

Acute Post Infectious Cerebellitis Presenting as Hydrocephalus: A Rare Presentation

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ABSTRACT

Acute post infectious cerebellitis, also known as acute cerebellar ataxia, is a rare inflammatory syndrome often characterized by cerebellar dysfunction of rapid onset. Although usually benign and self-limiting, acute cerebellitis may manifest a fulminant course. We describe a patient referred to our center with acute cerebellitis, complicated by hydrocephalus.

An 32-year-old female presented with headache, nausea, vomiting, and gait and limbs ataxia 2 weeks after following a respiratory tract infection. CT showed obstructive hydrocephalus. MRI showed bilateral cerebellar swelling with hyperintense signals on T2-weighted imaging. The diagnosis was acute post infectious cerebellitis. Following treatment with corticosteroids, the clinical signs resolved. The patient made an uneventful recovery, with elimination of neurologic deficit. *JMS* 2010;13(2):67-69

Key Words: Cerebellitis, ataxia, demyelination, steroids

Acute postinfectious cerebellitis is usually a self-limiting condition presenting with sudden-onset ataxia, and is characterized by complete recovery in the majority of cases. However, a few recent reports of acute cerebellitis with a critical or potentially critical course have changed the conception of this disease from an acute cerebellar ataxia with a benign course to a more serious disease entity with a heterogeneous pathogenesis.¹ A diagnosis of acute cerebellitis is difficult because clinical presentations often vary, resulting in diagnostic delay. Some patients may present with only mild cerebellar signs, whereas others may present with brainstem involvement. We report a patient referred to our department as a case of acute post infectious cerebellitis complicated by hydrocephalus.

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Case Report

A 32 year old female was referred to our department with complaints of severe occipital headache, unsteadiness of gait (swaying to either side) and vomiting for one month. The patient also had an episode of abnormal body movements in the form of generalized tonic clonic seizures. Two weeks before admission, she had manifested cough and fever for a few days which resolved spontaneously. There was no history of trauma, dysarthria or difficulty in swallowing and feeding.

On admission general physical examination and systemic examination was normal. Neurological examination revealed that the patient was conscious and oriented, and had pancerebellar ataxia and nystagmus. Deep tendon reflexes were hyperactive, with no signs of meningeal irritation. Her fundus examination was normal.

Laboratory results indicated mild leucocytosis with neutrophilia (12,400 cells per mm³ with 78% neutrophils). Results in terms of blood chemistry, urine analysis were normal. The cerebrospinal fluid examination was normal.

Oligoclonal bands in CSF were negative. A non contrast CT scan of her head that revealed hydrocephalus secondary to fourth ventricular obstruction because of cerebellar edema. The cerebellum showed a non enhancing hypodensity in both hemispheres. A diagnosis of obstructive hydrocephalus with an underlying cerebellar space occupying lesion was considered. An external ventricular drainage in the form of ventriculo-peritoneal shunt was performed. Magnetic resonance imaging of the brain (T2 and FLAIR picture) showed bilateral, symmetrical hyperintensities involving the cerebellar white matter and (C shaped hyperintensity).

A final diagnosis of acute post infectious cerebellitis was made. The patient was managed with intravenous methylprednisolone 1000mg for five days. The patient had a

remarkable recovery within two weeks of hospitalization and was ambulatory without support on discharge.

Discussion

Acute cerebellitis is defined as a neurologic condition that consists of nausea, headache, and altered mental status, including loss of consciousness and convulsions in addition to the acute onset of cerebellar signs. Also known as acute cerebellar ataxia, cerebellitis occurs most commonly in young children and may be difficult to diagnose on routine clinical and laboratory studies. According to the proposed consensus definition by the International Multiple Sclerosis Study group for central nervous system inflammatory demyelinating

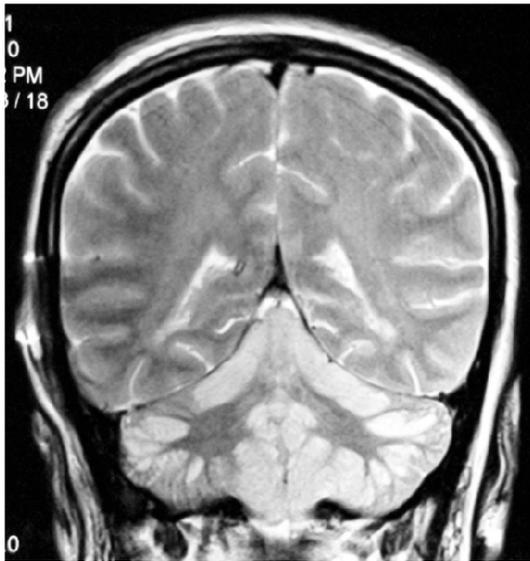


Figure 1:



Figure 2:

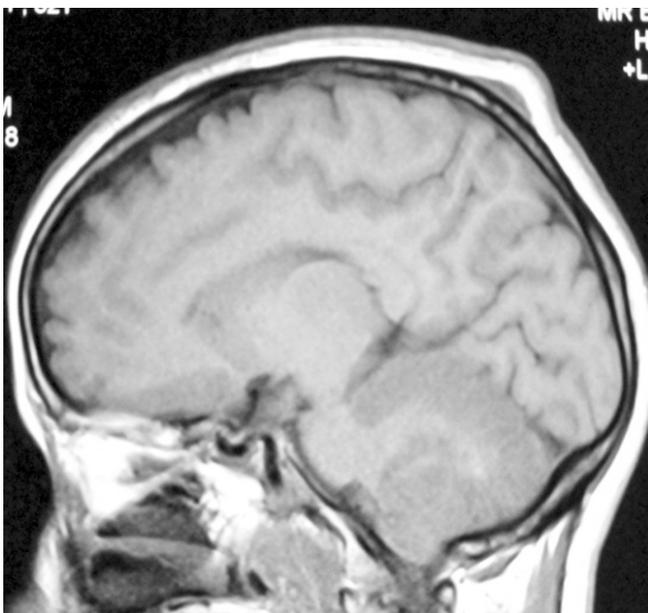


Figure 3: MRI pictures C shaped hyperintensity in cerebellum.

disorders of childhood in 2007, cerebellitis is a type of clinically isolated syndrome.²

Cerebellitis is an inflammatory syndrome resulting in acute cerebellar dysfunction, which can occur as a primary infectious, postinfectious, or postvaccination disorder. It may also occur without evidence of an antecedent or concurrent factor.³ Cerebellitis may occur due to a host of viral agents, including enteroviruses, herpesviruses, HIV, and rabies. Bacterial infections have also been associated with cerebellitis, including *Borrelia burgdorferi* (Lyme disease), *Mycoplasma pneumoniae*, *Legionella*, and *Coxiellaburnettii* (Q fever). In addition, cerebellitis, cerebellitis may follow immunizations such as hepatitis, smallpox, and measles vaccination.⁴ In many cases, however, the precise causative agent is not isolated. Differential diagnosis of postinfectious cerebellitis should include drug overdose. A history of recent exposure to drugs such as phenytoin, carbamazepine or alcohol must be ruled out in every patient.⁵

The sensitivity of MRI for the detection of cerebellitis is

not known. A few patients with cerebellitis have presented with a normal MRI. Abnormal noncontrast MRI findings in cerebellitis have only been described in a few case reports, many of which occurred in young children. Isolated cerebellar abnormalities were noted, including parenchymal hyperintensities on T2-WI, swelling, and secondary obstructive hydrocephalus. Abnormal MRI enhancement may be seen in some but not all cases in acute and subacute stages of the disease. In general the characteristic MRI T2 and FLAIR picture of postinfectious cerebellitis is bilateral, symmetrical hyperintensities involving the cerebellar white matter and taking exactly the shape of the cerebellar white matter and mapping it (C-shaped hyperintensity). The C-shape hyperintensity is due to cerebellar white matter vasogenic edema that develops secondary to the immune mediated inflammatory demyelination of the white matter of the cerebellum which results in breakdown of the blood brain barrier with subsequent development of vasogenic edema that follows the myelinated axons of white matter tracts and association fibers of the cerebellum, spreading them apart and extending alongside them resulting in the characteristic C-shape of the MRI T2 and FLAIR hyperintensities.⁶⁻⁸

On precontrast CT scan images the bilateral white matter cerebellar lesions appear as a bilateral symmetrical C shaped hypodensity and on precontrast MRI T1 images the cerebellar white matter lesions appear as a bilateral symmetrical C-shaped hypointensity. Due to breakdown of blood brain barrier, (which is responsible for the formation of vasogenic edema) some degree of contrast enhancement should be expected. Cerebellar swelling (which results from inflammation and edema) might induce compression of the 4th ventricle and variable degrees of obstructive hydrocephalus. Follow-up studies showed reversals of the acute changes and the development of atrophy years later in severe cases.

Post infectious regressive demyelination white matter diseases represent an autoimmune response to proteins, most probably myelin-basic proteins, in the CNS with perivenous inflammation and demyelination found in autopsy and biopsy studies. Demyelination may not be present in the first few days of the disease. Demyelination of the white matter is associated with breakdown of the blood brain barrier and the development of vasogenic edema. Vasogenic edema is the most common type of edema results from local disruption of the blood brain barrier.

The prognosis of acute cerebellitis is usually good. Even patients with severe symptoms and increased intracranial pressure can recover completely without any sequelae. Because acute cerebellitis has a variable clinical course, each case of acute cerebellitis must be treated and considered individually. Treatment modalities can range from medical care to aggressive surgical decompression.¹⁰ Mild cases without progression of clinical or radiologic findings should be closely

monitored with serial magnetic resonance images. Steroids (pulse methylprednisone with close observation) are the first line of treatment when signs are moderate to severe.^{3,9} However, most patients will recover without steroids or any specific treatment. Because of a lack of large series of patients with acute cerebellitis, the heterogeneity of the condition, and reports of patients worsening despite steroids, the exact role of steroids is unclear. In more severe cases, marked cerebellar swelling may result in obstructive hydrocephalus, requiring the placement of an external ventricular drain, with or without corticosteroid therapy. Such patients should be closely monitored in an intensive care setting because of the risk of upward herniation. Sudden deaths have been reported following fulminant cerebellitis.¹¹ Death in acute cerebellitis is usually due to severe cerebellar swelling resulting in transtentorial and transforaminal herniations.³

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