Acute pediatric cerebellitis and mutism. Case report and review of the literature

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ABSTRACT

Acute cerebellitis (AC) is a rare syndrome characterized by an inflammatory involvement of the cerebellum which can complicate infections or vaccinations. AC can be a life-threatening condition, presenting with speech disorders including long-lasting mutism, usually followed by dysarthria. Clinical examination, laboratory findings and Magnetic Resonance Imaging are essential for establishing an early diagnosis and initiating prompt and adequate treatment, thereby reducing morbidity and mortality. We report a case of a child with AC complicated by mutism and we present a review of the literature of pediatric cases with this condition.

Key words: cerebellitis, mutism, dysarthria, childhood

Introduction

Acute cerebellitis (AC) is an uncommon syndrome characterized by inflammatory involvement of the cerebellum, which can complicate infections or vaccinations. Its main manifestations are ataxia, a wide-based gait, dysmetria, tremor, fever, headache, nystagmus and speech disorders, including mutism. (1) Cerebellar mutism is a complete but transient loss of speech, usually followed by dysarthria, caused by direct lesions of the cerebellar vermis or hemispheres. (2) It is commonly reported as a complication after posterior fossa neurosurgery, while it is less frequent in cerebellar inflammation. (3) The neuroimaging gold standard for the diagnosis of AC is Magnetic Resonance Imaging (MRI), usually demonstrating altered signal intensity, swelling and abnormal diffusion weighted imaging of the cerebellum and surrounding areas. (4) Antibiotic, antiviral, antiedema and/or corticosteroid drugs are useful for treating AC, while rarely external ventricular drainage is necessary to reduce intracranial hypertension. (1) AC usually has a rapid onset and is self-limiting, but sometimes can be life-threatening. (5) Its symptoms and complications, especially mutism, can last for several months. We report a case of a child with AC complicated by mutism, and we present a review of the literature of pediatric cases with this condition.

Case report

A 6-year-old previously healthy girl presented with fever, diarrhea, seizures, neck stiffness, upward eye deviation and drowsiness. She was admitted to our Pediatric intensive care unit (PICU) (day 0), where she required two days of mechanical ventilation to control persisting seizures and respiratory complications. Cerebrospinal fluid (CSF) examination revealed 900 cells/mm3, protein 164 mg/dl and glucose 54 mg/dl. C-Reactive-Protein was increased (60.38 mg/L; normal value < 5 mg/L). Blood cell count and blood chemistry were normal. Suspecting viral encephalitis, therapy with Acyclovir was started. Electroencephalogram (EEG) showed beta and delta activity in medio-posterior brain regions, with right prevalence. Serological tests for cytomegalovirus, varicella-z virus, herpes simplex virus 6, adenovirus, rubella and mumps demonstrated past infections; bacterial and mycotic blood cultures and serology for toxoplasma gondii, cocusacke virus,
basic activity. A second MRI, performed on day 30, did not demonstrate significant differences from the first one. With persisting symptoms, a single photon emission tomography (SPET) was performed, showing the absence of cerebral and cerebellar regional deficit of perfusion. The child was discharged on day 34: her motor functions, walking and speech improved, she restarted drawing simple subjects but she still presented with dysarthria and equilibrium disturbances. At clinical follow-up, 6 months after the onset of symptoms, she was able to formulate sentences, although she still spoke slowly. Neurological examination was normal except for slight nystagmus laterally. MRI, performed at the same time, revealed the regression of cerebellar tonsillar dislocation; in addition, it showed slight widening of perifolial liquoral spaces and a minimal dilatation of the fourth ventricle (figure 1B). This study was approved by the Institutional Review Board (IRB) and written informed consent was signed by the child’s parents.

## Discussion
AC is an inflammatory syndrome which involves cerebellar structures usually bilaterally and symmetrically, as a consequence of viral infections or vaccinations. The main symptoms and signs are related to cerebellum dysfunction, associated with abnormal cerebrospinal fluid findings and an abnormal MRI. (1) AC is most frequent in childhood and is usually self-limiting; though sometimes, it can be a life-threatening event, leading to serious complications including death. (5) Its etiology is various, usually viral (herpes virus, rubella, Coxsackie virus); more rarely B. burgdorferi, Coxiella burnetii and Mycoplasma pneumonia have been involved. (5) MRI of the brain is the radiologic investigation of choice for the detection of inflammatory disorders of the cerebellum, and it is also useful for their follow-up. (4) Cranial Axial Computed Tomography (CAT) has been demonstrated to be often unrevealing. (4) Brain SPECT may be useful to better assess the neurological deficits as it shows marked reduction in cerebral perfusion. (6) Our case reveals some interesting characteristics because the child suffered from an episode of acute cerebellitis, likely consequent to an Echovirus infection, complicated by persistent mutism but without important alterations of SPECT findings. Only MRI showed herniation of the cerebellar tonsils, confirming the leading role of MRI in diagnosing AC. In the literature, we found 17 cases of pediatric acute cerebellitis presenting with mutism (table 1). (2,3,7-10) These cases concerned 12 girls and 5 boys with age range between 2 and 7 years. Mutism lasted between 3 days and 5 months (mean: 21 days) and was followed by dysarthria, with slow and difficult language recovery. The etiology was unknown in 13 cases (76.5%) and the involved agent was always Rotavirus, detected only in stool samples. All children underwent MRI examination, revealing, more frequently, hyperintense areas and swelling of the cerebellum in T2-weighted sequences. Only in one child was tonsillar herniation detected, as happened in our case. No other patients underwent a SPECT examination. In regard to treatment, only one child required external ventricular drainage due to intracranial hypertension, while everyone else received antibiotics, antiviral, antiedema and/or corticosteroid drugs. Follow-up reported in the literature differed according to timing and findings. Clinically, every child improved in his/her ability to walk and speak. Speech disorders often persisted, lasting several months, and perfectly fluent speech was not found at clinical follow-up. Radiological follow-up showed cerebellar atrophy in 16 children (94%).

In conclusion, we can state that AC is a dangerous syndrome, often complicated by long-lasting mutism affecting children’s quality of life. Clinical examination, CSF findings and MRI are essential for establishing an early diagnosis and initiating prompt treatment to reduce the morbidity and mortality associated with this condition.
Table 1. This table reports all acute pediatric cases of cerebellitis associated with speech disorders, such as mutism and dysarthria.

<table>
<thead>
<tr>
<th>REFERENCE</th>
<th>CASES</th>
<th>ETIOLOGY</th>
<th>SYMPTOMS</th>
<th>MUTISM DURATION</th>
<th>MRI</th>
<th>TREATMENT</th>
<th>FOLLOW-UP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Riva, Cortex 1998</td>
<td>F 4y 2m</td>
<td>n.r.</td>
<td>Gastroenteritis, Consciousness disturbance, Ocular dysmetria, Ataxia</td>
<td>4 w</td>
<td>Altered intensity, T2: ventricular dilatation, Edema</td>
<td>Acyclovir, Dexamethasone, Ceftriaxone, Mannitol, CSF shunt</td>
<td>11m: MRI: not completely resolved T2 images, CLINIC: mild hypotonia, deficient free verbal fluency, telegraphic spontaneous language, impoverished and elementary sentence construction</td>
</tr>
<tr>
<td>Papavasiliou, Pediatr Neurol 2004</td>
<td>F 3y</td>
<td>n.r.</td>
<td>Acute gastroenteritis, Ataxia and dysmetria, Involuntary choreo-athetoid type movements, Opioidotic posturing, Tremor, Inability to stand</td>
<td>2 w</td>
<td>Swelling, protruding tonsils</td>
<td>Acyclovir, Dexamethasone</td>
<td>6m: MRI: cerebellar atrophy, increase of the subarachnoid space of the posterior fossa and of the size of the fourth ventricle</td>
</tr>
<tr>
<td>Shihara, Brain Develop 2007</td>
<td>F 2y 7m</td>
<td>Rotavirus (faeces)</td>
<td>Diarrhea and vomiting, Fever, Convulsions, Consciousness disturbance, Equilibrium disturbance, Wide-base gait, Hypotonia</td>
<td>5 m</td>
<td>Normal –</td>
<td>Methylprednisolone</td>
<td>d 148: slow speech and dysarthria, right hand tremor, d 180: enlarged fourth ventricle and widened cerebellar sulci</td>
</tr>
<tr>
<td>Dimova, Pediatr Neurol 2009</td>
<td>F 7y 6m</td>
<td>n.r.</td>
<td>Fever, Vomiting, Dizziness, Headache, Consciousness disturbance, Hypotonia, Dysmetria, Altered reflexes, Laughing</td>
<td>2 w</td>
<td>Hyperintensity T2 and FLAIR</td>
<td>Cefotaxime, Ciprofloxacin, Methylprednisolone</td>
<td>6m: CLINICAL: Persisted movements and speech disturbances, MRI: marked cerebellar atrophy</td>
</tr>
<tr>
<td>Kuboda, BrainDevelopm 2010</td>
<td>M 3y 2m, F 2y 4m</td>
<td>Rotavirus (faeces)</td>
<td>Fever, Diarrhea and vomiting, Consciousness disturbances, Repeatedly flapped limbs, Aparic episodes, Seizures, Hypotonia, Intentional hand tremors, Ataxia</td>
<td>1 m – 16 d</td>
<td>DWI: hyperintensity</td>
<td>Methylprednisolone – Carbamazepine, Phenobarbital, High-dose Ig, Methylprednisolone</td>
<td>5m: CLINICAL: Persisted movements and speech disturbances, MRI: marked diffuse cerebellar atrophy, 3m: CLINICAL: persistened slurred explosive speech, 6m: MRI: slight cerebellar atrophy</td>
</tr>
</tbody>
</table>

d, days; D, dysarthria; F, female; M, male; m, months; Mt, mutism; n.r., not reported; w, weeks; y, years.
REFERENCES