EMG showed complete axonotmesic denervation of the right hemifacial muscles. MRI with contrast showed enhancement in the intratemporal part of the right facial nerve and disappearance of the previous alteration on the left (fig. 1b). Intravenous dexamethasone (8 mg for 5 days; 4 mg for 3 days) was given, during which time the patient reported improved facial mobility and disappearance of local tenderness.

Discussion

Differential diagnosis of our case considered Crohn's disease, sarcoidosis and Bell's palsy [7, 13–16]. There were no signs of intestinal disease and oral examination showed no ulceration or mucosal cobblestoning. We excluded sarcoidosis from the negative chest X-ray and normal ACE in serum and CSF.

The oligoclonal banding in our patient suggests abnormal Ig production by lymphocyte clones within the CNS, since neither serum Ig nor blood-brain barrier alterations were present (low CSF albumin). IgG synthesis in CSF was reported in 3/56 cases of peripheral facial palsy, but 2 had elevated Borrelia antibodies [16].

Transient alteration along the facial nerve, detected by MRI, has been described previously in MRS [17], while another patient with MRS had an increased level of CSF IgG as well as oligoclonal banding [18]. Our patient is the first in whom these two alterations are associated, and although we have no proof of a causal relation between them, in the absence of family history, collagenopathy and sarcoidosis, they suggest a primary immune disorder of the CNS as a possible cause of MRS.

References

1 Sussman GL, Yang WH, Steinberg S: Melkersson-Rosenthal syndrome: Clinical, pathologic, and therapeutic considerations. Ann Allergy 1992;69:187–193.
2 Cohen HA, Cohen Z, Ashkenasi A, et al: Melkersson-Rosenthal syndrome. Cutis 1994;54:327–328.
3 Pearce JM: Melkersson's syndrome. J Neurol Neurosurg Psychiatry 1995;58:340.
4 Meisel-Stosiek M, Hornstein OP, Stosiek N: Family study on Melkersson-Rosenthal syndrome. Some hereditary aspects of the disease and review of literature. Acta Derm Venereol 1990;70:221–226.
5 Cleary KR, Batsakis JG: Orofacial granulomatosis and Crohn's disease. Ann Otol Rhinol Laryngol 1996;105:166–167.
6 Zimmer WM, Rogers PS, Reeve CM, Sheridan PJ: Orofacial manifestations of Melkersson-Rosenthal syndrome: A study of 42 patients and review of 220 cases from the literature. Oral Surg Oral Med Oral Pathol 1992;76:610–619.
7 Balevi B: Melkersson-Rosenthal syndrome: Review of the literature and case report of a 10-year misdiagnosis. Quintessence Int 1997;28:265–269.
8 Wadlington WB, Riley HD Jr, Lowbeer L: The Melkersson-Rosenthal syndrome. Pediatrics 1984;73:502–506.
9 Winnie R, Deluxe DM: Melkersson-Rosenthal syndrome: Review of literature and case report. Int J Oral Maxillofac Surg 1992;21:115–117.
10 Levenson MJ, Ingerman M, Grimes C, Anand KV: Melkersson-Rosenthal syndrome. Arch Otolaryngol 1984;110:540–542.
11 Orlando MR, Atkins JS: Melkersson-Rosenthal syndrome. Arch Otolaryngol Head Neck Surg 1990;116:728–729.
12 Levy FS, Bircher AJ, Buchner SA: Delayed-type hypersensitivity to cow's milk protein in Melkersson-Rosenthal syndrome: Coincidence or pathogenetic role? Dermatology 1996;192:99–102.
13 Misra S, Ament ME: Orofacial lesions in Crohn's disease. Am J Gastroenterol 1996;91:1651–1653.
14 De Aloe G, Rubegni P, Mazzatenta C, Figiani M: Complete Melkersson-Rosenthal syndrome in a patient with Crohn's disease. Dermatology 1997;195:182.
15 Kohout J, Schober W: Zur Sarkoidose des Zentralnervensystems. Nervenarzt 1974;45:538–543.
16 Roberg M, Ernerudh J, Forsberg P, Fridell E, Fryden A, Hyden D, Linde A, Odkvist L: Acute peripheral facial palsy: CSF findings and etiology. Acta Neurol Scand 1991;83:55–60
17 Ferriby D, Pertuzon B, Clarisse J, Vermersch P: Magnetic resonance imaging of the facial nerve in a case of Melkersson-Rosenthal syndrome. Rev Neurol 1998;426–428.
18 Durelli L, Cocito D, Delsedime M: The Melkersson-Rosenthal syndrome: A case with increased CNS IgG synthesis. Ann Neurol 1985;18:623.

Gennaro Bussone, MD, C. Besta National Neurological Institute
Via Celoria 11, IT–20133 Milano (Italy)
Tel. +39 02 2394264, Fax +39 02 70638067
E-Mail bussone@istituto-besta.it

Eur Neurol 2004;51:43–45
DOI: 10.1159/000075086

Lyme Neuroborreliosis Mimicking Primary CNS Lymphoma

Ernst U. Walther[a], *Klaus Seelos*[b], *Karl Bise*[c], *Michael Mayer*[a], *Andreas Straube*[a]

[a] Department of Neurology, [b] Department of Neuroradiology and [c] Institute of Neuropathology, Klinikum Grosshadern, Munich, Germany

Case Report

In October 1994, a 25-year-old woman complained of 4 days of progressive fatigue, retro-orbital headaches, nausea, and intermittent vomiting. Frontal sinusitis was suspected and oral amoxicillin was administered for 4 days. Despite therapy, the patient became somnolent and was referred to hospital. MRI scanning showed multiple space-occupying lesions in the deep white matter with concomitant edema adjacent to both frontal horns (fig. 1). After steroid treatment, the patient's condition improved markedly. Eighteen days after onset of the initial symptoms she was referred to our clinic for further diagnostic evaluation of suspected multilocular CNS lymphoma.

On examination, she had papilledema bilaterally, and cogwheel eye pursuit. Tendon reflexes were more pronounced to the right, but all other findings were normal. The patient's past medical history was normal and she took no medication except birth control pills. Postcontrast MRI scans showed gadolinium enhancement of all lesions (data not shown).

Somatosensory and visually evoked potentials as well as central motor conduction velocities were normal. CSF leukocytes were mildly increased (8/μl; 57% lymphocytes, 39% monocytes, 2% macrophages and 2% granulocytes). No lymphoma cells were found. CSF glucose was normal. CSF protein was 44 mg/dl. Isoelectric focusing showed oligoclonal bands in the CSF. Antibodies to *Borrelia burgdorferi* were determined using an indirect immunofluorescence test after absorption with *Treponema phagedenis,* as well as ELISA. IgG and IgM titers ≥ 1:64 in serum and ≥ 1:4 in CSF are considered significantly elevated. In comparison, the patient had a serum IgG titer

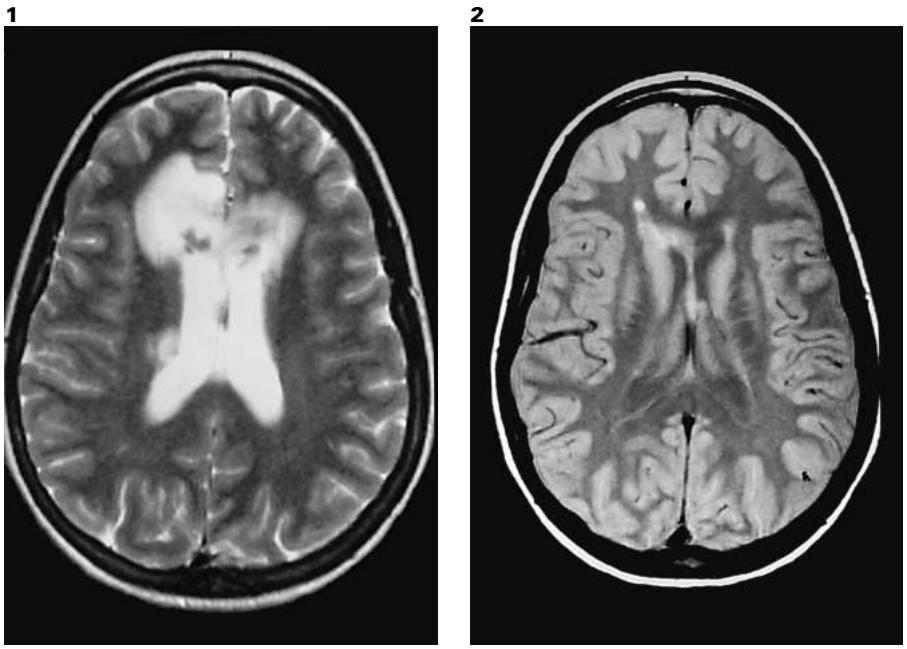

Fig. 1. T_2-weighted transverse MRI shows multiple space-occupying lesions in the deep white matter, adjacent to lateral ventricles.
Fig. 2. Proton-density-weighted transverse MRI shows no substantial residual edema in the frontal deep white matter.

1:2048, serum IgM 1:64, CSF IgG 1:64, and CSF IgM < 1:2. Intrathecal production of antibodies to *B. burgdorferi* was determined as published previously [1]. Briefly, the CSF/serum ratio of ELISA specific anti-*B. burgdorferi* IgG values was compared with the CSF/serum ratio of total IgG (CSF/serum index). The CSF/serum index in the patient was 3.0, indicating specific intrathecal antibody production in the CSF. Tests for syphilis, sarcoidosis, and HIV were negative. The patient underwent stereotaxic biopsy of the lesion in the right frontal white matter to exclude a CNS lymphoma because of pretreatment with corticosteroids. Histopathological examination showed an active inflammatory process with diffuse infiltration of granulocytes and monocytes, reactive gliosis, and petechial erythrocyte extravasation, consistent with Lyme neuroborreliosis. Direct microscopic or cultural proof of *B. burgdorferi* or other infectious agents failed. PCR was not performed. On the basis of the serological results, active Lyme disease was diagnosed. Since further microbiological analyses were pending at the time, intravenous amoxicillin was administered for 14 days to cover a potential *Listeria* infection. (*Listeria* serology and PCR results proved negative.) An MRI scan directly after discontinuation of therapy revealed no residual edema (fig. 2), but minor focal gadolinium enhancement at the biopsy site was noted, possibly related to mild gliosis (data not shown). A second examination 6 months later detected no gadolinium enhancement. Currently, the patient is in good health and not on treatment.

Discussion

Neurological manifestations of Lyme disease include meningitis, encephalitis, psychiatric disorders, cranial neuritis, polyradiculitis, and peripheral neuropathy [2]. The pathogenesis is not known, but both vasculopathy and direct brain invasion are possible mechanisms. Our patient provides an unusual example of a curable cerebral mass lesion. Papilledema is an uncommon manifestation of neuroborreliosis and usually not due to elevated intracranial pressure [3]. Space-occupying lesions and pseudotumor cerebri are extremely rare and have been described only in children up to 15 years of age [4]. Brain CT and MRI findings in patients with Lyme disease are usually normal, but abnormalities may involve periventricular lesions simulating multiple sclerosis [5, 6], hydrocephalus [7], and thalamic and basal ganglia lesions [5, 8]. However, space-occupying, contrast-enhancing lesions in contact with the subarachnoid space, predominantly in the cerebral hemispheres, basal ganglia, and corpus callosum, are characteristic of primary CNS lymphoma [9]. Corticosteroids and antibiotic therapy directed against *B. burgdorferi* led to complete remission of elevated intracranial pressure, as well as resolution of the inflammatory lesions on MRI scans. To our knowledge, this is the first case report of an adult patient with neuroborreliosis mimicking CNS lymphoma in cranial MRI. Brain imaging has not been used in previously published cases with abnormal CSF cytology [10]. Normal evoked potential studies on presentation and the favorable long-term clinical course of the disease are strong arguments against multiple sclerosis. We are not aware of any case of acute disseminated encephalomyelitis associated with *B. burgdorferi* infection; however, the clinical syndrome of our patient meets the criteria. As the infection can be cured, MRI may play an important role in making the diagnosis and differentiating Lyme neuroborreliosis from other diseases of the white matter.

Acknowledgment

We thank our colleagues at the Max von Pettenkofer Institute for Microbiology and Hygiene (Dr. B. Wilske) and at the Institute of Clinical Chemistry (Dr. M. Wick) for their CSF and serological analyses.

References

1 Wilske B, Schierz G, Preac-Mursic V, von Busch K, Kühbeck R, Pfister HW, Einhäupl K: Intrathecal production of specific antibodies against *Borrelia burgdorferi* in patients with lymphocytic meningoradiculitis (Bannwarth's syndrome). J Infect Dis 1986;153:304–314.
2 Halperin JJ, Luft BJ, Anand AK, Roque CT, Alvarez O, Volkman DJ, Dattwyler RJ: Lyme neuroborreliosis: Central nervous system manifestations. Neurology 1989;39:753–759.
3 Berglöff J, Gasser R, Feigl B: Ophthalmic manifestations in Lyme borreliosis. A review. J Neuroophthalmol 1994;14:15–20.
4 Kan L, Sood SK, Maytal J: Pseudotumor cerebri in Lyme disease: A case report and literature review. Pediatr Neurol 1998;18:439–441.
5 Finkel MF: Lyme disease and its neurologic complications. Arch Neurol 1988;45:99–104.
6 Pachner AR, Duray P, Steere AC: Central nervous system manifestations of Lyme disease. Arch Neurol 1989;46:790–796.
7 Danek A, Uttner I, Yousry T, Pfister HW: Lyme neuroborreliosis disguised as normal pressure hydrocephalus. Neurology 1996;46:1743–1745.
8 Wokke JHJ, vanGijn J, Elderson A, Stanek G: Chronic forms of *Borrelia burgdorferi* infection of the nervous system. Neurology 1987;37:1031–1034.
9 Bühring U, Herrlinger U, Krings T, Thiex R, Weller M, Küker W: MRI features of primary central nervous system lymphomas at presentation. Neurology 2001;57:393–396.
10 Szyfelbein WM, Ross JS: Lyme disease meningopolyneuritis simulating malignant lymphoma. Mod Pathol 1988;1:464–468.

Ernst U. Walther, MD, Department of Neurology, Klinikum Grosshadern
Marchioninistrasse 15, DE–81377 München (Germany)
Tel. +49 89 7095 3904, Fax +49 89 7095 3677
E-Mail ewalther@nefo.med.uni-muenchen.de

Table 1. Nerve conduction studies

Nerves	MCV m/s	CMAP, mV (distal/prox.)	d.l. ms	SCV m/s	SNAP μV	FCV m/s
Rt. median	[a]	1.0/n.e.	4.8	n.e.	n.e.	n.e.
Lt. median	[a]	1.1/n.e.	5.3	33.0	2.0	n.e.
Rt. ulnar	[a]	1.4/n.e.	3.4	n.e.	n.e.	n.e.
Lt. ulnar	[a]	1.0/n.e.	5.0	n.e.	n.e.	n.e.
Rt. PTN	35.0	2.6/1.9	5.0			39.6
Lt. PTN	30.8	2.6/2.2	8.2			40.4
Rt. DPN	41.0	1.8/1.7	5.5			
Lt. DPN	n.e.	n.e.	n.e.			
Rt. sural				n.e.	n.e.	
Lt. sural				42.6	5.0	

Rt = Right; Lt = left; PTN = posterior tibial nerve; DPN = deep peroneal nerve; MCV = motor conduction velocity; CMAP = compound motor action potential; d.l. = distal latency; SCV = sensory conduction velocity; SNAP = sensory nerve action potential; FCV = F wave conduction velocity; n.e. = not evoked.

[a] MCV could not be calculated due to complete conduction block between elbow and wrist.

Eur Neurol 2004;51:45–46
DOI: 10.1159/000075087

Bilateral Hypoglossal Nerve Involvement in Chronic Inflammatory Demyelinating Polyneuropathy

Kazuo Yamashiro[a], Yasuyuki Okuma[a], Kenji Fujishima[a], Keigo Goto[a], Yoshikuni Mizuno[b]

[a] Department of Neurology, Juntendo University Izu-Nagaoka Hospital, Shizuoka, and [b] Department of Neurology, Juntendo University School of Medicine, Tokyo, Japan

Although cranial nerves are occasionally affected in chronic inflammatory demyelinating polyneuropathy (CIDP), involvement of the bulbar muscles is uncommon [1–6]. Here we report a patient with CIDP who presented with weakness of the distal upper extremities and atrophy of the tongue. The features mimicked those of motor neuron disease. CIDP should be considered in patients who present with bulbar palsy because it is treatable.

Case Report

A 49-year-old man developed a slowly progressive weakness of the left hand followed by weakness of the right hand. Two years later, he noted speech disturbance, and another year later, he noted difficulty in swallowing. He was diagnosed as having motor neuron disease in our outpatient clinic because he showed atrophy of the tongue. At age 52, he was admitted to our hospital for further investigation. Neurologic examination revealed bilateral facial weakness, absence of the gag reflex, poor palatal elevation, and moderate atrophy and paresis of the tongue bilaterally without fasciculations. Moderate dysphagia and dysarthria were also noted. He showed a dropped wrist on the left side. The Medical Research Council (MRC) scale grade for the left forearm extensors was 1, and that for the right was 3, with bilateral wasting and weakness of the intrinsic hand muscles (MRC grades 1–2). There was mild impairment of pinprick and light-touch sensation over the median and the ulnar nerve territories on both sides. Muscle power in the lower extremities was preserved. All tendon reflexes were absent.

Nerve conduction studies (table 1) showed slowing of nerve conduction velocities and reduced sensory and motor action potential amplitude. Complete conduction block was seen between elbow and wrist in the median and ulnar motor nerves on both sides. The distal latency for the facial nerve was prolonged (6.3 ms, normal <4.0 ms. Needle electromyography revealed a reduced interference with long-duration polyphasic high-amplitude motor unit potentials in the tongue, sternocleidomastoid muscle, biceps brachii muscle, and the first dorsal interosseus muscle, indicating chronic partial denervation and reinnervation. No abnormal potentials were seen at rest. Cerebrospinal fluid studies showed an elevated protein content (76 mg/dl) with a normal white blood cell count. Magnetic resonance images of the brain and the spinal cord were unremarkable. Other laboratory studies including routine chemistries, immunoelectrophoresis, thyroid function tests, and tests for antinuclear antigen, rheumatoid factor, and angiotensin-converting enzyme revealed normal findings.

He was diagnosed as having CIDP, and was treated with high-dose intravenous immunoglobulin at 0.4 g/kg/day for 5 days. Subsequently, he received 60 mg of oral prednisolone for 1 month, after