Case report

Transient mutism, pathological laughter and cerebellar ataxia after primary varicella-zoster virus infection

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SUMMARY

Acute postinfectious cerebellitis is characterised by inflammatory involvement of the cerebellum and consequent cerebellar dysfunction, usually presenting a benign and self-limited course. Common symptoms described in the literature include ataxia, peripheral facial paralysis, aphasia, spinal dysfunction and eventually hydrocephalus. We describe a rare presentation of acute cerebellitis after varicella, in a 6-year-old child, who presented with ataxia, dysmetria, mutism and pathological laughter.

BACKGROUND

The main agents of acute postinfectious cerebellitis are Epstein-Barr virus, rubella, measles, coxsackie and varicella-zoster, the latter being the most frequent, with an incidence of approximately 1 in 4000 cases of varicella.¹

Typically, cerebellitis is characterised by the gradual onset of symptoms such as fever, tremor, trunk and limb ataxia, dysarthria, headache, nausea, vomiting and altered consciousness.¹ Although usually benign and self-limited, it can present severe complications.¹

Cerebellar mutism is associated with lesions in the vermis or cerebellar hemispheres and is usually characterised by complete but transient aphasia, followed by a period of dysarthria.² It most frequently follows posterior fossa surgery, trauma or cerebellar haemorrhage, but it has been rarely related to infectious or postinfectious cerebellar syndromes.^{2,3}

Pathological laughter corresponds to a disturbance of the expression of emotions, characterised by episodes of uncontrolled laughter with no apparent relation to a stimulus or triggered by an inappropriate stimulus.⁴ It is a rare clinical manifestation of brain-pons-cerebellar pathways that control laughter, most often seen in vascular lesions of the descending corticobulbar tract, multiple sclerosis, amyotrophic lateral sclerosis, cerebellar subtype of multiple system atrophy, gelastic epilepsy, cerebellar angle tumours and spinal cord tumours, typically adult conditions.⁴

Only one case of pathological laughter was described in children, although in situations of posterior fossa syndrome, mainly after midline cerebellar tumours surgery, children generally present emotional lability or maladaptive behaviour.⁴

We report this case in order to highlight a less common form of acute postinfectious cerebellitis, which requires follow-up and investigation of possible neurological sequelae.

CASE PRESENTATION

We present the case of a 6-year-old girl child, diagnosed with varicella 6 days before admission, who came to the emergency room due to vomiting, headache and ataxia since the previous day. She had no relevant personal or family background and no history of head trauma or drug intake. On admission, physical examination revealed generalised vesicular rash and some crusted papules, gait severely impaired (wide-based, unsteady), impaired coordination of voluntary movements with dysmetria on finger-nose testing, inappropriate laughter and mutism; verbal comprehension and ability to obey simple orders were preserved.

INVESTIGATIONS

The evaluation included normal hemogram and biochemistry findings; urine analysis was negative for barbiturates, benzodiazepines, carbamazepine, cocaine and phenytoin. No respiratory viruses were detected by FilmArray Respiratory Panel. Lumbar puncture revealed normal cell count ($5 \times 10^9/L$ white blood cells), normal protein and glucose content, as well as undetectable neurotropic viruses (FilmArray Meningitis/Encephalitis). Blood and cerebrospinal fluid cultures were sterile. She also performed an electroencephalogram and a CT scan that were normal, and later MRI (3 weeks after symptoms onset) that did not reveal focal brain lesions, namely cerebellar, nor alteration of the extra-axial spaces.

OUTCOME AND FOLLOW-UP

Treatment was not necessary because symptoms gradually improved, with resolution of mutism by D7 and ataxia by D10, but dysarthria was evident up to 1 month after discharge.

She was posteriorly reevaluated by europaediatrics, presenting normal psychomotor development and neurological examination.

DISCUSSION

The acute post-varicella cerebellitis hypothesis was established given the epidemiological and clinical context. However, this child presented a

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combination of atypical symptoms, suggestive of generalised cerebellar lesion with ataxia, dysmetria, mutism and pathological laughter. Other possible causes for pathological laughter were ruled out as there were no signs of focal lesions on neurological examination and electroencephalogram, also CT scan and MRI were normal. Only one similar case was described in literature.⁴ In that case, the mutism was also transient, followed by a long period of dysarthria, verbal comprehension was preserved even during the mutism phase, movements were ataxic and there were occasional episodes of pathological laughter. It should be noted the surprisingly low number of children with severe speech

disturbances in the course of severe acute cerebellitis, given the incomplete maturation of their cerebellar–cerebral circuits, and hence a greater vulnerability to speech dysfunction.⁴

Spontaneous recovery may occur, but the follow-up is essential, because of possible neurological sequelae, such as obstructive hydrocephalus.⁴

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Learning points

- ▶ Although a common condition in childhood and generally with a good prognosis, acute postinfectious cerebellitis may also give rise to severe global cerebellar dysfunction, leading to some short-term and long-term sequelae.
- ▶ Inappropriate laughter and mutism are not usually valued as manifestations of disease in children. This case report shows us that these two signs should be considered as possible clinical manifestations of postinfectious cerebellar disease, revealing the role of the cerebellum in the multiple domains of neurological function.

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