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Case report

Cerebellar fits in the 2000s

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Abstract

Acute compression on the brainstem or acute increase in intracranial pressure may induce non-epileptic events varying from tonic seizures to axial rigidity with motor automatism, sometimes clearly characterized by decerebrate or decorticate paroxysmal posturing. The EEG correlate is characterized by diffuse asynchronous slow waves of variable amplitude. The mechanism behind such events, known as "cerebellar seizures or fits", is linked to cerebellar herniation and brainstem compression and is not of cortical origin. Misrecognition of such entity may entail an incongruous therapeutic intervention in a life-threatening situation. We describe two emblematic paediatric cases of cerebellar fits caused by diffuse oedema and brainstem compression: a 10-year-old girl with acute disseminated encephalomyelitis (ADEM) and a 2-year-old girl with severe respiratory distress symptomatic of Fallot tetralogy. We also describe the EEG correlate recorded during the events.

Keywords: Cerebellar fits; Non-epileptic seizures; EEG; Intracranial hypertension

1. Introduction

In 1871 John Hughlings Jackson described the case of a 5-year-old boy with "tetanus-like seizures" secondary to a tubercular abscess of the cerebellum, characterized by opisthotonic posturing and preserved consciousness and lasting between 3 and 4 min. Several other cases were later published but only in 1880 a link between these types of seizure and compression of brainstem structures was hypothesized [1]. At the turn of the century the concept of "decerebrate rigidity" was also introduced under the influence of the British School of Experimental Physiology, largely based on animal models [1]. The few published human cases were characterized by episodes of tonic limb posturing with extension of the lower limbs and flexion and pronation of the upper limbs. Consciousness could be maintained during the attacks however respiratory irregularity was common and could be fatal. In more recent neurology textbooks cerebellar seizures are often only briefly described, however, while assuredly not a common phenomenon, they are still something that a child neurologist must be very aware of: immediate differential diagnosis with an epileptic seizure might mean the difference between the useless administration of antiepileptic drugs and the life-saving administration of mannitol.

2. Case 1

Patient 1 was a girl first seen in another hospital at the age of 10 for repeated episodes of headache with photophobia and vomit. Fundus oculi examination

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showed bilateral papillar edema. A brain MRI scan showed symmetrical white matter abnormalities over both hemispheres, more evident over the posterior parietal and occipital regions, the internal and external capsules, the subtentorial mid-cerebellar region and in the bulbar region. She was transferred to our hospital two days later for suspected demyelinating disease. Rachicentesis showed a liquoral pressure of 32 mmHg, a cell count of 0 and an increase in protein levels (71 mg/dl), which hinted towards an acute disseminated encephalomyelitis (ADEM). The following day she had severe headache and drowsiness and repetitive seizures lasting between 20 and 30 s, characterized by tonic axial contractions tending towards opisthotonic posturing, tonic upper limb contraction, lower limb hyperextension, with kicking automatisms and preserved consciousness At seizure onset the upper limbs were flexed, while in a later phase they were extended and pronated (Fig. 1). The EEG failed to show ictal etpileptic activity and demonstrated low-to medium amplitude polymorphic diffuse delta activity. Cerebellar seizures were suspected. A brain CT scan showed diffuse oedema and brainstem compression. She was intubated, administered mannitol and glucocorticoids. She was then transferred to the ICU and an intracranial pressure monitor was surgically placed. Her clinical condition gradually improved and 4 days after admittance she was extubated. The following day she underwent an MRI scan which confirmed the presence of diffuse white matter abnormalities, and thus the diagnosis of ADEM. After 2 years of follow-up she still shows full recovery from the acute event.

3. Case 2

Patient 2 was a two-year-old girl first seen in our hospital in relation to a Fallot tetralogy with severe infundibular stenosis and repeated episodes of respiratory distress. Given her adoption from a foreign institution one month prior to admission, her clinical history before contact with our hospital was unclear. Neurological examination at admittance was within limits. During hospitalization she had several episodes of asphyxia. In day 9, following severe respiratory distress, she had alteration of consciousness and was sent to the ICU where she had multiple seizures characterized by opisthotonic posturing, extension of the lower limbs and flexion/pronation of the upper limbs. Her EEG showed a basal activity of very low amplitude, and the seizures were associated with polymorphic delta activity with superimposed muscle artifacts (Fig. 2). A brain CT scan showed severe cerebral oedema, with extrusion of the cerebellar tonsils in the foramen magnus. Even though a ventricular shunt was immediately placed, intracranial pressure increased from 15 to 29 mmHg. Mannitol and 24-h therapeutic hypothermia were administered, but she still had severe metabolic acidosis. Her clinical condition gradually worsened, as did her basal EEG activity. Cerebral death was ascertained 17 days after admittance.

4. Discussion

Cerebellar fits are an ill-defined entity, there being no clear consensus among physicians in distinguishing the paroxysmal events described by Jackson from other signs of intracranial hypertension. Decerebrate posturing (DP) for example (also referred to as decerebrate rigidity), is caused by midbrain lesions and is manifested

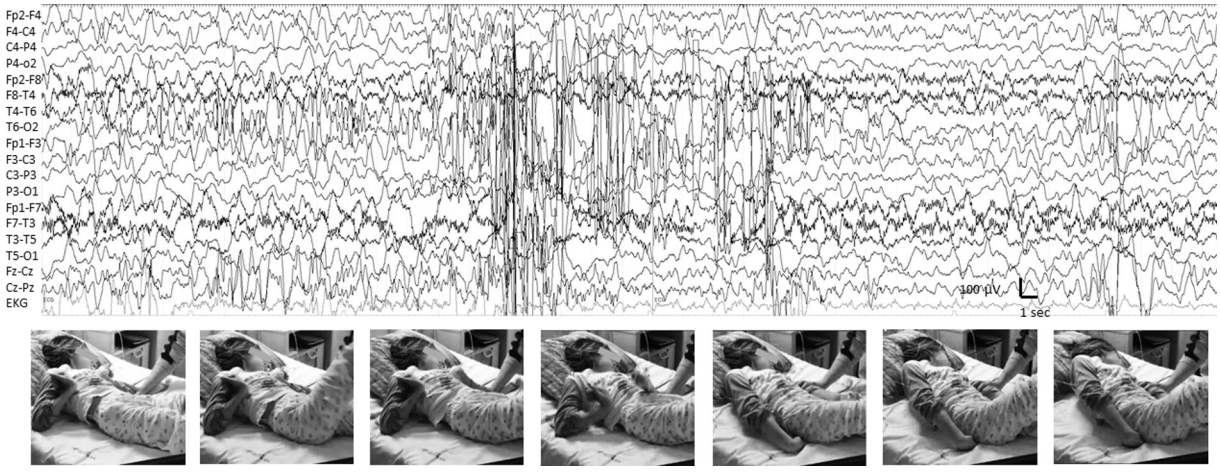

Fig. 1. Low-to medium amplitude polymorphic diffuse delta activity. The image sequence shows the clinical correlate, characterized by tonic axial contraction followed by tonic upper limb contraction, lower limb hyperextension, with kicking automatisms and preserved consciousness Of notice, upper limbs were initially flexed, while in a later phase they were extended and pronated.

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