The wobbling titan: Acute post-infectious cerebellar ataxia

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SUMMARY

Acute post-infectious cerebellar ataxia (APCA) is a transient neurological disorder commonly observed in children following viral or bacterial infections. This case report describes a five-year-old girl presenting with sudden-onset unstable gait, without focal neurological deficits or systemic symptoms, after a recent febrile illness. Clinical examination revealed a wide-based ataxic gait and bilateral lower limb weakness. Investigations showed no evidence of infection or other neurological pathology. The patient was diagnosed with APCA and managed supportively, leading to complete resolution of symptoms. This case underscores the importance of considering APCA in children presenting with acute ataxia, particularly following recent infections, and highlights the need for thorough evaluation to exclude alternative aetiologies. Early recognition and appropriate management are crucial for favourable outcomes in patients with APCA.

INTRODUCTION

Cerebellar ataxia frequently caused by impairment of cortical signals from the cerebellum that manifest as incoordination in movement, gait abnormalities, changes in speech and eye abnormalities.[1] Acute cerebellar ataxia is known as sudden benign self-limited neurological complications that has temporary cerebellar dysfunction less than 72 hours of duration, in a previously healthy person.[2] It typically occurs in children less than 6 years old with an incidence of 1 in 100,000 to 500,000 and is commonly associated with post infectious autoimmune mechanism following infection from varicella zoster, Coxsackievirus, Echovirus, Enteroviruses, Epstein–Barr virus (EBV), Herpes simplex virus I, Measles virus, Mumps virus, Borrelia burgdorferi, and severe acute respiratory syndrome Coronavirus 2 (SARS–CoV–2).[4] However, the relative mechanism underlying the pathophysiology of cerebellum disruption is still unknown.4 Acute post-infectious cerebellar ataxia (APCA) is usually characterized by sudden ataxia following a period of illness and may present with other neurological sign and symptoms such as nystagmus, hypotonia, tremor or scanning of speech.[5] It is a diagnosis of exclusion after excluding other causes of ataxia with a thorough history that supports recent infection, clinical examination and biochemical tests. Imaging and lumbar puncture are not specific and can be deferred with watchful waiting under a close neurological follow-up.[4,6] Recovery varies from more than 2 weeks to months and complete recovery may take up to several months to years.[6]

CASE PRESENTATION

A five-year-old girl was first seen at the primary clinic during an acute presentation. She was brought in by her mother, who reported a complaint of unstable gait for the past two days. This episode was the first occurrence, and its sudden onset was noticed by the mother after the child woke up from sleep. The mother denied any worsening of the child's unstable gait. There were no speech abnormalities, dysphagia, headaches, gastrointestinal issues, or urinary/bowel incontinence reported. Additionally, there were no seizure episodes, traumas, fevers, or syncopal attacks. The child appeared to be active as usual with a normal appetite. The mother denied any clumsiness or recurrent falls prior to this incident. However, there was a history of fever with upper respiratory tract symptoms two weeks before the current presentation, lasting only five days. The child was treated symptomatically with paracetamol and subsequently recovered well.

Further history revealed that the child was born via emergency lower segment caesarean section (LSCS) due to failed induction at term, with a good birth weight. There was no prolonged admission following birth. However, the child had a history of meningitis at two months old, with no resulting neurological impairment, and she recovered well thereafter. She is immunocompetent, and her immunizations are up to date. There was initially noted to be a delay in speech until three years old; however, the child is now able to speak in four to five word sentences. Otherwise, gross motor, fine motor, and social skills are normal.

Returning to her current examination, she appeared well, with a normal colour, no respiratory distress, and no dysmorphic features. No saccadic abnormalities or meningeal signs were observed. During her gait examination, it was noted that the child had an unstable, wide-based ataxic gait. Vital signs were normal, and her height and weight were within the normal range. Examination of the throat, lungs, and cardiovascular systems revealed no abnormalities. Due to the child's lack of cooperation, cranial nerve and cerebellar examinations were not performed. However, no muscle atrophy was observed. Examination of the central nervous system in both the upper and lower limbs demonstrated a power of 4/5 in the upper limbs and proximal lower limbs and 3/5 in the distal lower limbs bilaterally. Both the upper and lower limbs had normal tone, reflexes, and sensation, with no fasciculations observed. The provisional diagnosis was acute cerebellar ataxia, and the child was referred to the emergency department for further evaluation.

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Upon admission, the child was afebrile, alert, and active. Her ataxia was observed to be gradually improving, and no new neurological symptoms developed during her hospital stay. Initial neurological assessment showed muscle strength graded at MRC 3–4/5; however, this was likely due to coordination difficulties from ataxia and limited cooperation during examination, leading to a false impression of muscle weakness. The child's progressive clinical improvement supported that the apparent weakness was functional and secondary to ataxia, rather than indicative of a primary neurological deficit. Throughout her admission, she demonstrated consistent neurological findings, including a wide-based ataxic gait without clonus or upper motor neuron signs. Power and tone were preserved, and no cranial nerve deficits were observed. Despite initial unsteadiness, she was able to climb onto the bed, walk with support, and eventually ambulate with minimal assistance. Her vital signs remained stable, and no signs of clinical deterioration were noted. Basic laboratory work-up including full blood count, renal and liver function tests, inflammatory markers (C-Reactive Protein, Erythrocyte Sedimentation Rate), and electrolytes were performed and yielded normal results. These findings helped rule out infective, inflammatory, or metabolic causes, further supporting a diagnosis of exclusion. In view of the reassuring progression, absence of red flags, and results indicating a ruling out or resolved infective process, neuroimaging was deemed unnecessary at that point.

The paediatric neurology team decided not to perform neuroimaging or lumbar puncture during the acute phase because of the clear signs of clinical improvement and the absence of any concerning features (such as altered consciousness, severe headache, focal deficits, or signs of increased intracranial pressure). She was diagnosed as Acute post infectious cerebellar ataxia (APCA) supported by the improvement of her motor exam over time reinforcing the likelihood of a pure cerebellar ataxia rather a concurrent motor pathology. She was managed conservatively with paracetamol and regular clinical monitoring. Physiotherapy was initiated during the admission and continued as outpatient post-discharge. A 4-day inpatient observation was sufficient to ensure symptom stabilization. The child achieved full recovery within two weeks of discharge. At the three-month follow-up, she remained clinically well with no residual ataxia. At the six-month consultation conducted via phone, the child was reported to be well with no residual impairment.

DISCUSSION

Acute post-infectious cerebellar ataxia (APCA) emerges as the primary consideration in children presenting with acute ataxia.[3] APCA may arise following viral or bacterial infections, or even post-vaccination, typically manifesting within days to weeks.[7] Notably, studies have demonstrated autoimmune reactivity targeting cerebellar Purkinje cells post-infection, shedding light on the underlying pathophysiology. However, the precise mechanisms driving cerebellar disturbance remain elusive.[4]

While cerebrospinal fluid (CSF) analysis has been investigated in cases of APCA, its diagnostic yield appears limited, with lumbar puncture generally warranted only in cases of suspected meningitis.[8] Life-threatening or surgical pathology as an etiology for acute ataxia in children is rare, but it is important to remain vigilant to avoid missing the diagnosis. Life-threatening etiologies of ataxia can be classified into four categories: strokes, neoplasms, infections and/or inflammations, and toxic ingestions.[9]

For instance, ataxia due to a brainstem lesion, cerebral hemorrhage, brain infarction, or posterior fossa tumors (such as medulloblastoma or cerebellar astrocytoma) is usually accompanied by warning neurological signs and symptoms such as progressive headache, vomiting, altered consciousness, papilledema, cranial nerve involvement, or long tract dysfunction. In this case, none of these worrisome signs and symptoms were present apart from the ataxia.[9]

Moreover, the presentation of ataxia was acute and non-progressive, and upon close observation, her condition was improving. These characteristics are highly suggestive of APCA and are unlikely to be associated with structural lesions, which usually cause a progressive or steadily worsening course of neurological deficits rather than spontaneous recovery. Therefore, a surgical mass lesion or cerebellar tumor was deemed clinically unlikely, eliminating the urgent need for neuroimaging. On the other hand, toxic ingestion and trauma were considered the least likely based on the history and normal systemic examinations.[9]

Furthermore, a thorough differential diagnosis of acute cerebellar ataxia is essential, encompassing conditions such as Guillain-Barré syndrome, acute cerebellitis, acute disseminated encephalomyelitis (ADEM), and labyrinthitis. It is crucial to distinguish APCA from acute cerebellitis, as the latter represents the most severe end of a continuous clinical spectrum originating from APCA.[9] In general, children with acute cerebellitis present with more severe symptoms such as vomiting, fever, altered mental status, other signs of meningeal irritation, or seizures, and appear more ill compared to those with APCA.[9] In such cases, neuroimaging such as brain MRI typically shows inflammatory changes and may require urgent intervention.

Opsoclonus-myoclonus syndrome (OMS) is another immune-mediated paraneoplastic encephalopathy that may be difficult to distinguish from APCA in the early stages. However, unlike APCA, OMS does not show rapid improvement and is usually accompanied by additional symptoms such as feeding difficulties, regression, sleep disturbances, rapid saccadic eye movements, and paroxysmal movements. Hence, if there is minimal improvement with paroxysmal limb or eye movements, it is essential to consider OMS.[9]

Immune-mediated forms such as acute disseminated encephalomyelitis (ADEM) are typically severe at onset and occur in the context of viral or post-viral infections. The most typical symptoms of ADEM include bilateral or unilateral pyramidal signs (60–95%), acute hemiplegia (76%), ataxia (18%–65%), and cranial nerve palsies.[9]

Guillain-Barré syndrome (GBS) is another immune-mediated condition that presents with symmetrical weakness and progressive ataxia. Younger children may present with

symptoms such as meningeal signs, leg pain, vomiting, and agitation, while neurological examination reveals reduced or absent muscle stretch reflexes. Additional symptoms such as oculomotor paresis may increase suspicion for the variant of GBS known as Miller-Fisher Syndrome (MFS).[9]

Labyrinthitis presents similarly to APCA but is distinguished by additional symptoms such as vertigo, more severe systemic illness, and prominent vomiting. While nystagmus can occur in both conditions, it is typically more pronounced and persistent in labyrinthitis. In contrast, in APCA, if nystagmus is present, it is usually mild and not a dominant feature.[9]

Distinguishing among these entities necessitates meticulous history-taking, physical examination, and further investigative workup.[10] Neuroimaging, such as brain MRI, was not performed in our case because APCA usually shows no abnormalities on MRI. However, severe forms, such as acute cerebellitis, can present with neurological and systemic symptoms along with cerebellar or meningeal inflammatory changes on MRI.[6]

APCA, particularly in children, is often a clinical diagnosis based on a history of rapid-onset ataxia following a prodromal viral illness, with exclusion of serious or life-threatening conditions.[7] Neuroimaging is typically not required unless symptoms are atypical or concerning.[7] In addition, the clinical decision to avoid neuroimaging was reinforced by ongoing improvement during observation, supporting a transient and functional disturbance rather than a structural lesion. This approach is supported by evidence from Nussinovitch et al., which recommends neuroimaging only in cases where there is no improvement in 1–2 weeks, and Segal et al., who concluded that observation is safe in typical APCA cases in young children with recent viral illness and no additional neurological findings.[2, 5] These evidence-based recommendations support the decision of the clinician to carefully monitor the patient in the hospital for any new neurological signs or deterioration.

The child showed significant clinical improvement, suggesting a transient dysfunction rather than a structural lesion, thus making imaging unnecessary. A conservative approach was adopted and continuously reassessed to avoid unnecessary sedation and imaging, provided the child's safety was not compromised. In this case, the absence of focal neurological signs, negative blood investigations for infection, and the patient's uneventful clinical course along with complete resolution of the ataxic gait after discharge support the diagnosis of acute post-infectious cerebellar ataxia (APCA), which typically requires only conservative symptomatic treatment and regular follow-up to ensure full recovery.[6]

Empirically, there is still a lack of strong evidence on the effectiveness of corticosteroids or IVIG as treatment for APCA.[1] Nonetheless, several case reports have noted marked improvement following treatment with corticosteroids or IVIG. These considerations may be discussed on a case-by-case basis in the absence of consensus protocols. Furthermore, some research has indicated that autoimmune cerebellar ataxia may be effectively treated with IVIG. Therefore, clinicians may consider steroids, antivirals, or IVIG in cases with persistent symptoms, signs of inflammation, or suspected immune-mediated pathology.[1]

While APCA stands as the leading cause of acute ataxia in children, its association with various infections underscores the importance of vigilance among clinicians. Nevertheless, before reassuring families and initiating supportive care, a thorough evaluation to rule out more ominous pathologies remains paramount.

CONCLUSION

In summary, acute post-infectious cerebellar ataxia (APCA) is a common cause of acute ataxia in children, often triggered by viral or bacterial infections. While the exact causes are not fully understood, autoimmune reactions targeting cerebellar cells post-infection are suspected.

Diagnosing APCA relies on thorough clinical evaluation and ruling out other possible causes. In our case, the absence of serious symptoms, coupled with negative test results and the child's swift recovery, supported the diagnosis of APCA.

Early recognition and proper management led to the child's complete recovery, emphasizing the importance of prompt diagnosis and follow-up for children with acute ataxia. Clinicians must remain vigilant for APCA in such cases, ensuring thorough evaluation before reassuring families and providing supportive care. Further research is needed to better understand APCA and improve treatment strategies.

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