Case Report

Is that a scolex? a case of clinically isolated syndrome

Van K. Ma*, Lourdemillard Bellevue, Maria Espiritu Fuller

Department of Pediatrics, Children’s Hospital of New Jersey at Newark Beth Israel Medical Center, Newark, New Jersey, USA.

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*Correspondence:
Dr. Van K. Ma,
Email: vankma@gmail.com

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ABSTRACT

Clinically Isolated Syndrome is an initial demyelinating event of the central nervous system that has been associated with the future development of multiple sclerosis. Diagnostic studies include clinical and paraclinical studies. Patients with lesions on MRI of the brain at baseline will more likely develop multiple sclerosis compared to patients without findings. We report a case of a 10-year-old female of Colombian ancestry and origin, who presented with indiscernible neurological clinical signs and symptoms, with MRI brain with and without contrast showing demyelinating lesions with one lesion “suggesting” a scolex.

Keywords: Cysticercosis, Clinically isolated syndrome, Encephalopathy, Multiple sclerosis, Neurocysticercosis, Scolex

INTRODUCTION

Clinically Isolated Syndrome (CIS) is defined as an initial presentation of disease with characteristics of an inflammatory demyelination pathology, but without dissemination in time. Patients who present with a clinically isolated attack, by definition, do not have multiple sclerosis. However, 30-70% of patients with a clinically isolated syndrome go on to be diagnosed with multiple sclerosis (MS). One longitudinal study of a cohort of 236 patients ages 10-56 years old showed that 17.8% did not have a relapse within 25 years. We present the case of a patient with MRI brain with and without contrast “suggesting” a scolex.

CASE REPORT

We present a case report of a 10-year-old female with no significant past medical history who initially presented to the emergency room with a one-week history of difficulty with articulation and swallowing, worsening weakness of the right upper and lower extremities, intermittent pain of her feet, and frequent falls. She had an episode of generalized weakness and ataxia with associated vomiting one week prior to admission but all symptoms had self-resolved within two days. She immigrated to the United States from Colombia five years prior and had not since returned. Her physical exam was significant for the following: bilateral end-gaze nystagmus, slurred speech, decrease gag reflex, right-sided weakness (lower more than upper extremity), pronator drift, hyperactive patellar reflexes (right more than left), positive Babinski bilaterally, right foot drop, and toe-walking with a wide-stance gait on ambulation. Patient was immediately started on ceftriaxone and acyclovir for possible infectious causes and transferred to the PICU. Neurology was consulted and levetiracetam was added for seizure prophylaxis.

CBC, CMP, and urine toxicology were unremarkable. LDH was mildly elevated. Blood culture, and stool for ova and parasites x3 were negative. Serological studies (IgM/IgG) for Lyme disease and toxoplasmosis were negative. Cysticercosis IgG x2 were negative. Initial CSF analysis was normal. Herpes simplex virus and enterovirus CSF PCR, and IgG CSF were all negative.
48-hour video EEG was suggestive of diffuse encephalopathy. CT head without contrast showed confluent white matter hypodensities of uncertain chronicity (Figure 1).

![Figure 1: CT head without contrast showing confluent white matter hypodensities of uncertain chronicity.](image)

MRI brain with and without contrast showed innumerable enhancing lesions with one “suggesting” a scolex which led to a diagnosis of neurocysticercosis (Figure 2).

![Figure 2: MRI brain “suggesting” a scolex.](image)

Infectious disease was consulted, and the patient was started on albendazole 7.5 mg/kg every 12 hours to complete a 15 day-course in conjunction with steroids initiated at 20 mg/kg per day and tapered on day three. After three days of therapy, improvement of right lower extremity was noted.

In light of nonspecific findings and to further explore the extent of the disease, a whole-body MRI was done which showed “multiple short segments T2 and STIR hyperintense lesions in the cervical and thoracic cord without cord expansion or enhancement”. A repeat CSF analysis showed four well-defined gamma bands, corresponding to abnormal synthesis of gammaglobulins. Serum electrophoresis showed hypogammaglobulinemia but was notable for an elevated alpha-1-globulin. Myelin basic protein was negative.

Given the continued improvement in her symptoms, she was transferred to a children’s inpatient rehabilitation facility with the working diagnosis of acute encephalopathy. She remained at the facility for 18 days. Roughly six weeks after the onset of illness, the patient had one follow up visit with neurology at which time a repeat MRI brain was ordered but not completed due to loss of medical insurance. Multiple attempts were made to contact the family and not until 18 months later, the primary care team was finally able to reach the mother who endorsed complete return to pre-illness baseline with no further recurrence of symptoms. Mother was encouraged to continue working to obtain medical insurance. In the interim, the patient has remained lost to follow-up.

**DISCUSSION**

As with many neurological disorders, no definitive diagnostic tests are available for the diagnosis of CIS, and diagnosis is primarily based on clinical and paraclinical data. While the scolex-like enhancing lesion and the patient’s background initially led us to think this could be a case of neurocysticercosis, the patient’s serology remained negative. In a study comparing patients with neurocysticercosis and other neurological disorders, the sensitivity of both serum and CSF samples was 25.0% in inactive neurocysticercosis and 90.9% in active cases, specificity was 100% and 95.8% respectively.\(^3\) When serum and CSF samples were combined, the sensitivity was 100% in active neurocysticercosis.\(^3\) In our case, a CSF sample sent for cystercerosis PCR would have been helpful in a diagnosis of neurocysticercosis.

In the past, in the absence of other diseases, clinical dissemination in space and time of an inflammatory-demyelinating process has been the gold standard for the diagnosis of MS. Based on the initial presentation of a database of 1844 patients with MS diagnosed using the Poser criteria, 18% initially presented with optic neuritis, 52% with long-tract signs and symptoms, 9% with brainstem syndrome, and 21% with multifocal abnormalities.\(^4\) The International Panel of McDonald and colleagues have incorporated MRI evidence of dissemination in time and space to allow for a diagnosis of MS in patients with CIS. The abnormalities on MRI of the brain in CIS are compatible with those associated with early MS. Based on clinical presentation and MRI findings at baseline, 3 months, 1 year, and 3 years follow-up, 21%, 48%, and 58% of patients were eventually diagnosed with MS respectively using the McDonald criteria.\(^5\) The development of MS with the McDonald MRI criteria after 1 year in those with a CIS had a sensitivity of 83%, specificity of 83%, positive predictive value of 75%, negative predictive value of 89%, and accuracy of 83% for clinically definite MS at 3 years.\(^5\) A study following patients for 14
years, found that 49 of 50 (98%) patients with abnormalities on MRI at baseline had clinical or radiologic evidence consistent with MS. As a result, white-matter lesions on MRI in young adults with isolated syndromes are highly suggestive of multiple sclerosis.

In addition, various CSF abnormalities have been found in patients with CIS. Two or more IgG oligoclonal bands in the CSF without corresponding bands in the serum is found in 60–70% of patients with CIS and is associated with an increased risk of developing MS. The presence of oligoclonal bands in conjunction with MRI findings in diagnosing MS was found to increase specificity and negative predictive value. Myelin-basic-protein (MBP) and proteolipid-protein antibodies in CSF have been found in more patients with possible onset symptoms of MS and clinically definite MS. In addition, antibodies against myelin-oligodendrocyte-glycoprotein (MOG) have been found in acute lesions in patients with multiple sclerosis. In a study of 103 patients with a CIS, oligoclonal bands in their CSF, and positive MRI scans, 21 of 22 (95%) patients with both serum MOG and MBP antibodies had a second episode. In contrast, 35 of 42 (83%) patients positive for MOG antibodies alone had a second episode. In light of our negative MBP result, it would have been worthwhile to measure the serum MOG in our patient. Even though the patient’s definitive diagnosis was never confirmed, the case highlights the inconclusiveness of her presentation, medical work-up, and the importance of adequate follow up. Based on her clinical presentation, neuroimaging studies, and literature review, this patient likely suffered a CIS. While she has not had any clinical symptoms within the last 18 months, data on CIS is based on prospective studies with patients having relapses in the near future. The presence of demyelinating lesions on her whole-body MRI and oligoclonal bands in her CSF are consistent with findings in patients who went on to develop clinically definite MS. It is pivotal for her to be monitored for any relapses.

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REFERENCES
