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ACUTE CEREBELLAR ATAXIA: AN UNUSUAL PEDIATRIC CASE

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☐ **Abstract—Background: Acute cerebellar ataxia is a clinical syndrome with sudden onset of uncoordinated gait and normal mental status in young children. Although it has a benign clinical course, it often requires an exhaustive diagnostic work-up in order to rule out potentially life-threatening etiologies that present similarly. The wide differential encompasses causes from infections, brain masses, drugs, toxins, trauma, paraneoplastic syndromes, as well as hereditary or congenital disorders. Case Report: We report on a 4-year-old boy with recent hand-foot-mouth disease who presented with acute cerebellar ataxia. In addition to his marked truncal ataxia and wide-based, staggering gait, he had slowness of speech, which is not commonly reported with this condition in the literature. Why Should an Emergency Physician Be Aware of This?: Emergency physicians have a unique role in being first to evaluate pediatric ataxia and can make a significant impact on identifying potentially fatal mimickers of acute cerebellar ataxia. This article will attempt to outline major diagnostic considerations in order to aid emergency physicians through their clinical approach.** © 2016 Elsevier Inc.

☐ **Keywords—pediatric; cerebellar ataxia**

INTRODUCTION

Acute cerebellar ataxia (ACA) presents infrequently in the pediatric setting; it is characterized by lack of coordination of movement that is not due to paresis, alteration in tone, sensory loss, or presence of involuntary movements in a relatively well-appearing child (1). ACA accounts for 30%–50% of all childhood ataxia cases (2). Despite being the most common cause of childhood ataxia, its similarity to other serious illnesses makes it a diagnosis of exclusion. That differential diagnosis includes intracranial tumors, toxic exposures and ingestions, trauma, acute cerebellitis, opsoclonus-myoclonus, and cerebellar stroke. These conditions have high morbidity and mortality rates (3,4). This article provides insight into a case of ACA and offers a diagnostic approach in an emergency or pediatric inpatient setting. Additionally, our patient presented with a slow rate of speech, which is uncommon with ACS and has not been reported in the literature.

CASE REPORT

A 4-year-old boy with no significant medical history presented with instability of gait beginning 2 days before admission to our hospital. His gait was severely unsteady, with a tendency to veer to the right that was not modified in any way. He denied head trauma. He was reported to have normal mental status with exception of slowed speech. He had recently had hand-foot-mouth disease diagnosed on history and physical findings by his primary care provider, which resolved 10 days before admission. He had been taking loratadine for seasonal allergies but did not have access to other substances. There was no

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history of focal weakness, seizure activity, fevers, sleep abnormalities, travel, sick contacts, dizziness, or headache. On his physical examination, the vital signs were normal for age and specifically were reported as temperature 98.7°F, pulse 92 beats/min, respiratory rate 18 breaths/min, blood pressure 105/60 mm Hg, and pulse oximetry 99%. His pupils were equal, round, reacted to light, and were 3 mm bilaterally. His ear examination showed bilateral tympanostomy tubes. The tympanic membranes were nonpurulent and were not bulging, the canals had no discharge or foreign bodies bilaterally. The mastoid was nontender and not boggy bilaterally.

He exhibited marked truncal ataxia when sitting unassisted and a wide-based, staggering gait with difficulty standing independently. He had an inclination to fall forward without ability to stop himself. He had bilateral cerebellar signs of dysmetria and heel-shin incoordination. He did not exhibit nystagmus, proptosis, or dysdiadokinesia or pronator drift. There was no report of loss of sensation and on physical examination there was no sensory deprivation. There were no deficits of the cranial nerves as tested (PERRL [pupils equal, round, reactive to light], EOMI [extraocular movements intact], no facial weakness/droop/asymmetry, no loss of facial sensation, no hearing deficit to whisper test, no deviation of uvula, no tongue deviation, and shoulder shrug intact), and there were no signs of meningeal irritation. The deep tendon reflexes were normal and strength was 5/5 in all extremities. He is able to form words appropriately, although he spoke more slowly than normally. Specifically, parents and examiner appreciated prolonged phonation of vocabulary words. He exhibited no expressive or receptive aphasias. The remainder of the physical examination was unremarkable.

Laboratory analyses (complete blood count and comprehensive metabolic profile) revealed a blood glucose of 78 mg/dL (normal range, 65–99 mg/dL), a relatively low white blood cell count (4.5×10^9/L), and normal hemoglobin. Erythrocyte sedimentation rate, serum urea, and urinary analyses (including vanillylmandelic acid [VMA] and homovanillic acid [HVA] levels) were normal. Rapid urine drug screen was negative. An initial computed tomography (CT) and subsequent sedated magnetic resonance imaging (MRI) scans were both reported as normal. Lumbar puncture was performed (opening pressure was not measured), and cerebrospinal fluid (CSF) had two white blood cells (100% lymphocytes), two red blood cells, and normal glucose (49 mg/dL) and protein (15 mg/dL); CSF culture was sterile. An Enterovirus polymerase chain reaction was sent and was negative.

The child was admitted with the diagnosis of ataxia and was hospitalized for 4 days after the CT was completed. With supportive care, there were signs of clinical improvement of gait instability and he was able to ambulate unassisted by discharge. The diagnosis of ACS was made on the basis of the clinical findings and the negative laboratory and imaging studies during the course of hospitalization. He received physical therapy for several months after discharge and by 6 months after discharge he had fully recovered.

DISCUSSION

"Pure" ACA is defined as sudden or "explosive" onset with predominant symptoms of truncal ataxia uncoordinated gait and normal mental status that is not associated with fever, seizures, or other systemic signs (5). It is most commonly seen in children aged 2–4 years old and almost exclusively in those younger than age 6 years. While it is a relatively benign cause of ataxia and diagnostic work-up is generally negative, studies have shown that normal MRI in ACA presentations might be present despite an underlying condition with potentially fatal outcomes (6). Due to the wide differential diagnosis of ACA, the diagnostic work-up may be exhaustive.

With every presentation, an accurate and thorough history of the present illness is essential for understanding the severity of the illness and establishing a working diagnosis. Questions should focus on recent immunizations, illnesses, signs of increased intracranial pressure (headache, vomiting, diplopia), drug ingestion, and recent head or neck trauma, as the second and third most common causes of ataxia are intoxication and intracranial masses. Most often associated with ACA is recent infection with viral agents, such as varicella, Epstein-Barr virus, Echovirus, Enterovirus (Coxsackievirus), 1–3 weeks before onset of symptoms (2). This was the case with our patient, who recently had hand-foot-mouth disease, which is usually caused by Coxsackievirus A16 virus or less commonly Enterovirus 71 virus. It is thought to be due to a postinfectious cerebellar demyelination phenomenon.(7) Prognosis of ACA with this association is generally good (7). Symptoms of intracranial pressure or a recent history of trauma suggest a more severe etiology and warrant an immediate imaging study. Additionally, questions about previous episodes in the child or family history are important to rule in or out congenital or hereditary differential diagnosis. Table 1 lists causes ataxia frequently identified in children (5,8,9). Familiarity of these causes can help guide a physician in the working diagnosis, particularly while formulating questions to elicit a clear history of present illness and the subsequent work-up.

Physical examination should focus on evaluating the patient's mental status in which a normal mental status helps differentiate from more serious conditions and favors a diagnosis of ACA. Providers should evaluate