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Case Report

Infectious Cerebellitis Rare Entity About Two Cases

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Abstract

Introduction: Cerebellitis is an inflammatory pathology of cerebellar structures more common in children, frequently of post-infectious origin or following vaccination, seldom during a viral or bacterial infection. Ataxia is most frequently caused by dysfunction of the complex circuitry connecting the cerebellum, basal ganglia and cerebral cortex. Results: We report the case of 2 patients aged 3 and 10 years presenting acute cerebellitis of infectious origin: Ataxic syndrome associated with balance disorders and speech disorders and fever. The evolution was quickly favorable with corticosteroid therapy and proprioceptive physiotherapy. The absence of fatalities in our case report suggests early diagnosis, and steroid treatment can increase the chance of recovery. Discussion: Clinical presentations are deceptive and variable with cerebellar symptoms of acute kinetic and static ataxia with inflammatory signs. The treatment is based on steroids when symptoms are moderate to severe. Antimicrobial therapy should always be considered, because ataxia can be a presenting sign of both viral encephalitis and bacterial meningitis. The prognosis for acute cerebellitis is generally favorable. The courses vary from what is commonly a benign and self-limiting disease to what occasionally is fulminant disease resulting in several cerebellar damage or even sudden death. Conclusion: Recognizing ataxia in children can be difficult, which is why a lumbar punction should be considered if infectious cerebellitis is suspected. Most people fully recover, however there is a risk of lasting disability.

Keywords

Lumbar Punction, Magnetic Resonance Imaging, Cerebellitis, Acute Cerebellar Ataxia

1. Introduction

Acute cerebellitis is defined as an acute neurologic condition characterized by cerebellar ataxia or dysfunction that is attributable to a recent or concurrent infective illness, a recent vaccination or ingestion of medication, and in which there is MRI evidence of predominantly cerebellar inflammation [1]. There is two pediatric observations of infectious cerebellitis, their mode of revelation, diagnosis and evolution.

2. Case Report

2.1. Patient 1

The first case reported is about a 3-year-old female child, with no particular history including good psychomotor development, well vaccinated, no recent infectious episodes, no medication or toxic substances. At the department, she presented for an ataxic syndrome with paraparesis of both legs, evolving for 5 days in a context of apyrexia, associated with

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balance disorders without visual or swallowing disorders. Clinical examination revealed a conscious apyretic patient, with ataxic gate, abolished ROT without sensory deficit. Cerebrospinal fluid analysis revealed 233 White cells/mm^3 (95% lymphocytes), Proteinorrachia was 0.32 g/L and glycorrhachia was 0.5 g/L. Direct examination and culture were negative. Brain Computed Tomography scan was normal with negative CRP. The ENMG route was normal. The patient received corticosteroid therapy (Methyl prednisone) at a rate of 2mg/kg/8h for 1 week and proprioceptive physiotherapy with full recovery.

2.2. Patient 2

The second case reported is about a 10-year-old girl, with no particular history, no recent infectious episode, no medication or toxic substances. Initially she presented for an intracranial hypertension syndrome of vomiting and headache in a helmet without associated visual disturbances, evolving in a context of fever estimated at 39 °evolving one week before her hospitalization. The course was marked 4 days later by an ataxic syndrome associated with balance disorders and dysarthria without visual or swallowing disorders. Clinical examination on admission revealed a patient who was conscious, feverish, with the following neurological examination: Ataxic walking, standing that was difficult to maintain, ROT present without sensory deficit. The eye fundus was normal. Brain CT scan showed areas of right cerebellar hypodensity repressing V4 with ptosis of cerebellar tonsils. Analysis of cerebrospinal fluid (CSF) found 40 elements/mm^3 of which 80% lymphocytes, proteinorachia was 0.4 g/l, glycorrhachia 0.78 g/l. Direct examination and culture were negative. Brain magnetic resonance imaging (MRI) study showed a bilateral and asymmetrical diffuse cortical cerebellar thickening without systematization, in discrete T1 hypo signal, T2 hypersignal and FLAIR, with discrete meningeal contrast in view, a mass effect on the brainstem and V4 without upstream dilation and ptosis of the cerebellar tonsils at the level of the Mc Rae line. CSF multiplex PCR isolated Haemophilus influenzae. The patient received corticosteroid therapy (methylprednisone) at a rate of 2mg/kg/8h for 1 week and proprioceptive physiotherapy with full recovery.

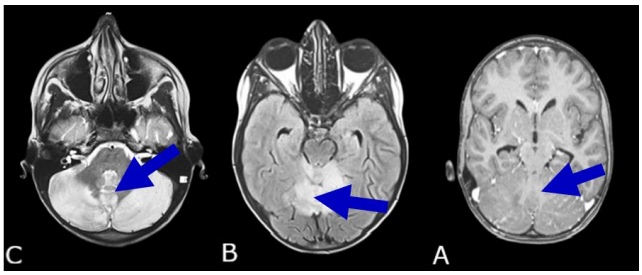

Figure 1. *Magnetic resonance imaging (MRI) axial cut. Patient 2. (A) T1 injected, (B) T2 FLAIR Sequence, (C) T2 Sequence. Bilateral and asymmetrical diffuse cerebellar cortical thickening without systematized character, in discrete T1 hypo signal, T2 hypersignal and FLAIR, with discrete meningeal contrast taking in front of it, a mass effect on the brainstem and V4 without upstream dilation and ptosis of the cerebellar tonsils at the level of the Mc Rae line.*

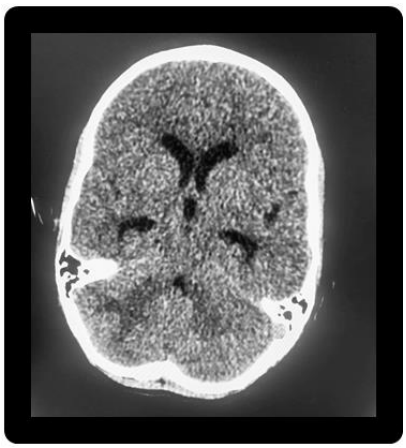

Figure 2. *Brain CT scan. Helical acquisition without contrast injection. Ranges of right cerebellar hypodensities repressing V4 with ptosis of cerebellar tonsils.*

Table 1. *Clinical and biological features of infectious cerebellitis in 2 cases.*

Observation	Patient N °1	Patient N °2
Age / Gender	3 Years /Girl	10 Years /Girl
Clinical	Disorder Walking Right Paraparesis Fever at 39 °(5 days)	CITH syndrome: helmet headache + jet vomiting, Fever at 38.5 °. Ataxic walking (7 days)
LCR	White cells = 233 (95% Lympho) Negative CRP	White cells = 40 (80% Lympho) CRP at 38; PCR: Haemophilus Influenzae.
CT / Brain MRI	Normal Brain CT	Cerebral CT: cerebellitis with tonsillar ptosis Brain MRI: Infectious cerebellitis of viral origin.

Observation	Patient N °1	Patient N °2
Treatment	Méthylprednisone Proprioceptive Physiotherapy	Méthylprednisone Proprioceptive Physiotherapy
Evolution	Favorable	Favorable

3. Discussion

Acute cerebellitis, one of the most common causes of acute cerebellar damage in infants, can be infectious, post-infectious, or post vaccination [2, 3]. It's typically viral. Patients may be completely symptom-free; although they frequently experience mild to severe cerebellar symptoms such as irregular spontaneous eye movements, ataxia, wide walking, and tremors, which are accompanied by headache, drowsiness, stiff neck, and vomiting.

The term acute refers to symptoms that last from a few hours to two days.

It normally has a self-limiting course, but it can infrequently become fulminant. Acute infectious cerebellitis is typically differs from post-infectious cerebellitis by the presence of fever and the absence of a latent period following a non-specific infection.

Infectious agents associated with acute cerebellitis are: Epstein-Barr virus, varicella-zoster virus, herpes simplex virus-1, Coxackies virus, influenza, respiratory syncytial virus, cytomegalovirus, rotavirus, West Nile virus, Streptococcus pneumoniae, Borrelia burgdorferi, M. pneumoniae, and Salmonella typhi [4, 5]. An autoimmune mechanism has also been proposed for acute cerebellitis given its post-infectious origin and detection of autoantibodies in some patients targeting glutamate receptors, Purkinje cells, centrosomes, cardiolipin, gangliosides, and glutamic acid decarboxylase.

The mechanism underlying this cerebellar dysfunction is yet unknown. In some situations, the infectious agent continues to play a direct role until it is isolated in the CSF [6]. Elsewhere, an immunological alteration similar to that of acute disseminated encephalomyelitis (EMAD) persists [7].

In the case of acute infection-like symptoms, the examination should also include sensory or vestibular dysfunction, which can produce ataxia. In most infections, motor dysfunction will predominant; however, when dentate nuclei are selectively implicated, the cognitive-affective syndrome may influence the clinical presentation [8].

The classic clinical presentation includes cerebellar symptoms of acute kinetic and static ataxia with inflammatory signs. Clinical manifestations are often bilateral and symmetrical, however unilateral cases of acute hemi ataxia with hemiparesis have been reported [9, 10]. Other symptoms may include dysarthria, headache, nausea, myoclonus, abnormal eye movements and sometimes transitory altered consciousness or even convulsions, suggesting that the process is not necessarily limited to the cerebellum; fever and meningeal syndrome may or may not exist. This often-benign pathology can evolve to severe forms, with fulgurating evolutions [11] or potentially serious motor or cognitive sequelae [12]. Ataxia is not always in the foreground, and may even be omitted from the picture. frequently, the notion of a recent viral illness with upper airway infection is discovered.

CSF is normal or shows moderate lymphocytosis.

Although typically very suggestive, this clinical picture is not always complete nor specific, and the diagnosis of cerebellitis has traditionally been one of exclusion [13].

Imaging exploration, particularly MRI, now allows us to support the diagnosis and to quickly relate the intracranial hypertension syndrome, when it is isolated and in the foreground, to cerebellitis and to quickly rule out other etiologies of cerebellar involvement such as vascular or tumor.

Cerebral computed tomography, which is less sensitive than MRI, particularly in the posterior fossa, is often normal or shows cerebellar hemispheric hypodensity (owing to frequent artefacts in this region), with or without V4 compression. Little is known regarding the sensitivity of MRI in detecting cerebellitis, and data are restricted and based on isolated cases [14-16].

Differential diagnoses include EMAD, drug-induced inflammatory processes (overdose or chemotherapy), particular leukodystrophies affecting white and grey matter, lead poisoning [13, 14], X-ray histiocytosis, and invasive gliomas or lymphomas.

The prognosis for acute cerebellitis is generally favorable. Even patients with severe symptoms and increased intracranial pressure often recover completely, with no complications. Steroids are the first line of treatment when symptoms are moderate to severe [17]; however, most people will recover without steroids or other specific medication.

Antimicrobial therapy should always be investigated since ataxia can suggest viral encephalitis or bacterial meningitis [18, 19].

Aside from immediate problems that can be fatal, long-term neurological abnormalities following acute cerebellitis have been recorded in 6 to 33% of cases, depending on the study [20].

4. Conclusions

Recognizing ataxia in children can be difficult, which is

why a lumbar punction should be considered if infectious cerebellitis is suspected. It may be overlooked, particularly in very young children, and mistakenly linked to a lack of co-ordination. Physical examination and proper maneuvers are useful in identifying its clinical symptom. There are multiple causes of ataxia, each with a different consequence that can range from transitory and innocuous to exceedingly severe and scary. Most instances are serious; however, in severe cases, early detection and treatment are vital to avoiding possibly lethal consequences. Most people fully recover, however there is a risk of lasting disability.

Abbreviations

MRI	Magnetic Resonance Imagery
V4	Fourth Ventricle
CSF	Cerebrospinal Fluid
PCR	Polymerase Chain Reaction
CRP	C reactive Protein
ENMG	Electroneuromyogram
ROT	Osteotendinous Reflex
Brain CT	Brain Computed Tomography
EMAD	Acute Disseminated Encephalomyelitis

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Abdelhak Abkari: Project administration
Daoud Bentaleb: Resources
Wilhem Gilius: Resources

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Data Availability Statement

The data supporting the outcome of this research work has been reported in this manuscript.

Conflicts of Interest

The authors declare no conflicts of interest.

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