CASE REPORT

Pseudotumor cerebri as the presentation of Lyme disease in a non-endemic area

Marta Ezequiel, Ana Teresa Teixeira, Maria João Brito, Catarina Luís

Department of Pediatria, Hospital Professor Doutor Fernando Fonseca, EPE, Amadora, Portugal

Correspondence to
Dr Marta Ezequiel, marta.ezequiel@gmail.com

Accepted 16 March 2018

SUMMARY

Intracranial hypertension is a rare entity in prepubertal children, and its differential diagnosis includes a number of systemic diseases, drugs, vitamin deficiencies and excesses, and hereditary conditions. Infectious aetiology is rare. The case of a 9-year-old boy with intracranial hypertension secondary to acute neuroborreliosis is described. He presented with daily pulsatile frontotemporal headache, pallor, photophobia and phonophobia. His neurological examination revealed papilledema with no nuchal rigidity. The lumbar puncture showed increased pressure (50 cm H_2O) and lymphocytic pleocytosis. Serum and cerebrospinal fluid (CSF) *Borrelia burgdorferi* antibodies were positive. This kind of infection is rare in Portugal but a trip to an endemic area was identified. A careful history, considering the exposure to rural areas together with the intracranial hypertension and inflammatory CSF, are important clues to the diagnosis, allowing the institution to select appropriate treatment.

BACKGROUND

In Europe, Lyme disease is a tickborne illness caused by three pathogenic species of the spirochete *Borrelia—Borrelia burgdorferi*, *Borrelia afzelii* and *Borrelia garinii*. Borreliosis has a broad spectrum of clinical manifestations with multisystem involvement, especially skin in 80% or more of infected individuals, joints (less commonly in Europe), heart, eyes and the nervous system.[1] The symptom most commonly associated with the central nervous system is subacute lymphocytic meningitis, with or without related cranial palsies.[2]

This case report was presented as a poster at the 34th Annual Meeting of the European Society for Paediatric Infectious Diseases in 2016.

CASE PRESENTATION

A previously healthy 9-year-old boy with a medical history of cow's milk and flucloxacillin allergy was admitted with daily pulsatile frontotemporal headache, pallor, photophobia and phonophobia, without night awakening, vomiting or visual changes. He had been to the French Pyrenees 2 months earlier and in rural areas in Portugal in the last 6 months and had had one visit to a pedagogical farm and holidays in the countryside. He did not recall bite marks or cutaneous rash preceding the symptoms. His neurological examination revealed papilloedema and was otherwise normal, namely with no nuchal rigidity. Head CT scan was normal. A lumbar puncture was performed with clear cerebrospinal fluid (CSF), cell count 30, predominantly mononuclear, proteins 29 mg/dL, glucose 55 mg/dL and an increased opening pressure (50 cmH_20). Blood tests were normal except for an erythrocyte sedimentation rate of 36 mm/hour. He started acetazolamide and was admitted for investigation. Serum *B. burgdorferi* antibodies detected by ELISA were positive (IgG 30.59 U/mL, IgM 76.31 U/mL) and confirmed by Western blot. CSF *B. burgdorferi* IgG 28.38 U/mL and IgM 12.19 U/mL and Western blot were also positive confirming the diagnosis. Head MRI was normal. Other bacterial and viral agents were excluded by serologic, PCR and cultural testing, namely *Mycoplasma pneumoniae*, *Epstein-Barr virus*, *Cytomegalovirus* and *Enterovirus*. Serum immunoglobulins were normal. The diagnosis of neuroborreliosis was assumed and a 21-day course of intravenous ceftriaxone was started and completed with full recovery.

OUTCOME AND FOLLOW-UP

He had follow-up for 9 months, in which he remained asymptomatic with no papilloedema and persisted with serum IgG positive until 3 months later.

DISCUSSION

Neuroborreliosis is defined as infiltration of the nervous system by *Borrelia*. The most frequent manifestation is a subacute lymphocytic meningitis, frequently associated with cranial nerve involvement.[2] Isolated cranial nerve involvement is also possible, especially as facial palsy.[3] In adults, neuroborreliosis can be present as a painful radiculopathy or Bannwarth's syndrome, a clinical entity very rarely described in children.[1 3–6]

Contrary to the American presentation, European patients most often do not recall a tick bite or present the typical rash or *eritema migrans*, so a careful epidemiologic history is very important and should be investigated, as in this case.[2]

Intracranial hypertension is rare but has been described in neuroborreliosis.[5 7–10] Sixth cranial nerve palsy, a common finding in idiopathic intracranial hypertension, may also be present.[10] In some cases, the intracranial hypertension occurs in association with lymphocytic meningitis, although clinical manifestations are scarce, with most authors describing an absence of nuchal rigidity, as occurred in our patient.[2] The remaining patients have no inflammatory changes in the CSF.[7 10] In both cases,

To cite: Ezequiel M, Teixeira AT, Brito MJ, *et al*. *BMJ Case Rep* Published Online First: [*please include* Day Month Year]. doi:10.1136/bcr-2017-222976

the hypertension is attributed to the inflammatory changes in the arachnoid, interfering with CSF absorption.

Pseudotumor cerebri is a disorder defined by clinical criteria that include symptoms and signs isolated from those produced by increased intracranial pressure, such as headache, papilloedema, vision loss and elevated intracranial pressure with normal CSF composition. Differential diagnosis for pseudotumor cerebri is vast and includes infectious diseases such as meningitis, otitis media and mastoiditis, obstruction of venous drainage such as venous sinus thrombosis and hyperviscosity, endocrine disorders, obesity, nutritional disorders such as hypervitaminosis A and medications.[7 9] Obesity is a frequent finding in postpubertal patients, but is rare in prepubertal children, like our patient.[7 11] CSF changes are not common, so in their presence, it is compulsory to investigate an infectious origin, as happened in this case, as the intracranial hypertension might be the only symptom of a central nervous system infection.

Learning points

- Pseudotumor cerebri can be the sole manifestation of neuroborreliosis.
- A history of tick bite is often absent in many cases.
- Central nervous system involvement can occur with no cutaneous manifestations.
- *Borrelia* infections should be actively investigated in children with central nervous system disease even in non-endemic areas.

Contributors ME and ATT: conception, design, analysis and interpretation of lab results. ME: drafting the article. MJB and CL: revising it critically for important intellectual content. ATT, MJB and CL: final approval of the version to be published. All authors read and approved the final manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1 Baumann M, Birnbacher R, Koch J, *et al*. Uncommon manifestations of neuroborreliosis in children. *Eur J Paediatr Neurol* 2010;14:274–7.
2 Pachner AR, Steiner I. Lyme neuroborreliosis: infection, immunity, and inflammation. *Lancet Neurol* 2007;6:544–52.
3 Belman AL, Iyer M, Coyle PK, *et al*. Neurologic manifestations in children with North American Lyme disease. *Neurology* 1993;43:2609.
4 Pfister HW, Rupprecht TA. Clinical aspects of neuroborreliosis and post-Lyme disease syndrome in adult patients. *Int J Med Microbiol* 2006;296(Suppl 40):11–16.
5 Castaldo JE, Griffith E, Monkowski DH. Pseudotumor cerebri: early manifestation of adult Lyme disease. *Am J Med* 2008;121:e5–6.
6 Francisco T, Marques M, Vieira JP, *et al*. Bilateral facial palsy: a form of neuroborreliosis presentation in paediatric age. *BMJ Case Rep* 2013;2013:bcr2012008060.
7 Kan L, Sood SK, Maytal J. Pseudotumor cerebri in Lyme disease: a case report and literature review. *Pediatr Neurol* 1998;18:439–41.
8 Nord JA, Karter D. Clinical Infectious Diseases. 2003;37:e25–6.
9 Abadie I, Marc E, Bursztyn J, *et al*. Hypertension intracrânienne isolée par maladie de Lyme. *Archives de Pédiatrie* 2003;10:791–3.
10 Şahin B, İncecik F, Hergüner ÖM, *et al*. A presentation of lyme disease: pseudotumor cerebri. *Turk J Pediatr* 2015;57:522–4.
11 Ramgopal S, Obeid R, Zuccoli G, *et al*. Lyme disease-related intracranial hypertension in children: clinical and imaging findings. *J Neurol* 2016;263:500–7.