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Lyme disease in a child presenting with bilateral facial nerve palsy: MRI findings and review of the literature

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Abstract We report a 7-year-old boy with neuroborreliosis presenting with headache and bilateral facial nerve palsy. MRI demonstrated tentorial and bilateral facial and trigeminal nerve enhancement.

Key words Lyme disease · Magnetic resonance imaging

Case report

A 7-year-old boy with no relevant previous medical history was admitted because of bilateral peripheral facial nerve palsies. For one week, he had felt unwell, lost his appetite, became lethargic and complained of severe headache. He also had bilateral conjunctivitis. The initial presumptive diagnosis, before the appearance of the facial nerve palsies, was of heat stroke, since at that time we had a heat wave. On admission, the child had no fever and showed normal strength in the limbs. No other cranial nerve palsy was found. There was no papilloedema. The electroencephalogram was normal

MRI at 1.0 T with 5-mm axial and 4-mm-thick sagittal T1 and T2-weighted images was normal. After intravenous administration of 0.1 mmol/kg gadodiamide, enhancement of both trigeminal nerves, the right side of the tentorium and both facial nerves in the internal auditory canal was seen (Fig. 1). No parenchymal lesions were evident.

Because his mother remembered removing a 'splinter' the size of a pin-head from his leg 1 month previously, during a holiday in the south of Belgium, a tentative diagnosis of borreliosis was made. Serum and cerebrospinal fluid (CSF) were examined for *Borrelia burgdorferi* antibodies. The boy was treated intravenously with ceftriaxone for a 2-week period before the results of the serology. During the first few days starting antibiotic therapy he developed minimal weakness of both hands, and arthralgia in the right knee. Transient erythema of both cheeks also appeared. The patient slowly recovered from his bilateral facial nerve paralysis. The serum showed no evidence of infection. CSF examination showed an elevated protein content (61 mg/ml), while the white blood cell count (3 lymphocytes/microlitre) and glucose concentration (0.62 g/dl) were normal. Antigen detection (polymerase chain reaction amplification) for *B. burgdorferi* in the CSF was negative. IgM ELISA (enzyme-linked immunosorbent assay) was positive, IgG ELISA negative for *B. burgdorferi* in the serum. Epstein-Barr virus serology showed immunity and Herpes simplex serology was negative. Western blot confirmed the positive IgM serum findings for *B. burgdorferi.*

Discussion

Lyme disease is a tick-transmitted spirochaetal illness caused by *B. burgdorferi.* Its worldwide distribution correlated with the geographic ranges of certain ixodid ticks. *Ixodes damnini* and *Ixodes pacificus* are the principal vectors in the USA, whereas *Ixodes ricinus* and *Ixodes persulcatus* are the vectors throughout Europe and Asia, respectively. Central nervous system (CNS) presentations occur in 10–15 % of cases; 23 % of reported cases were in children and adolescents [1, 2].

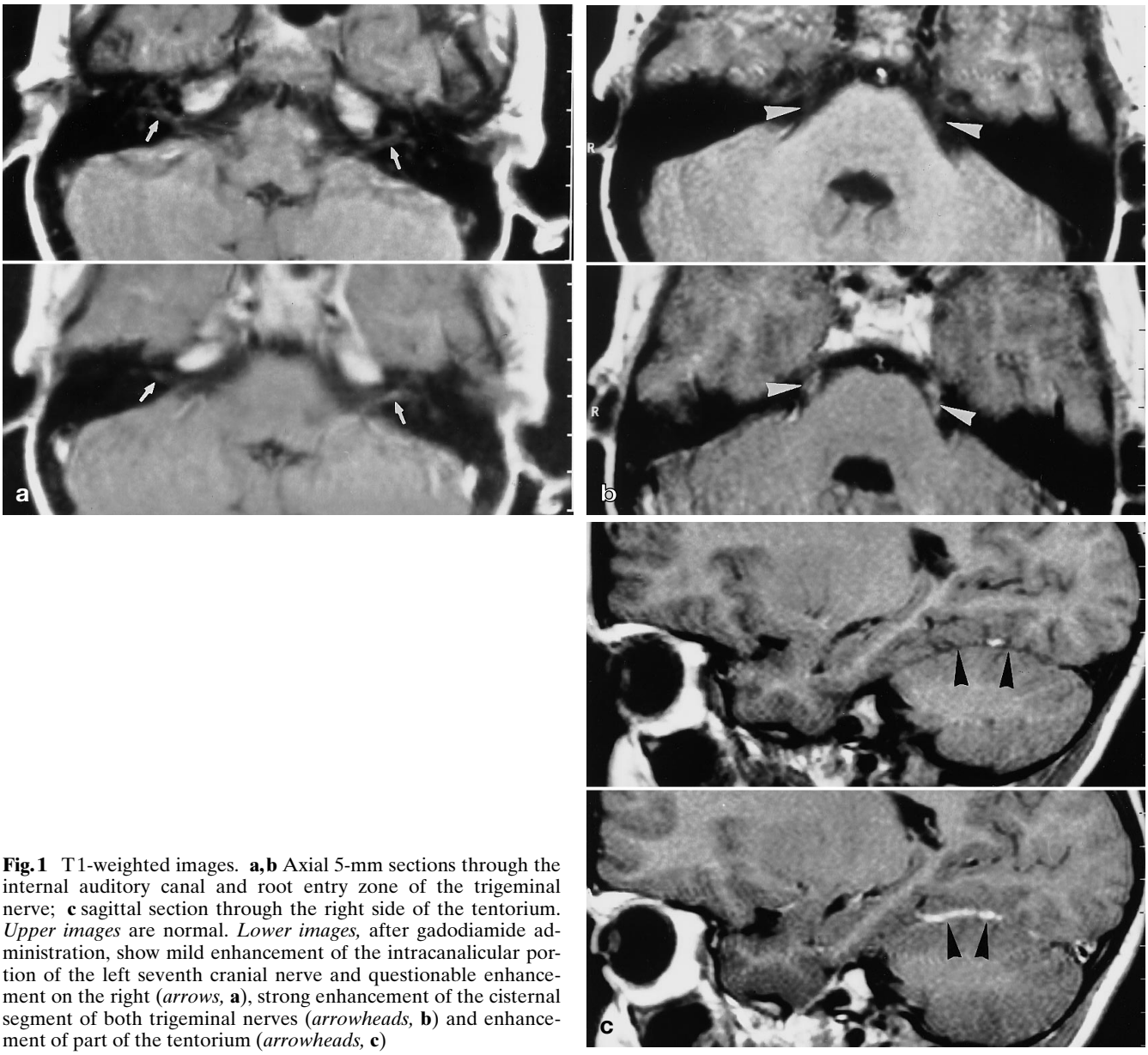

Fig. 1 T1-weighted images. **a, b** Axial 5-mm sections through the internal auditory canal and root entry zone of the trigeminal nerve; **c** sagittal section through the right side of the tentorium. *Upper images* are normal. *Lower images,* after gadodiamide administration, show mild enhancement of the intracanalicular portion of the left seventh cranial nerve and questionable enhancement on the right (*arrows,* **a**), strong enhancement of the cisternal segment of both trigeminal nerves (*arrowheads,* **b**) and enhancement of part of the tentorium (*arrowheads,* **c**)

As with other spirochaetal infections, the disease occurs in stages with remissions and exacerbations and different clinical manifestations at each stage. Incubation varies from 3 to 32 days, after which a characteristic expanding skin lesion presents, accompanied by 'flu-like' or 'meningitis-like' symptoms (stage 1). After several weeks to months, neurological abnormalities and cardiac involvement are seen in 15% and 8% of patients, respectively. Migrating musculoskeletal pain is common during this phase (stage 2). Within weeks or up to 2 years of the onset of infection, about 60% of the patients develop arthritis (stage 3).

Neurological manifestations include meningitis, encephalitis, cranial neuritis – including unilateral or bilateral facial palsy – motor and sensory radiculoneuritis, mononeuritis multiplex, chorea and myelitis. The usual pattern consists of fluctuating symptoms of meningitis accompanied by facial nerve palsy and peripheral radiculoneuropathy [1]. In 96 children reported by Belman et al. [3] the most frequent neurological symptom was headache and the most frequent neurological sign facial palsy. Less common manifestations in children were sleep disturbances and papilloedema, with increased intracranial pressure. The latter seems to be unique to North American paediatric Lyme disease. Peripheral

neuropathy and meningoradiculitis, in contrast with adults, were rare [3].

The diagnosis is made by recognition of a characteristic clinical picture with serological confirmation [1]. With CNS involvement, local intrathecal production of specific antibody appears to be a frequent, consistent finding, in contrast with peripheral nervous system disease [4].

The treatment of neurological abnormalities is intravenous antibiotics (ceftriaxone, penicillin G) and steroids. Other manifestations can be treated with oral antibiotics (doxycycline, amoxicillin, cefuroxime, erythromycin). About 15% of patients experience a Herxheimer-like reaction during the first 24 h of therapy, presenting as fever, chills, myalgia, headache, tachycardia, tachypnoea and mild vasodilatation with mild hypotension. An increased circulating neutrophil count is found [1]. The weakness of both hands, arthralgia and erythema in our patient could be side effects of the antibiotics or represent a mild Herxheimer reaction.

Several reports have described lesions on MRI in the cerebral white matter in Lyme disease, showing as focal areas of high signal on T2-weighted images [4, 5]. Fernández et al. [6] reported brain stem and white matter lesions in six of 14 patients [6]. The lesions can simulate tumours, demyelinating disease, acute disseminated encephalomyelitis, vasculitis and ischaemic lesions [5, 7–13, 14]. We did not see abnormal signal on T2-weighted images.

Few reports mention contrast-enhancing lesions. Third, fifth and seventh cranial nerve enhancement has been reported [15, 16]. Of three patients reported by Demaerel et al. [9] two showed pial enhancement along the brain stem, extending along the spinal cord in one; the third showed peripheral enhancement of white-matter lesions. Nelson et al. [16] demonstrated tentorial enhancement [16]. Rafto et al. [17] described interpeduncular leptomeningeal enhancement [17]. Murphy [18] investigated 11 patients with facial paralysis, including one with Lyme disease, finding a poorer prognosis for complete return of function when enhancement of the mastoid segment of the facial nerve was seen. Patients with enhancement of the labyrinthine segment, geniculate ganglion or proximal tympanic portion of the nerve, including the patient with Lyme disease, showed complete return of function. In our case, enhancement of the intracanalicular portion was present on the left and questionable on the right. The bilateral facial paralysis recovered, but later on the left, the side with more pronounced contrast enhancement. The enhancement of both trigeminal nerves at the root entry zone was not expressed clinically.

Reports of MRI findings in the CNS in children and adolescents with Lyme disease are rare. Belman et al. [19] reported eight children, two of whom showed multiple foci of increased signal on T2-weighted images [19]. Other papers describe single cases or limited series [4, 6–9, 11, 15, 20]. Right third cranial nerve enhancement in a child with recurrent oculomotor palsy [15], presumed demyelinating lesions in the pons and cerebrum [8, 20], pial enhancement and white matter lesions with peripheral enhancement [9], ischaemic foci [11], tumour-like lesions [7], and high-signal brain stem and white matter lesions on T2-weighted images have also been described [4, 6, 9].

Kruger et al. [5] reported follow-up of 27 treated patients, but only three underwent MRI at the moment of diagnosis; they showed unchanged multiple sclerosis-like lesions on MRI 2–4 years after treatment. Six patients without clinical CNS involvement at the time of diagnosis and positive MRI at follow-up were found to have smaller, less numerous white matter lesions than 11 patients with positive MRI at follow-up and initial CNS symptoms. In these latter patients additional lesions in the basal ganglia, and capsular regions were found [5]. Halperin et al. [4] repeated MRI in six of seven patients with high-signal lesions on T2-weighted images at the time of diagnosis; mean follow-up time was 5.3 months. In three patients the lesions disappeared [4].

MRI appears to be the examination of choice to assess the response to antibiotic therapy. Contrast enhancement disappeared in two patients reported by Demaerel et al. [9] who had follow-up. White matter lesions, initially enhancing in one of these patients, persisted on the T2-weighted images but enhancement was not seen 103 days after the onset of symptoms. Pial enhancement along the brain stem and unenhancing white matter lesions in the other patient had disappeared completely 86 days after the onset of symptoms. Both patients recovered fully. Oksi et al. [7] found progression of unenhancing white-matter lesions and atrophy 1 year after initial MRI in a patient who later died of chronic relapsing Lyme disease. In two other patients, with contrast-enhancing tumour-like lesions, the lesions disappeared. Both underwent craniotomy for suspected malignancy. One had a normal MRI 5 months after antibiotic therapy and surgical intervention; the other showed multifocal lesions which disappeared after repeated antibiotic therapy during 2 years of follow-up [7]. No follow-up MRI examination was performed in our patient because of his favourable progress.

Different mechanisms of involvement of the central nervous system have been suggested, such as direct spirochaetal invasion [7, 21], vasculitis [4, 5, 7, 10] or an immune-mediated process [22]. Debate continues.

Meningeal enhancement and multiple cranial nerve enhancement, as in our patient, were reported only by Nelson et al. [16] and Demaerel et al. [9]. Recognising these subtle imaging findings, combined with high clinical suspicion, is the key to diagnosis and allows early treatment.

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