Clinical and radiology findings in acute childhood cerebellitis

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Authors: M. Fernández Hernando, A. Saiz Ayala, S. C. G. Costilla, E. Santamarta; Oviedo/ES
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Learning objectives

- To know the main clinico-etiological features in acute cerebellar childhood ataxia.
- To analyze the most common imaging findings in this entity and to show which have a higher diagnostic performance.
- To evaluate the clinical and radiological evolution of this patients.
- To analyze the clinical and radiological findings of the eleven cases of acute childhood hemicerebelitis published to date in the international literature.

Background

Hemicerebellitis is a rare entity only reported in less than fifteen cases in the literature in the pediatric age.

It is a cerebellar effusion involving only one of the two cerebellar hemispheres, with an homolateral ataxic gait clinic.

Ataxia may be congenital or acquired. Congenital ataxia is usually associated with central nervous system malformations. Ataxia can be classified as acute, episodic, or chronic. Episodic and chronically progressive ataxias are uncommon in childhood and are usually caused by inherited metabolic or genetic disorders.

Acute cerebellar ataxia, defined as unsteadiness of walking or of fine motor movement of less than 72 hours’ duration in a previously well child, is most common in pre-school children (2-4 years) but may be seen in older children and adolescents. Boys are more commonly affected.

Postinfectious cerebellitis presents with fast onset of gait abnormalities, ranging in severity from unsteadiness and a wide-based stance to complete inability to walk. Symptoms are maximal at onset and may be more severe in cases following varicella infection. The limbs are less affected than the trunk. Associated findings can include truncal instability, head titubation, intention tremor, dysmetria, and nystagmus.
Abnormalities of ocular movement, including ocular flutter (horizontal) and opsoclonus (multidirectional), are occasionally seen. A small minority of patients has associated cranial nerve palsies or long-tract signs. Mental status is normal. Acute cerebellar ataxia is not associated with fever, seizures, or other systemic upset in a high percentage.

Acute cerebellar ataxia usually resulting from postinfectious cerebellar demyelination, less commonly occurring as a result of direct infection of the cerebellum. Postinfectious cerebellar demyelination is thought to be an autoimmune phenomenon incited by infection or immunization, with subsequent cross-reaction of antibodies against cerebellar epitopes, although specific autoantibodies have only rarely been identified.

A history of antecedent illness in the 5 to 21 days before presentation is obtained in about 70% of patients.

Many infectious agents have been implicated in the pathogenesis of this entity such as mumps, measles, coxsackie virus, poliovirus or *Mycoplasma pneumoniae*.

Most cases are preceded by varicella. Rarely, the development of ataxia precedes the eruptive phase of varicella infection. The introduction of universal immunization against varicella is likely to render varicella-related cerebellar ataxia uncommon.

The differential diagnosis of acute cerebellar ataxia should include multiple etiologies. Recent immunizations should be noted, as should the child's general state of health in the weeks and months prior to presentation. Recurrent or persistent headache and vomiting or diplopia suggests an intracranial mass lesion and possible elevation of intracranial pressure. The history should therefore include inquiry as to antecedent or current symptoms of systemic infection, including fever, rash, and gastrointestinal upset. A common cause of acute ataxia is inadvertent or deliberate drug ingestion. Recent head or neck trauma should prompt consideration of vertebral artery dissection. A history of previous similar episodes in the patient or other family members should also be identified if present.

Imaging techniques allow us to exclude tumors, vascular lesions or abscesses and allow us to recognize the benign nature of this clinical syndrome in most children.

**Imaging findings OR Procedure details**
We are going to analyze the most significant radiological findings in acute childhood cerebellitis based on three cases reported in the Radiology Department of the Hospital Central de Asturias.

**CASE 1:**

A 5 year old girl presented fever (maximum 39ºC) associated with vomiting and mild diarrhea. Twenty-four hours later she is diagnosed of faringoamigdalytis and is prescribed antibiotic treatment. One day later she is admitted to the hospital with dizziness, lost of conscience and one episode of a few seconds long, of general stiffness associated with eyes deviation. Her personal and family morbid histories were unremarkable and there were no vaccination in the previous days.

The blood and urine chemistry, blood cultures (virus and bacteria), fundoscopy and electroencephalogram were normal.

During her stay in that hospital she suffered another two seizures, so she was referred to the Neuropediatric Unit of our hospital.

Examination revealed a mild left ataxic gait as well as a left dymetria, and no other focal neurologic abnormalities were found. Other physical evaluation was normal.

The laboratory tests were repeated with the following results: normal blood chemistry, CRL chemistry with 9 leucocytes/mm$^3$ (normal glucose and protein levels), positive CRL culture (Enterovirus), electroencephalogram, electromyogram and nerve conduction velocity were also normal.

The association between the clinics and the Enterovirus isolation in CRL made us suspect a cerebellar involvement, so a brain MR was made (one week after the onset of neurologic symptoms), which showed a high signal intensity on T2 and FLAIR-weighted sequences in left cerebellar hemisphere. Ventricular size and white matter were normal (Figure 1).

Clinical evolution was very positive, with gait and dysemetria improvement in a few days. In the clinical control four months later all the neurological symptoms had disappeared. The MRI control four months after showed a cerebellar atrophy on T1, T2 and FLAIR-weighted sequences and hippocaptation in that hemisphere in diffusion sequences (Figure 2).

**CASE 2**
A 9 years old boy presented to his hospital with a five days evolution history of frontal and occipital headache associating photophobia. This pain wakes him up sometimes in the night. The previous days he had suffered a gastroenteritis. In the Emergency Unit a Brain CT was performed because of the persistence and worsening of the headache and revealed a masa in the left cerebellum hemisphere, so he was referred to the Neuropediatric Unit of our hospital.

On admission the patient was oriented, his speech was fluent and all the neurological examination was normal except an important left hemiataxia being unable to stand up firmly. No nystagmus was observed.

The blood chemistry, blood and urine cultures (virus and bacteria), tumoral markers, PEV and PESS were normal.

The initial brain MRI showed high signal intensity on T2 and FLAIR in the anterior lobe of the left cerebellar hemisphere and cerebellar vermis associated with discrete vasogenic edema of the white matter which conditioned a moderate mass effect on the fourth ventricle, a discreet triventricular ventriculomegaly and partial effacement of the cistern of the quadrigeminal plate. There was also a striated pattern with alternating hyperintense and hypointense lines with linear enhancement of the cerebellar folia. (Figures 3 and 4). The diagnostic possibilities are rhomboencephalitis vs Lhermitte-Duclos.

High doses or intravenous corticoids and intravenous acyclovir were given prophilactically during ten and fifteen days respectively. Clinical evolution was good and at the moment of discharge the boy was able to walk normally although his gait skill was worse in fast turns and presented fine tremor in the left hand with a slight dysmetria.

The clinical control six months after the patient was absolutely asymptomatic. The MRI performed showed an important atrophy on the anterior lobule of the left cerebellar hemisphere (Figure 5).

**CASE 3:**

A 17 year old girl presented occipital headache, nausea, vomiting and fever feeling. Analgesic treatment was prescribed but the patient is admitted hours later in the emergency department for worsening of these symptoms and the appearance of unsteady gait.

Physical examination: horizontal rotary nystagmus in all gaze positions with unstable tandem drive to the right.
Blood count, biochemistry, coagulation, blood cultures (viruses and bacteria) and complete blood serology and CSF were normal.

Biochemistry and CSF culture: normal.

Cranial CT: normal.

Cranial MRI was performed showing a hypointense signal and poor differentiation of cortico-subcortical right cerebellar hemisphere and hyperintensity on T1 sequences and hyperintensity on T2 sequences, FLAIR and DW. Important leptomeningeal enhancement was found after administration of gadolinium in this cerebellar hemisphere and more discreet on the left. (Figure 6).

The most significant clinical and radiological findings of the cases reported to date in the literature are summarized in table 1.

**Images for this section:**
Fig. 1: Brain MRI in the onset of symptoms showed a high signal intensity on T2 and FLAIR-weighted sequences in left cerebellar hemisphere. Ventricular size and white matter are normal.
Fig. 2: The MRI control four months after showed a cerebellar atrophy on T1, T2 and FLAIR-weighted sequences and hipocaptation in that hemisphere in diffusion sequences.
**Fig. 3:** Brain MRI showed high signal intensity on T2 and FLAIR in the anterior lobe of the left cerebellar hemisphere and cerebellar vermis associated with discrete vasogenic edema of the white matter which conditioned a moderate mass effect on the fourth ventricle, a discreet triventricular ventriculomegaly and partial effacement of the cistern of the quadrigeminal plate. There was also a striated pattern with alternating hyperintense and hypointense lines with linear enhancement of the cerebellar folia.
Fig. 4: Brain MRI showed high signal intensity on T2 and FLAIR in the anterior lobe of the left cerebellar hemisphere and cerebellar vermis associated with discrete vasogenic edema of the white matter which conditioned a moderate mass effect on the fourth ventricle, a discreet triventricular ventriculomegaly and partial effacement of the cistern of the quadrigeminal plate. There was also a striated pattern with alternating hyperintense and hypointense lines with linear enhancement of the cerebellar folia.
Figure 5: Control brain MRI six months after showed an important atrophy on the anterior lobule of the left cerebellar hemisphere. The patient was absolutely asymptomatic.

Fig. 5: Control brain MRI six months after showed an important atrophy on the anterior lobule of the left cerebellar hemisphere. The patient was absolutely asymptomatic.
Fig. 6: Cranial MRI showing a hypointense signal and poor differentiation of cortico-subcortical right cerebellar hemisphere on T1 sequences and hyperintensity signal on T2 sequences, FLAIR and DW. Important leptomeningeal enhancement was found after administration of gadolinium in this cerebellar hemisphere (more discreet on the left).
Table 1: Clinical features on previous reported patients with hemiataxia

<table>
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<tr>
<th>Case</th>
<th>Age</th>
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<tr>
<td>Lester</td>
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<td>M</td>
</tr>
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<td>Jabbour</td>
<td>13 y</td>
<td>M</td>
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<td>9 y</td>
<td>M</td>
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<tr>
<td>Case 1</td>
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</tbody>
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Table 1: Clinical features on previous reported patients with hemiataxia.
Conclusion

Acute childhood hemicerebellitis is a rare entity that should be included on hemiataxies and acute ataxias differential diagnose.

RM is the gold standard, because is the best complementary imaging test valuing the posterior cerebral fosa. In addition, has an important role in the evaluation of potentially lethal complications requiring surgery and allows an accurate diagnosis over other differential diagnoses.

CT is an inadequate technique with very low performance in this entity.

The later cerebellar atrophy has been reported in some patients, even though it does not imply, in our current knowledge, motor or movement control disturbances. Later evolution use to be good.

Personal Information

References


