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Index of Suspicion

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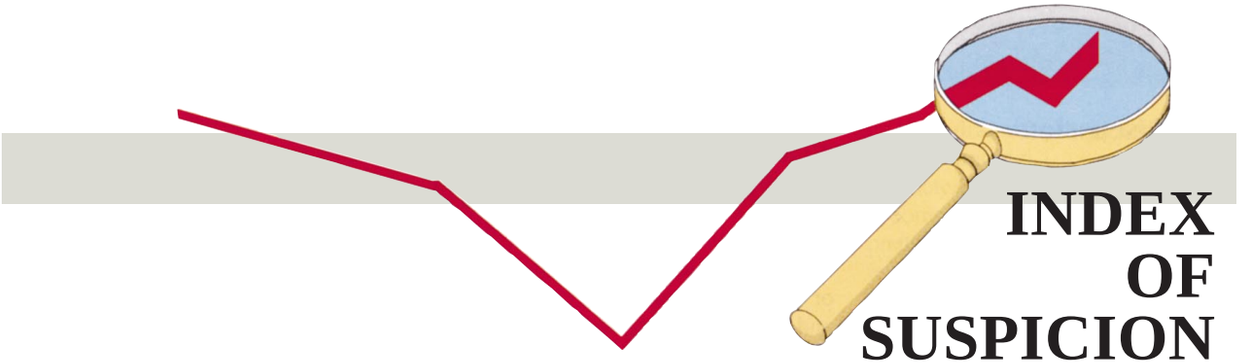

INDEX OF SUSPICION

This section of Pediatrics in Review *reminds clinicians of those conditions that can present in a misleading fashion and require suspicion for early diagnosis. Emphasis has been placed on conditions in which early diagnosis is important and that the general pediatrician might be expected to encounter, at least once in a while. The reader is encouraged to write possible diagnoses for each case before turning to the discussion, which is on the following page.*

We invite readers to contribute case presentations and discussions.

Case 1 Presentation

A 5-year-old girl is seen because of 5 days of fever and worsening cough accompanied by nausea, poor appetite, and decreased fluid intake. She complains of right-sided back and shoulder pain and has difficulty walking. She has had no contact with ill persons. On physical examination, her rectal temperature is 38.9˚C (102˚F), pulse is 151 beats/min, respirations are 52 breaths/min, and oxygen saturation by pulse oximetry is 95% on room air. Her respirations are labored, rapid, and shallow, with nasal flaring, retractions, and occasional grunting. Inspiratory crackles are heard over the right lung base. Scattered palatal petechiae are present. Her abdomen is diffusely tender, without guarding or rebound tenderness.

Her leukocyte count is 7,900/mm^3, with 76% band forms, 5% metamyelocytes, and 2% myelocytes. Chest radiographs demonstrate complete opacification of the right middle lobe with a small right pleural effusion. The child is admitted for intravenous rehydration and broad-spectrum antibiotic therapy.

Despite therapy, she develops moderate respiratory distress, vomiting, and oliguria. Computed tomography of the chest demonstrates a large right pleural effusion with early left mediastinal shift. A right thoracostomy tube is placed and 400 mL of fluid is evacuated.

Despite these measures, the child's condition continues to deteriorate. Her platelet count is 18,000/mm^3; hematocrit, 15%; direct bilirubin level, 22 mg/dL; aspartate aminotransferase level, 200 U/L; prothrombin time, 15 sec; and serum creatinine level, 1.5 mg/dL. Blood, cerebrospinal fluid, urine, and pleural fluid cultures show no growth. Further testing reveals the diagnosis.

Case 2 Presentation

A previously healthy 9-month-old girl comes to the urgent care center with a 5-day history of diarrhea and 1 day of nonbilious vomiting. She vomited eight times this afternoon and gradually has become lethargic. She is admitted for intravenous (IV) rehydration.

On physical examination, the infant is sleepy but arousable. Her blood pressure is 111/53 mm Hg, pulse is 120 beats/min, respiratory rate is 48 breaths/min, and temperature is 38˚C (100.4˚F). She is mildly dehydrated. Results of cardiopulmonary examination are normal. Her abdomen is soft, and no organs or masses are felt; distinct bowel sounds are heard. Initial blood chemistries are: sodium, 146 mmol/L; potassium, 2.4 mmol/L; chloride, 114 mmol/L; bicarbonate, 19 mmol/L; urea nitrogen, 18 mg/dL; creatinine, 0.2 mg/dL; and glucose, 110 mg/dL. Her stool is guaiac-negative.

Twelve hours later, the child's urinary output has decreased despite IV rehydration. She has become very irritable, her abdomen is rigidly distended, and no bowel sounds are heard. A rectal examination is normal. An abdominal radiograph reveals an air-fluid level in the left upper quadrant, with the left diaphragm partially obscured by a round soft-tissue mass superimposed on the heart shadow. A gastrografin enema is normal. An attempt to place a nasogastric (NG) tube during fluoroscopy is unsuccessful. The child is taken to surgery, where the diagnosis is made.

Case 3 Presentation

A 3-year-old boy is brought to the emergency department because of the sudden onset of left-sided weakness. On waking him this morning, his parents noticed that he could not stand or move his left arm or leg. There is no history of trauma nor any suspicion of abuse. The child has had a recent cough and nasal congestion that were treated with a syrup containing pseudoephedrine and dextromethorphan, but he has not appeared ill. He has developed normally and always has been healthy.

Physical examination reveals an afebrile, fearful boy who does not appear toxic and who shows no signs of trauma. Neurologic evaluation reveals decreased strength (grade I-II/IV) in his left arm and leg. Deep tendon reflexes are brisker on the left side. No sensory loss is demonstrable. No signs of increased intracranial pressure or meningeal irritation are present. The remainder of the physical examination is normal.

Results of the following studies are normal: complete blood count; levels of electrolytes, blood urea nitrogen, creatinine, and glucose; liver function tests; prothrombin time; partial thromboplastin time; and bleeding time. A lumbar puncture yields normal cerebrospinal fluid. Results of computed tomography of the head are normal.

The boy is admitted to the hospital. That evening he is much improved and the next day is running about normally. One week later, he is readmitted for a different disorder that explains his previous episode of transient weakness.

Case 1 Discussion

As the child's clinical course evolved, the constellation of jaundice, renal compromise, anemia, and thrombocytopenia raised concern about leptospirosis. Serologic titers for antibodies to *Leptospira interrogans* were obtained and were positive. Further investigation revealed that she lives downhill from a pig slaughterhouse and that water seeps into the yard, especially after rainstorms. In addition, a dead rat had been found near the house prior to the girl's becoming ill.

Leptospirosis is a zoonotic disease caused by the spirochete *L interrogans.* The true incidence of the disease in the United States is unknown, although it has been noted to occur most commonly in the summer months. The highest reported incidence is in the south Atlantic, Gulf, and Pacific coastal states.

Sources for infection include both wild and domestic mammalian species, especially dogs, rats, and livestock, who excrete the organism in their urine. Human transmission occurs through direct contact of mucosal surfaces and cut or abraded skin with the urine or carcass of infected animals. In addition, transmission may occur indirectly from swimming or wading in streams or puddles that have been contaminated by the urine of infected animals. Persons at risk include individuals engaging in outdoor recreational activities such as freshwater swimming, canoeing, kayaking, fishing, and hunting. Furthermore, certain individuals are predisposed by occupation, including farmers, ranchers, trappers, veterinarians, loggers, sewer workers, rice-field workers, and military personnel. Infected people usually excrete the organism in their urine for several weeks, but person-to-person transmission is rare.

The incubation period for leptospirosis usually is 7 to 12 days. Following penetration of skin or mucosal surfaces, the organism invades the bloodstream and spreads throughout the body. The spectrum of clinical illness includes subclinical infection, a self-limited anicteric febrile illness with or without meningitis (85% to 90% of cases), and a severe, potentially fatal illness known as Weil syndrome, which is characterized by a hemorrhagic diathesis, renal failure, and jaundice (5% to 10% of cases).

The onset of the self-limited form of leptospirosis usually is abrupt and involves nonspecific flu-like symptoms, including high fever, chills, headache, severe myalgia, and malaise. Gastrointestinal symptoms are common. Conjunctival suffusion is a characteristic physical finding, but is present in fewer than 50% of the cases. Physical examination also may reveal the presence of a maculopapular rash, pharyngeal injection, hepatosplenomegaly, lymphadenopathy, and myositis. These signs and symptoms are prominent for 4 to 7 days during this septicemic stage. The organism may be isolated from both blood and cerebrospinal fluid (CSF) during this stage.

After the initial septicemic stage, the patient defervesces and may show signs of clinical improvement. Following an asymptomatic period of 1 to 3 days, the patient enters the immune stage of illness (notable for the appearance of immunoglobulin M [IgM] antibodies). Aseptic meningitis, characterized by fever, headache, and vomiting, is the hallmark of this stage. The CSF cell count is greater than 500 cells/mm^3 in most cases, with a predominance of polymorphonuclear cells early in the illness, followed later by a predominance of mononuclear cells. Blood and CSF cultures usually are negative during this stage, which lasts from 4 to 30 days. Leptospiruria develops and typically persists for 1 to 6 weeks.

In approximately 5% to 10% of cases, as in this child, leptospirosis follows a more severe clinical course that may be characterized by hepatic involvement (jaundice, abnormal liver function tests, hepatomegaly, and liver failure) and renal dysfunction (proteinuria, pyuria, azotemia, oliguria, and anuria). This form, called Weil syndrome, has a mortality of 5% to 10% and also involves anemia, thrombocytopenia, and leukocytosis with neutrophilia. Initially, the patient presents with nonspecific, flu-like symptoms, but between the third and seventh day of illness, the clinical course becomes more severe, with the development of jaundice, azotemia, and oliguria. In severe cases, a hemorrhagic diathesis and shock may ensue. Chest radiographs usually are abnormal and may show evidence of large confluent areas of consolidation, as in this case. More commonly, however, there is evidence of bilateral parenchymal disease.

The diagnosis of leptospirosis frequently is missed and should be considered in the differential diagnosis of patients who have severe headache, myalgia, and fever and whose occupational, recreational, or household activities suggest risk factors for exposure. Leptospirosis may account for as many as 10% of cases of aseptic meningitis and, therefore, should be considered in these cases as well. Blood and CSF should be cultured on special media in the first 7 to 10 days of illness, as should urine after the first week and during convalescence.

Serologic tests for the detection of leptospiral antibodies are available, and the diagnosis can be made by documenting a fourfold or greater rise in titers of antibody in acute- and convalescent-phase sera taken from a patient who has a clinical presentation compatible with leptospirosis.

Supportive care and close monitoring of these patients is essential. The hepatic dysfunction in Weil syndrome usually resolves and rarely is the cause of death. Renal failure may require temporary hemodialysis, but it also usually resolves spontaneously. Penicillin is the drug of choice for patients who have severe illness (1.5 million units administered intravenously every 6 hours for 7 days). In patients who have mild illness, oral doxycycline or amoxicillin may be effective in shortening the course of the illness. The child in this case responded to a 7-day course of high-dose intravenous penicillin. Her renal and hepatic dysfunction resolved with supportive care, and she had no further complications.

Vaccination of dogs and livestock prevents disease, but not infection. Furthermore, vaccination programs have not been shown to prevent transmission to humans. For this reason, protective clothing, boots, and gloves should be worn to reduce

occupational exposure. In adults, doxycycline 200 mg orally once a week is an effective prophylaxis and should be considered for high-risk groups who have short-term exposure. However, tetracycline drugs should not be administered to children younger than 8 years of age. (*Stephanie R. Starr, MD, Derek S. Wheeler, MD, United States Naval Hospital, Guam*)

Case 2 Discussion

During laparotomy, a gastric volvulus was found and derotated. Cloudy fluid was present in the peritoneal cavity. The stomach was massively distended and infarcted irreversibly. A subtotal gastrectomy was performed, sparing a section of the gastric cardia and antrum. A partially infarcted spleen, which was attached to an eventrated left hemidiaphragm, was separated from the eventration. A splenectomy was believed to be unnecessary. The anterior gastric wall was anchored to the peritoneum. A gastrostomy tube (G-tube) was placed. The patient was admitted to the pediatric intensive care unit for continued intubation and further management.

Gastric volvulus is a rare disorder in childhood. It is caused by an abnormal rotation of one part of the stomach around another along its coronal or sagittal axis. Normally, the esophageal hiatus, the pylorus, and four gastric ligaments hold the stomach in position. The ligamentous attachments are gastrophrenic, gastrohepatic, gastrosplenic, and gastrocolic. The stomach loses its anchoring when the ligaments are absent or become attenuated, which allows abnormal mobility of the stomach. Gastric volvulus usually is classified as organoaxial or mesentricoaxial. Rarely, combinations of these two anatomies may be encountered. Organoaxial gastric volvulus is the most common form and accounts for two thirds of the cases. In these patients, absence or laxity of the gastrohepatic and gastrosplenic ligaments allows the stomach to rotate along its longitudinal axis. In patients who have mesentricoaxial volvulus, laxity of the gastrophrenic ligament and duodenal attachments allows rotation around the stomach's transverse axis, with displacement of the pyloric region anteriorly, in front of the esophagus.

Other factors that can contribute to the development of gastric volvulus include diaphragmatic hernia or eventration, esophageal hiatal hernia, and pronounced distention of the stomach or adjacent intestines. In this case, an attenuated gastrosplenic ligament, a left hemidiaphragmatic eventration, and a massively distended stomach predisposed this child to gastric volvulus.

Prompt recognition of gastric volvulus is required to prevent infarction. In adults, the Borchardt triad of unproductive retching, acute localized epigastric distention, and inability to pass a NG tube is pathognomonic for gastric volvulus. However, these clinical features are not present in all children who have gastric volvulus. Children usually present with a subset of the following symptoms: colicky abdominal pain, upper abdominal distention, vomiting, regurgitation, hematemesis, and failure to thrive. This child presented with vomiting and later developed abdominal distention. The inability to place a NG tube, especially during fluoroscopy, should alert the physician to the possibility of a gastric volvulus.

Abdominal radiography is essential for diagnosing children who have gastric volvulus because the clinical presentation is nonspecific. On an erect abdominal plain radiograph, a mesentricoaxial volvulus demonstrates a large gastric shadow with double air-fluid levels in the fundus and antrum. A barium study shows an inverted stomach and possibly an obstruction. An organoaxial volvulus demonstrates a large horizontal stomach with a single air-fluid level. The gastroesophageal junction lies lower than the normal position, and the antrum and duodenum might be distorted. In addition to the volvulus, signs of diaphragmatic hernia or eventration also might be noted on an abdominal radiograph. This child's radiographic findings were suggestive of an organoaxial gastric volvulus with a left diaphragmatic eventration.

Treatment of gastric volvulus is surgical. After derotation, the stomach is decompressed with NG suction or gastrostomy. To prevent recurrences, the stomach is fixed in position with anterior gastropexy and, if necessary, gastrostomy. Any associated intra-abdominal abdominal abnormalities are corrected at this time.

This child did well postoperatively. She was given total parenteral nutrition and broad-spectrum antibiotics for 8 days. By her ninth postoperative day, she could tolerate G-tube and oral feedings. She was sent home 12 days after surgery. Her diet has been supplemented with vitamin B12 and iron.

Although gastric volvulus is rare, this case reminds the clinician of the wide spectrum of disorders that can obstruct the intestinal tract and of the necessity to keep obstruction in mind when children manifest repeated vomiting or persistent abdominal pain. Nonbilious vomiting, as experienced by this patient, suggests obstruction of the gastrointestinal tract proximal to the ampulla of Vater. Plain abdominal radiographs may provide significant clues about the specific diagnosis, but often contrast radiography is required for definitive treatment. (*Mary E. Rimsza, MD, John Pohl, MD, Cindy Duke, MD, Maricopa Medical Center, University of Arizona College of Medicine, Phoenix, AZ*)

Case 3 Discussion

The boy was readmitted to the hospital after developing complex partial seizures with secondary generalization that involved the left side of his body. Interictal electroencephalography revealed multifocal spikes in the right temporal lobe. A detailed family history obtained at this stage revealed a positive family history of seizures on both sides of the family, although neither parent had experienced seizures.

In retrospect, the weakness responsible for his previous admission was designated as Todd paralysis following an unobserved seizure that had occurred during sleep. The parents had observed only the residual paralysis, which characteristically resolved completely in 24 hours. Todd paralysis typically occurs in a patient who has nonprogressive neurologic disease that involves

focal seizures. The seizures are followed by a brief postictal paralysis. Recovery is complete, with no neurologic abnormalities persisting beyond 24 hours.

The cause of Todd paralysis is not completely understood. Initially it was believed that the paralysis was due to "exhaustion" of the involved neurons, but now the paralysis is attributed to an inhibitory phenomenon, possibly related to neurotransmitter dysfunction. The duration and severity of the seizure do not correlate with the degree of postictal paralysis. On the contrary, postictal paralysis often is more pronounced after an arrested partial seizure than after a complete, more severe seizure. The paralysis usually, but not always, is noted in the area of the focal seizure activity. A patient may experience weakness in an extremity other than the one involved in the seizure. Aphasia also may occur.

On physical examination, patients who experience Todd paralysis frequently will respond to plantar reflex testing during the time of weakness with extension of the first toe. After recovery, the reflex reverts to the normal plantar flexion response.

It is helpful for clinicians to use a grading system to describe variations in the strength of different limbs or muscle groups. The grades range from 0 to IV. 0 rating indicates that no active movement is possible; I indicates movement, but not against gravity; II denotes movement against gravity but not against resistance; III indicates movement against partial resistance; and IV designates movement against full resistance.

The term hemiplegia traditionally has been used to describe an upper motor neuron type of weakness that involves both limbs on the same side of the body. The etiology of acute hemiplegia in a child is diverse; no cause can be established in a significant percentage of cases.

Several primary neurologic conditions can mimic Todd paralysis. Hemiparetic seizures, which are characterized by acute lateralized weakness and a normal mental state, are a type of partial epilepsy, but the paresis in this disorder is ictal rather than postictal. Alternating hemiplegia of childhood may mimic Todd paralysis by causing repeated episodes of hemiplegia that last a few minutes to a few days. A similar transient neurologic deficit may be seen after a migraine attack, but the presentation usually involves the typical headache and a positive family history of migraine. Transient ischemic attacks (TIAs) also may present with a focal, nonconvulsive neurologic deficit due to interruption of the cerebral perfusion. The paralysis may occur abruptly, and the patient may recover completely within 24 hours. However, recurrent TIAs can leave slight residual paresis.

Embolic and thrombotic events caused by disorders of different organ systems can lead to hemiplegia. Embolization of blood vessels in the brain may occur in association with cardiac arrhythmias, bacterial endocarditis, or cardiac catheterization. Other cardiac disorders, such as cyanotic heart disease with polycythemia and hypoxia, may predispose to thrombus formation. Hematologic disorders such as sickle cell anemia, hemolytic-uremic syndrome, and polycythemia also can cause thrombus formation. Lesions in the carotid artery, such as from accidental trauma to the neck or tonsillar area, may lead to thrombus formation. Intracranial hemorrhage is a rare cause of hemiplegia and usually is seen in patients who have leukemia, bleeding disorders, and vascular malformations. The mode of onset of the hemiplegia is helpful in making a diagnosis. Embolic phenomena have the quickest onset, occurring in minutes, whereas thrombus formation usually occurs over several hours.

Experiences with patients such as this underscore the value of a detailed history and thorough examination, emphasize the need to consider all systems when evaluating a child who has a puzzling condition, and remind the clinician that two quite different clinical problems can have a common pathogenesis. (*Harminder S. Dhaliwal, MD, Columbia Medical Center, Lewisville, TX*)

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