0.6 U/L	ELISA: IgG: 249 U/L, IgM: 0.2 U/L
4	ELISA: IgM: positive, IgG: 7 U/L, Western blot: positive	ELISA: ($N < 1$), IgM: 2.25, IgG: 8.45	ELISA: IgM: negative, IgG: 50	

Western blots were considered positive when they showed at least two IgM bands and three IgG bands. CSF: cerebrospinal fluid.

disease. Neurological manifestations occur in 8–46% of cases of Lyme disease [4]. Meningoradiculitis is the most common neurological manifestation, accounting for 85% of cases of neuroborreliosis [4]. Diagnostic criteria for neuroborreliosis have been developed in Europe and the US [4]. Lymphocytic meningitis, cranial nerve involvement, radiculopathy, and meningoradiculitis are among the clinical criteria. In the American criteria set, one of the following is required: demonstration of *B. burgdorferi* in a tissue or cerebrospinal fluid specimen, IgM or IgG antibodies to *B. burgdorferi* in the serum or cerebrospinal fluid, or a significant change in antibody titers. Cerebrospinal fluid lymphocytosis and intrathecal production of specific antibodies are required by the European criteria set. Our patients met these criteria. In addition to meningoradiculitis, which is the most common neurological manifestation, other acute neurological syndromes have been reported, including meningitis, isolated involvement of a cranial or spinal root, acute myelitis, and acute encephalitis [4]. The other neurological manifestations of Lyme disease occur at the third phase and run a chronic course. They include chronic encephalomyelitis, neuropathies, and polyradiculoneuropathy. The link with Lyme disease is controversial for a number of manifestations (encephalopathies, psychiatric disorders, amyotrophic lateral sclerosis, multiple sclerosis, and cerebrovascular accidents). Parsonage–Turner has rarely been reported as a peripheral neurological manifestation of Lyme disease [5].

Parsonage–Turner syndrome is a disease of multiple nerve trunks that predominantly involves the brachial plexus. The annual incidence is 2–3/100,000 [2,3]. Risk factors that may play a triggering role are identified in 30–80% of cases. In a review of 246 patients, a possible cause was identified in 53% of cases [2]. In 43.5% of cases, the suspected cause was an infection [2]. Many potential infectious causes have been reported: tuberculosis, typhoid fever, yersiniosis, leptospirosis, smallpox, mumps, cytomegalovirus infection [6] Epstein–Barr virus infection [7], parvovirus B19 infection [8], and HIV infection [9]. Herpes viruses and the Epstein–Barr virus may lead to cross-reactivity for IgM antibodies against *B. burgdorferi*; in this situation, IgG antibodies by ELISA and Western blot results are negative for *B. burgdorferi* [10].

Lyme disease has rarely been reported as a cause of Parsonage–Turner syndrome. We found five previously reported cases, all of which were published more than a decade ago [11–14]. Four occurred in France and one in Japan. There were 3 men and 2 women aged 28–70 years. Two patients had involvement of both shoulders. The pain lasted for a few days to 3 months. None of the patients reported erythema chronicum migrans. Serological tests for Lyme disease were positive in all 5 patients. Electrophysiological testing was consistently abnormal. Cerebrospinal fluid abnormalities were found in 3 patients. Antibiotic therapy ensured a favorable outcome within 2–12 months. These characteristics are similar to those of our 4 patients. Given the spontaneously favorable outcome of Parsonage–Turner syndrome within 1–2 years [2,3], Lyme disease therapy is unnecessary in the absence of suggestive symptoms such as erythema chronicum migrans (which was noted in only 1 of 9 cases).

The contribution of Lyme disease to the occurrence of Parsonage–Turner syndrome is probably underevaluated, since tests are not done routinely. Serological testing is the main diagnostic tool. However, cerebrospinal fluid is normal in idiopathic Parsonage–Turner syndrome, and lymphocytosis with protein elevation suggests Lyme disease [3]. However, our findings suggest that cerebrospinal fluid testing for antibodies may have a lower yield than serum testing. A local inflammatory process is suspected in Parsonage–Turner syndrome [2,3]. Neuroborreliosis is related to inflammation induced by the microorganism [15,16]. Thus, a causative role for Lyme disease in Parsonage–Turner syndrome is biologically plausible.

In sum, these data indicate that Parsonage–Turner syndrome should be added to the list of neurological manifestations of Lyme disease. Serological tests for Lyme disease should be performed in patients with Parsonage–Turner syndrome, most notably those living in areas of high endemicity of Lyme disease. Although Parsonage–Turner syndrome resolves spontaneously, patients with positive serological tests should receive treatment to prevent the development of other neurological complications related to Lyme disease.

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