ILLUSTRATIVE CASE

Occipital Epilepsy, the Great Mimicker: Case Report and Review for the Pediatrician

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CASE 1

The patient is a 4-year-old ex-28 week boy with developmental delay who presented to the emergency department with headache, altered mental status, abdominal pain, and bilious emesis. He was hard to wake the morning of presentation and was less interactive than normal but opened his eyes and pointed to his head and abdomen when asked if he had pain.

He had been having similar episodes of pain, vomiting, and depressed mental status associated with body tensing and nonrhythmic shaking since 2 years of age, and the episodes were now occurring monthly. Episodes typically began in the morning, with several hours of pain and 1 to 2 days of somnolence. Occasionally, his whole body would tense without tongue biting or loss of urine. He recovered completely to his baseline between episodes. His mother could identify no triggers. His development had always been globally delayed, but he continued to make progress. He had an unremarkable NICU course, and his family history was unknown because he had lived with his foster mother since discharge.

He had been admitted twice for these episodes: once he was treated for dehydration and diagnosed with constipation on the basis of abdominal radiograph results. The second time, malrotation and brain mass were ruled out with an upper gastrointestinal series and brain MRI, the results of metabolic lab work were negative, and he was diagnosed with a viral syndrome.

On current presentation, he was pale, tachycardic, and afebrile with otherwise normal vitals. He had delayed capillary refill and was rousable only to tactile stimulation. He was diffusely hypotonic with 1+ reflexes but no focal deficits and had an abdominal examination with benign results. His blood count showed a slight leukocytosis of 15×10^9/L with an 88% concentration of neutrophils, and his metabolic panel showed a low bicarbonate level of 16 mmol/L without an anion gap; venous gas also showed metabolic acidosis with a pH of 7.3, and the results of urinalysis and urine toxicology were negative. He received a fluid bolus and was admitted for further rehydration and workup, including lumbar puncture.

CASE 2

The patient is a 6-year-old, normally developing girl who presented to the neurology clinic with episodic headaches and altered mental status. The first episode occurred at 5 years old, when she was riding in a car with her mother. She yelled out suddenly, and her mother noted her head twitching with eye deviation to the right. She could not be

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aroused and fell limply when her mother tried to stand her up. An ambulance was called, but the spell resolved after a few minutes and no further evaluation was sought. Similar episodes of focal twitching, gaze deviation, emesis, and transient vision loss with limp unresponsiveness recurred weekly, with each occurrence lasting up to 1 hour. The patient did not remember the spells after they occurred but was at her baseline between episodes without developmental regression.

Her previous history was unremarkable; she was a term infant and had no relevant family history. In between the episodes, a neurologic examination showed normal results, and no examination during an episode was available.

Questions: What is the differential for abdominal pain, headache, vomiting, and altered mental status, and which conditions should be emergently ruled out? How does this differential change if the symptoms are episodic with interim wellness between episodes?

Discussion

Gastrointestinal diagnoses that should not be missed include bowel obstructions such as intussusception or malrotation and an intraabdominal mass. Less emergent explanations for symptoms include cyclic vomiting syndrome and hepatitis. Neurologic emergencies such as subarachnoid hemorrhage, dural venous sinus thrombosis, and inborn errors of metabolism should be ruled out. Other neurologic causes of such symptoms include epilepsy, classic migraine, and abdominal migraine. Emergent endocrinopathies such as diabetic ketoacidosis and Addison disease, or other low cortisol states, could also present this way. Infectious emergencies such as meningitis or encephalitis should be considered, although more common causes include strep pharyngitis or viral gastroenteritis, with subsequent dehydration causing altered mental status. Acute ingestion of alcohol or iron should also be excluded.

Our patients had self-limited episodes of illness recurring over many months. This intermittence rules out emergencies that progressively worsen without intervention, such as bowel obstruction and intracranial hemorrhage, and disorders that recur less frequently, such as viral gastroenteritis.

Importantly, both patients were healthy without developmental regression between episodes. This makes inborn errors of metabolism such as urea cycle defects, mitochondrial diseases, or organic acidemias extremely unlikely, because these disorders typically show progressive neurologic decline after each episode without a return to the patient's baseline.[1] These children often have some degree of global developmental delay, growth disturbance, and exercise intolerance, and they may have dysmorphic facies.

Migraine can cause headache, abdominal pain, and emesis. Migraine is common in prepubescent children and affects boys and girls almost equally.[2] Distinguishing features include photophobia, phonophobia, scotomas, severe headache, and durations of 1 to several hours. Migraine variants include ocular migraine with episodic blindness, abdominal migraine with severe emesis and minimal headache, and basilar migraine with severe vertigo.[2,3] Children with migraines can appear somnolent but will interact appropriately with symptom control and gentle coaching. This contrasts with our patients, who both had true altered mental status with less severe headache.

Cyclic vomiting syndrome is a preschool-aged variant of migraine. Patients often develop classic migraines by adolescence and have a family history of migraine.[3] These previously healthy children acutely develop multiple attacks of vomiting per day, lasting for several days and often recurring monthly. Laboratory investigations show no acute cause, and the emesis stops abruptly, often without specific treatment. In contrast to our patients, repeated vomiting is the main symptom, and altered mental status is atypical. The syndrome's defining feature is the stereotyped nature of recurrent vomiting attacks without apparent trigger or cause of remission. The diagnosis of migraine or cyclic vomiting is based on careful clinical history. Brain MRI can be obtained to rule out brain lesions if the history is atypical.

It is also important to consider psychogenic nonepileptic seizures (pseudoseizures), conversion disorder, and Munchausen-by-proxy syndrome in the proper psychosocial context, usually as a diagnosis of exclusion.

Finally, this differential includes recurrent seizures, which manifest as brief, discrete episodes of awareness loss with associated symptoms that are stereotypic or reproducible with each episode, followed by full return to baseline. Associated motor, sensory, and visceral symptoms reflect the particular brain region activated by the epileptic discharges and could include headache, emesis, and abdominal pain. While most seizures last from 30 seconds to under 2 minutes, occipital epilepsy of childhood is one exception and can present with seizures lasting several hours.[4]

CASE 1 CONTINUATION

The patient's mental status gradually improved over the next 2 days of admission. His cerebrospinal fluid cell count; differential; and glucose, protein, lactate, and amino acid levels were unremarkable. Metabolic testing of ammonia, pyruvate, serum amino acids, acylcarnitine profile, urine amino acids, and urine organic acids revealed normal results. Microarray analysis yielded unremarkable results. A neurologist was consulted and recommended an EEG, which showed rare bilateral posterior quadrant spikes and wave discharges during sleep.

CASE 2 CONTINUATION

The patient's neurologic workup included an EEG showing right-sided occipital epileptiform activity with propagation to the right central and frontal regions and normal brain MRI. No further infectious or metabolic workups, including lumbar puncture, were done because of the brief nature of the episodes and the intervals of wellness each time the patient was seen in clinic.

Question: Which childhood conditions show occipital EEG activity?

Discussion

Panayiotopoulos syndrome (PS) and Gastaut Syndrome (GS) are 2 childhood occipital epilepsies. Typical EEG findings in PS include occipital spikes in one-third of patients, occipital and other spikes in another one-third of patients, and spikes in other regions or no EEG findings at all in the last one-third of patients.[5,6] EEG abnormalities typically exist both during and between seizures and are accentuated during sleep.[4] In contrast, EEG findings in GS classically show occipital spikes exacerbated by visual stimulation from sources such as lights or television.[5] Occipital spikes can also be seen in brain lesions and in up to 1.2% of normal children without a history of seizures.[4]

CASES 1 AND 2 CONTINUATION

Both patients were diagnosed with occipital epilepsy of childhood. Patient 1 exhibited symptoms typical of PS, including prolonged episodes of altered mental status with headache, emesis, abdominal pain, and autonomic changes of pallor and tachycardia. Patient 2 had symptoms most consistent with GS, including shorter seizure duration, gaze deviation, focal twitching, transient vision loss, and initial resistance to medications. Occipital EEG abnormalities supported both diagnoses.

Question: What are the typical presentation, diagnostic workup, and prognosis of childhood occipital epilepsy?

Discussion

PS and GS are the 2 major subtypes of childhood occipital epilepsy and represent 2 ends of the clinical severity spectrum observed in this condition. PS is a more common and benign variant, found in 6% of children 1 to 15 years old with nonfebrile seizures.[4] The usual age of onset is 4 to 5 years, with equal prevalence in boys and girls.[5] No definite genetic basis for PS development is known at this time, and there is usually no family history of seizures associated with the condition.[6] Typical episodes involve headache, nausea and/or vomiting, eye deviation, and autonomic changes. These symptoms usually occur near the beginning of the seizure and include pallor more commonly than flushing or cyanosis; mydriasis or miosis; cardiac or respiratory rate changes like tachycardia and apnea; or incontinence and hypersalivation.[4–9] Generalized convulsions, stiffening or decreased tone with cyanosis or pallor, and visual hallucinations can also occur.[4–6] Spells last ~2 hours on average.[5]

In contrast, patients with GS are usually older (8–11 years of age) and experience much shorter seizures lasting <1 minute. Seizures typically include eyelid fluttering, focal twitching, and gaze deviation and can be accompanied by simple geometric visual hallucinations or transient blindness. The autonomic disturbances seen in PS are rare in GS and are often limited to syncopal limp unresponsiveness.[8,10]

The diagnosis of childhood occipital epilepsy is based on a suggestive history when other more emergent etiologies have been ruled out. An abnormal EEG with occipital spikes supports the diagnosis but is not required. Brain MRI is recommended because 10 to 20% of patients with autonomic seizures do have underlying brain pathology.[4] Lumbar puncture for infectious and metabolic workup may be done in an acute first-time presentation but may not be required in children with known recurrent, self-limited spells disrupted by interim wellness.

A patient diagnosed with PS has an excellent prognosis: one-third of patients have only a single episode, one-half of patients have 2 to 5 episodes, and most patients experience remission 1 to 2 years after symptom onset, at an average age of ~6 years.[5,6] Notably, 20% of patients develop a different seizure disorder, typically consisting of Rolandic seizures that disappear in the teenage years.[4,5] Given this benign course, prophylactic treatment is not required but should be considered in patients with frequent episodes that result in repeated invasive testing, hospitalization, missed school, and psychosocial stress to the patient's family. In this case, carbamazepine is recommended as the first line of treatment.[4,5] In contrast, GS is less benign because of its higher seizure burden. Only 50 to 80% of patients achieve seizure remission 10 years after diagnosis, and some degree of learning impairment often persists into adulthood as a result.[5,10]

CASE 1 RESOLUTION

Patient 1 was started on oxcarbazepine with improvement in episode frequency from 2 to 3 times per month to no occurrences in the past 8 months at the time of manuscript submission.

CASE 2 RESOLUTION

Patient 2 was trialed on oxcarbazepine, gabapentin, and clobazam without improvement but had better control with topiramate, which reduced spell frequency from weekly to once every 4 to 6 months.

CONCLUSIONS

Occipital epilepsy of childhood is a common yet underrecognized pediatric seizure syndrome. Its 2 major subtypes, PS and GS, have distinct presentations that include headache, altered mental status, emesis, autonomic disturbance in PS, and visual hallucinations in GS. They also differ in treatment response and prognosis. Diagnosis requires a suggestive history and can be supported by occipital discharges on EEGs. The prognosis for children diagnosed with occipital epilepsy is good overall, and the remission of seizures is particularly likely for a PS variant. Timely diagnosis is essential to avoid repeated invasive testing, initiate appropriate treatment if needed, and reassure the patient's family.

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