

An eight-year-old girl with probable Lyme disease in Hong Kong

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ABSTRACT

An eight-year-old girl presented with acute cerebellar syndrome and meningitis. Lumbar puncture showed elevated pressure. Examination of the cerebrospinal fluid showed lymphocytosis, raised protein and decreased glucose. She was initially treated with acyclovir and antituberculosis drugs with partial response. Subsequently, the antibody assay for *Borrelia burgdorferi* was positive and the antimicrobial regime was changed to ceftriaxone. She made a full recovery with no neurological deficit. A probable diagnosis of Lyme disease was made and the diagnostic approach to Lyme disease is discussed.

Keywords: acute cerebellar syndrome, *Borrelia burgdorferi*, Lyme disease, meningitis, tick infection

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INTRODUCTION

Lyme disease is a systemic infection caused by the spirochaete, *Borrelia burgdorferi* (*B. burgdorferi*). It is transmitted by the ticks of the genus *Ixodes*. There are three clinical stages: early-localised, early disseminated and late-persistent.⁽¹⁾ However, stages may overlap and most patients do not experience all stages. In the early localised stage, erythema migrans (EM), an enlarging erythema occurring at the site of the tick bite, is the only manifestation. Neurological, cardiovascular, and musculoskeletal involvements are the features of the other two stages. It is a rare disease in Hong Kong. We present an eight-year-old girl with acute cerebellar syndrome and meningitis probably caused by *B. burgdorferi*.

CASE REPORT

An eight-year-old girl was transferred to our unit from another hospital for further management of her acute cerebellar syndrome. She enjoyed good health in the past with no recent travel history. No rash was ever noticed. She had chicken pox infection at the age of three years. She initially presented with fever, headache and vomiting. She was managed conservatively as a case of upper respiratory tract infection. On day five of fever, she was noticed to have "tremor of eyes" by her parents. Acute

cerebellar syndrome was diagnosed by her attending paediatrician and she was transferred to our unit.

Upon admission, she was lethargic but orientated in time, place and person. Temperature was 38.7°C, Glasgow coma scale was 15/15. Other vital signs were stable. Neck stiffness and cerebellar signs, i.e. nystagmus, intention tremor, past pointing, dysidiadochokinesia and unsteady gait, were positive. Examination of other systems was normal. Serum sodium level was decreased to 125 mmol/L. Other blood results including complete blood count, erythrocyte sedimentation rate, C-reactive protein, clotting profile, liver function, renal function, glucose and blood culture were all normal. Computed tomography of her brain did not show any space-occupying lesion. Lumbar puncture showed an elevated opening pressure of 22 cm water. Microscopy of cerebrospinal fluid (CSF) showed white blood cell of 37/mm³ (10% polymorphs, 90% lymphocytes), red blood cell of 16/mm³. CSF protein was raised to 1.63 g/L, glucose decreased to 2.4 mmol/L (33% of blood glucose level). CSF Gram stain, acid-fast bacilli smear and latex test for *Neisseria meningitidis*, *Streptococcus pneumoniae* and *Haemophilus influenzae* were all negative. *Mycobacterium tuberculosis*, and herpes simplex polymerase chain reaction (PCR) assay results were negative. Bacterial, viral and *M. tuberculosis* cultures were negative. Blood for mycoplasma, Epstein-Barr virus, viral titre were negative. Blood and urine cultures were negative. Blood and urine toxicology were unremarkable. Her Mantoux test was negative. Two serial blood specimens were sent for *B. burgdorferi* antibody detection by enzyme-linked fluorescent immunoassay (ELFA) (VISAS[®] Lyme IgG and IgM, bioMerieux Inc, MO, USA), a quantitative assay with a sensitivity of 96.4% and specificity of 97.9% according to the manufacturer. Both were positive. Western blot confirmation was not done as it was not available in Hong Kong.

Intravenous acyclovir, cefotaxime, erythromycin and anti-tuberculosis (ATB) drugs with prednisolone and pyridoxine were started. Acyclovir, erythromycin and cefotaxime were stopped subsequently as the results were negative, while the ATB drugs were continued. She improved clinically with defervescence, resolution of neck stiffness and decreased severity of cerebellar signs. ATB drugs were stopped one month later, when

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the blood test for antibody against *B. burgdorferi* was positive. She was then treated for Lyme disease with two weeks intravenous ceftriaxone. Upon discharge, she still had mild intention tremor and unsteady gait. The cerebellar signs progressively resolved and she recovered completely six months later. At the last follow-up three years later, she remained well.

DISCUSSION

Lyme disease is a rare disease entity in Hong Kong, although it is commonly reported in North America and Europe. In the United States, it is an important endemic infection. Lyme disease takes its name from the town Lyme in Connecticut, USA, where it was identified in the mid 1970s.⁽²⁾ The diagnosis of Lyme disease requires the presence of the EM rash diagnosed by a physician or the presence of other clinical manifestations accompanied by positive Lyme-serology testing with confirmatory immunoblotting.⁽³⁾ EM was only reported in 60%–80% of children with Lyme disease.^(4,5)

Our patient presented with lethargy, neck rigidity and signs of cerebellar dysfunction. Her CSF showed elevated protein and depressed glucose and lymphocytosis. Tuberculosis (TB) would be the most likely candidate in Hong Kong and hence ATB treatment was started. However, the negative Mantoux test, the negative *M. tuberculosis* PCR results and cultures made the diagnosis less likely. The positive *B. burgdorferi* antibody titre provided the alternative explanation. However, the presentation of our case was not a typical one. In at least 80% of patients in the USA, the Lyme disease began with EM, where 15%, 5%, 60% of them had neurological, cardiac and musculoskeletal involvement, respectively.⁽⁶⁾ For the neurological symptoms of Lyme disease, there is a wide spectrum of manifestations, including aseptic meningitis, meningoencephalitis, Bell's palsy and other cranial neuropathies, radiculoneuritis or myelitis.⁽⁷⁾ However, the current case did not have any dermatological or musculoskeletal symptoms. The patient did not live in an endemic area and there was no recent travel history. Acute cerebellar syndrome is also a rare presentation of Lyme disease.⁽⁸⁾ Nevertheless, Lyme disease was the most likely diagnosis as TB was practically ruled out by the negative *M. tuberculosis* culture, PCR assay, and Mantoux test results. This was further supported when that the child remained well one month after treatment for

TB was discontinued.

Positive culture of *B. burgdorferi* provides the confirmatory evidence for Lyme disease.⁽⁹⁾ However, culture of *B. burgdorferi* involves incubating a specimen in Barbour-Stoenner-Kelly medium, which is not available in Hong Kong. The alternative diagnostic approach involves detection of a diagnostic level of antibody against the spirochaetes by the two-test approach of enzyme-linked immunosorbent assay (ELISA) followed by Western blotting to increase specificity.⁽¹⁰⁾ The main limitation of the current case was the absence of the second step of the more specific Western blot because it was not available in Hong Kong. Therefore, confirmation of Lyme disease was difficult in Hong Kong. Absence of the confirmatory test in Hong Kong dictated that the serological test, i.e. ELISA, must be correlated with the clinical features, and the current case could only be regarded as a probable case of Lyme disease. In conclusion, Lyme disease is an uncommon disease in Hong Kong. It involves the neurological, cardiac and musculoskeletal systems. As the definitive laboratory diagnostic test is not available in Hong Kong, Lyme disease can only be classified as probable in Hong Kong.

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