Intracranial hypertension in neuroborreliosis

Case report

Christoph Härtel*;
Stefan Schilling, Department of Paediatrics;
Birte Neppert, Department of Ophthalmology;
Bettina Tiemer, Institute of Medical Microbiology;
Jürgen Sperner, Department of Paediatrics, University of Lübeck Medical School, Lübeck, Germany.

**Correspondence to first author at* Department of Paediatrics, University of Lübeck Medical School, Ratzeburger Allee 160 23538 Lübeck, Germany.
E-mail: haertel@paedia.ukl.mu-luebeck.de

Neuroborreliosis is an infection of the nervous system caused by the spirochete Borrelia burgdorferi, from which patients most commonly develop lymphocytic meningitis, radiculoneuritis, or cranial neuropathy. In this report a 9-year-old male with an unusual neurological complication of neuroborreliosis – benign intracranial hypertension (BIH) – is described. Clinical symptoms of BIH, which consist of increased CSF pressure in the absence of an intracranial mass or obstruction to the circulation of CSF, resolved completely after antibiotic therapy with ceftriaxone.

Benign intracranial hypertension (BIH) is a clinical entity characterized by: (1) increased intracranial pressure, (2) papilledema and visual disturbances, and (3) no pathological findings in cranial CT or MRI. Various aetiological factors have been associated with BIH in children including obesity, arterial hypertension, otitis media, head injuries, antibiotics, corticosteroid withdrawal, deficiency or excess of vitamin A, iron deficiency anaemia, and disturbances of endocrinological metabolism (Cinceripini et al. 1999). However, in a significant number of children BIH has no detectable aetiology, therefore it is defined as idiopathic. In neuroborreliosis – an infection of the nervous system caused by the spirochete Borrelia burgdorferi – patients most commonly develop lymphocytic meningitis, radiculoneuritis, or cranial neuropathy. Many patients, however, present with unusual focal neurological symptoms. In this report we describe a 9-year-old male who had BIH with trochlear palsy due to neuroborreliosis.

Case report

A 9-year-old male with no history of significant illness was admitted to our children's hospital in January 2001 because of diplopia. The patient's mother remembered the child had received two tick bites during the previous summer, about 7 and 9 months earlier. Physical examination revealed no fever, no arthralgia, nor erythema, but an infection of the upper respiratory tract and a palsy of the left trochlear nerve resulting in vertical and torsional diplopia in the right gaze. The patient had visual acuity of 1.25 and intact visual fields in both eyes. In ophthalmoscopic and ultrasound examination a marked bilateral papilledema (right > left, 1/0.6mm) of the optic disc was found (Fig. 1), whereas retinal, vascular, or chorioidal abnormalities were not obvious. Visually evoked potentials (VEP) revealed normal amplitudes for both eyes, however, a slight prolongation of latencies in both eyes was noted, which was not consistent with optic neuritis. On cranial CT and MRI examination there were no sellar or suprasellar abnormalities,

normal-sized lateral ventricles, nor subarachnoid spaces. A lumbar puncture showed an elevated opening CSF pressure of 28cm H_2O (horizontal position, no sedatives administered) with nine lymphocytes/μL and normal protein (0.25g/L), glucose (61mg/dL), and lactate (1.2μmol/L) concentrations. Furthermore, EEG and biochemical findings including differential blood cell count; blood coagulation; serum electrolytes; urine analysis; renal, hepatic, and thyroid functions; antibodies against cardiolipin; mitochondria; liver–kidney microsomes; and antinuclear antigens were normal. For treatment of BIH, a therapy with acetazolamide 10mg/kg body weight/day and furosemide 1mg/kg body weight/day was initiated. The patient was still disturbed by diplopia on gaze to the right after day 5 of therapy, when IgM and IgG antibodies in CSF and serum against Borrelia burgdorferi were proved to be highly positive by specific enzyme-linked fluorescent assay (Index 2.1, regarded as positive if >1; Vidas Lyme IgG and IgM Kit; bioMerieux, Paris, France). The diagnosis of neuroborreliosis was verified by immunoblot (IgM positive for Osp C and p41 protein-fragments; IgG positive for p83/100, p41, p39, Osp C protein-fragments) of serum and CSF. Serological findings for neurotropic viruses such as adenovirus, cytomegalovirus, herpes simplex, Coxsackie, measles, parotitis, and varicella zoster virus were not evident for an acute infection. Thus, an intravenous antibiotic therapy with 2g ceftriaxone (80mg/kg body weight/day) was initiated, while the treatment with acetazolamide and furosemide was withdrawn. After 14 days of ceftriaxone therapy the diplopia resolved promptly and the patient was discharged from the hospital. In a clinical control examination 3 months later, the trochlear nerve palsy, the bilateral papilledema, as well as the slight VEP latency prolongation were no longer evident and the child remains well until now.

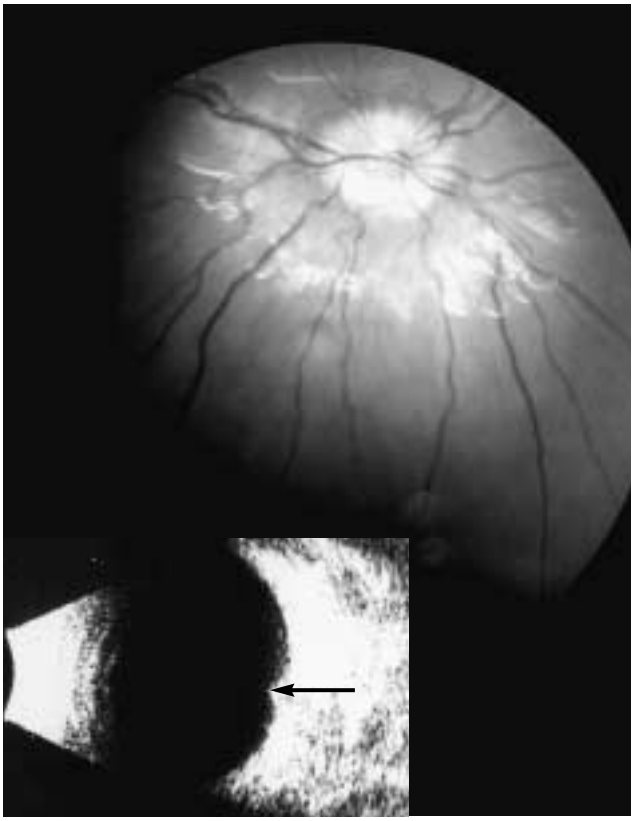

Figure 1: *Ophthalmoscopic and ultrasound findings in benign intracranial hypertension. A papilledema of right optical nerve (1mm) is demonstrated by ophthalmoscopy and ultrasound examination. (Detail: papilledema marked by arrow.)*

Discussion

Neuroborreliosis has become the most frequently diagnosed arthropod-borne infection of the nervous system in Europe and the USA (Christen 1996). In children, the yearly incidence of neuroborreliosis is about five cases per 100 000 children. Lymphocytic meningitis and radiculoneuritis (Bannwarth syndrome) as well as cranial neuropathy with the facial nerve being much more affected than other cranial nerves, account for nearly 90% of all paediatric cases with neuroborreliosis. We described the rare association of neuroborreliosis presenting with BIH and an isolated unilateral trochlear nerve palsy. In our patient oral therapy with furosemide and acetazolamide was not beneficial for precipitation of BIH and therefore withdrawn. Several investigators recommend the use of steroids for BIH in order to prevent injury of the optic nerve (Cinceripini et al. 1999). However, the rapid resolution of neurological symptoms upon ceftriaxone in our patient did not support the additional use of steroids.

In summary, this case report clearly demonstrates that neuroborreliosis can masquerade as BIH (Kan et al. 1998). Furthermore, it underlines the need to rule out infection, including by serology, for treatment of BIH before considering the use of steroids which could have serious consequence for progression of the infectious disease.

Accepted for publication 26th April 2002.

References

Cinceripini GS, Donahue S, Borchert MS. (1999) Idiopathic intracranial hypertension in prepubertal pediatric patients: characteristics, treatment, and outcome. *American Journal of Ophthalmology* **127:** 178–82.

Christen HJ. (1996) Lyme neuroborreliosis in children. *Annals of Medicine* **28:** 235–40.

Kan L, Sood SK, Maytal J. (1998) Pseudotumor cerebri in Lyme disease: a case report and literature review. *Pediatric Neurology* **18:** 439–41.

Notice

7th Congress of the Société Européenne de Neurologie Pédiatrique (SENP)

Palais des Congrès, Paris, France, December 1–4, 2002

Congress languages: French – English

Contact Dr A Arzimanoglou, Child Neurology and Metabolic Diseases, Hôpital Robert Debré, 48 Boulevard Sérurier, 75019 Paris, France.

E-mail: brigitte.tricot@rdb.ap-hop-paris.fr

Websites: www.senp-neuropediatrie.com

www.neuropediatrie-metabolisme.org