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Case 1 Presentation

A 4-month-old girl is brought to the ED because of fever and loss of appetite. Yesterday she was less playful than usual and whined instead of crying. She took only 3 oz of formula, after which she vomited twice. She refused other feedings and urinated only once during the day. Four days ago, she had rhinorrhea, but had no cough, diarrhea, or sick contacts. She was lethargic this morning and had a temperature of 99.5°F (37.5°C), prompting the visit.

On physical examination, the baby appears lethargic, but is afebrile and has stable vital signs. Her fontanelle is full, and she has bruises on the helix of her left ear and a left hemotympanum. In the ED, she has a tonic seizure with leftward gaze and absence of right-sided movement. Lorazepam is administered, she is intubated, and she receives a loading dose of fosphenytoin.

Laboratory findings include: WBC count of 26.08×10^9/mcL (26.08×10^9/L) with 80% neutrophils, 5% monocytes, and 14% lymphocytes; hemoglobin of 9.5 g/dL (95 g/L); hematocrit of 29.7% (0.297); and platelet count of 387×10^9/mcL (387×10^9/L). Prothrombin time is 15.3 seconds, and partial thromboplastin time is 26.1 seconds. Basic metabolic panel results are normal, as are findings on urinalysis. Rapid tests for respiratory syncytial virus and influenza as well as blood and urine cultures are negative. An additional procedure reveals the reason for her condition.

Case 2 Presentation

A 14-year-old girl is seen for a 5-day history of vertigo, nausea, vomiting, headache, and ataxia. The onset of her illness was sudden and dramatic, with her symptoms awakening her from sleep. Paramedics felt that she had a flulike syndrome and did not take her to the hospital. Later that day, she experienced clumsiness in walking and right facial weakness. On day 2 of the illness, she had worsening vertigo and was unable to stand; on day 3, an ED evaluation yielded a diagnosis of urinary tract infection, and she was treated with intravenous fluids and oral antibiotics. She denies any recent fever, illness, or trauma and uses no medications or illicit drugs.

On physical examination, the girl is afebrile and has stable vital signs. She is alert and oriented, with intact cognition and language, but has mild dysarthria. She has severe right-sided facial weakness, including the forehead (lower motor neuron pattern), and a mild decrease in sensation over the right cheek. She demonstrates severe incoordination of the right arm and leg, with difficulty walking, but has full motor power. Her deep tendon reflexes are symmetric, with downgoing plantar responses.

One diagnostic procedure reveals the source of her bothersome symptoms and signs.

Case 3 Presentation

A 4-year-old boy from Honduras, who is severely developmentally delayed, is admitted for an elective Blalock-Taussig shunt for his tetralogy of Fallot. He has had a recent dental procedure. On admission, he has mild rhinorrhea and is febrile, but has no other symptoms.

On physical examination, the child is irritable, but consolable. He appears cachectic and has macrocephaly, with a large, open, flat anterior fontanelle. He has a fever, tachycardia, and hypoxia. He exhibits cyanosis of his lips and nail beds and has marked clubbing of all nails. He has pectus carinatum and a grade II/VI systolic ejection murmur.
heard best at the left upper sternal border that radiates. His abdomen is protuberant. He moves all extremities and is able to grasp objects. There is bilateral ankle clonus. He sits without support, but cannot stand independently and is unable to walk. He speaks fewer than five words, and his developmental age is estimated to be approximately 9 months.

A metabolic panel and urinalysis yield normal results. His CBC shows normal leukocyte counts and polycythemia. All blood cultures are negative. He continues to have fevers despite being given broad-spectrum antibiotics. A tuberculin test is negative. Chest radiography is normal. Echocardiography shows no cardiac valvular vegetations. An imaging procedure is performed.

**Case 1 Discussion**

CT of the head revealed a small left frontotemporal subdural hematoma with blood in the falk; cerebral edema on the left with midline shift; subfalcine herniation; and infarction of the left cerebrum, right frontal lobe, and right cerebellum. MRI demonstrated edema of the entire left cerebrum, inferior right frontal lobe, area posterior to the right lateral ventricle, and right cerebellar hemisphere; a small left posterior fossa subdural hematoma; a subdural hematoma along the entire left cerebrum; multiple possible areas of subarachnoid hemorrhages of the middle cranial fossa; and a midline shift with uncal herniation.

The patient lives with her mother, grandmother, and three older siblings ages 2, 4, and 5 years old. All children were with the father over the weekend when the patient became ill. Both parents denied prior Child Protective Service (CPS) involvement, domestic violence, and drug or alcohol use.

Nevertheless, inflicted head trauma was suspected based on the medical findings and the lack of trauma history. A report was made to the CPS agency in the jurisdiction of the child’s father, and the hospital’s child abuse specialist was consulted. As part of the CPS and police investigation, the patient’s siblings were taken to the local child advocacy center for a forensic interview, during which the oldest brother reported that his 4-year-old brother was pushing the baby vigorously in a swing, which he demonstrated. The baby flew out of the swing, landing on the side of her face, and her father picked her up. She then slept all day. Her injuries were consistent with the mechanism described by her brother. CPS has substantiated neglect on the father’s part for inadequate supervision, delayed seeking of medical help, and failure to disclose the injury. CPS continues to monitor the family.

**The Condition**

The triad of unilateral ear bruising, retinal hemorrhages, and ipsilateral cerebral edema is consistent with a form of inflicted head trauma termed “tin ear syndrome.” The incidence of tin ear syndrome is unknown; it has been reported in the literature only as individual case reports. Similar to shaken baby syndrome, the mechanism of injury involves rotational acceleration of the head. In the case of tin ear syndrome, however, the head is set in motion by a direct impact to one side of a child’s head.

Because the infant brain is not yet myelinated, rotational acceleration results in diffuse axonal injury and tearing of bridging cortical veins, which causes subdural hematomas (SDHs). The immature neck muscles and relatively large head permit the brain to move within the skull when forces of rotational acceleration and deceleration are applied. SDH, although more common in shaken baby syndrome, also can occur in tin ear syndrome. The distribution of SDH from shaking typically is bilateral and along the falk; SDH from direct contact force to the head typically is ipsilateral in children.

In this patient, although physical abuse was suspected, the subsequent disclosure from her brother provided a plausible accidental mechanism for
injuries more commonly seen with abuse. The rotational acceleration of her head occurred when she was catapulted from the moving swing and landed on the left side of her head.

Evaluation
Any child who experiences acute altered mental status along with other signs of increased intracranial pressure should undergo head CT, which can demonstrate intracranial hemorrhage, cerebral edema, and soft-tissue swelling of the scalp. Subdural collections of different densities visible on CT raise concerns about hemorrhages of different ages and the possibility of multiple episodes of trauma. MRI is likely to be helpful at this point. However, it is not possible to date intracranial hemorrhages definitively by neuroimaging; patient signs and symptoms are the best indicators for dating inflicted injuries. If the distribution of the hemorrhages raises suspicion for vascular injury or malformations, MRA can be useful in assessing the cerebrovascular anatomy.

Because all retinal hemorrhages do not result from obvious inflicted head trauma, a dilated funduscopic examination should be performed on children who have acute changes in mental status as soon as possible to evaluate for and describe retinal hemorrhages. A skeletal survey should be completed for all children younger than 2 years of age for whom abuse is suspected to look for occult skeletal injuries. A “babygram,” which is a single anteroposterior view of the chest and abdomen, is insufficient because the entire skeleton is not included.

Treatment
Severe traumatic brain injury requires resuscitation that includes intubation and ventilation when the Glasgow Coma Scale score is less than 8 or if there is cardiopulmonary compromise. Intravenous access to maintain normal blood pressure is required. Patients may need blood products as well as dopamine or epinephrine to maintain blood pressure. Anticonvulsants should be administered for seizure activity. The greatest risk for mortality is transtentorial herniation. Thus, patients must be monitored closely for any signs of increased intracranial pressure. If signs of herniation are present, acute hyperventilation until the signs resolve, sedation, muscle relaxation, and administration of an osmotic agent are required.

Lessons for the Clinician
It is important for clinicians to remain nonjudgmental in situations that raise the suspicion of abuse, focusing on the mechanism required to cause the injury. Reasonable suspicion, not proof, of abuse and neglect is what “mandated reporters,” including physicians, must report to CPS agencies. As part of the police/CPS investigation, forensic interviews of possible witnesses, including other children, can be invaluable. (Allison Jackson, MD, MPH, Zarir Khademian, MD, Rachel Y. Moon, MD, Children’s National Medical Center, Washington, DC)

Case 2 Discussion
Diffusion MRI of the brain demonstrated lesions in the right cerebellum, right pons, left thalamus, and left corpus callosum (Fig. 2A, B, C), all structures supplied by the posterior (vertebrobasilar) circulation. MRA suggested absent flow in the right vertebral artery (Fig. 2D). Cer- ebral angiography demonstrated irregular narrowing of the right vertebral artery (Fig. 2E) and an intraluminal thrombus (Fig. 2F).

The clinical and MRI findings confirmed the diagnosis of multiple acute arterial ischemic strokes (AIS) occurring at different times over the preceding week. Angiography confirmed vertebral artery dissection, with artery-to-artery emboli from the intraluminal thrombus traveling downstream to cerebellar, brainstem, and thalamic arteries.

The girl was given anticoagulation treatment with unfractionated heparin as well as neuroprotective strategies that included maintenance of normal blood pressure, blood glucose concentrations, and temperature. Results of additional investigations, including echocardiography and prothrombotic testing, were negative. She showed modest clinical improvement over 2 weeks and was discharged to the rehabilitation facility on low-molecular weight heparin (LMWH) for secondary stroke prevention.

The Condition
Stroke has emerged as a relatively common but underrecognized cause of neurologic disability in children. An incidence of 3 to 8 per 100,000 children per year suggests that ischemic strokes are more common than pediatric brain tumors. Most ischemic strokes in children are arterial, but 20% occur as a result of cerebral sinovenous thrombosis. The focus of this discussion is on ischemic stroke and not on episodes caused by intracranial bleeding.

The sudden onset of focal neurologic deficits in a child should be considered to represent a stroke until proven otherwise. Children who suffer strokes usually present with abrupt onset of hemiparesis, but other focal deficits, including hemisensory changes, visual loss or diplopia, loss or slurring of speech, and imbalance or incoordination, should raise suspicion. Confounding signs and symptoms such as headaches,
seizures, and altered consciousness are common.

The differential diagnosis of an acute neurologic syndrome is lengthy (Table). The initial vertigo and ataxia in this patient raised additional considerations of acute cerebellitis, posterior fossa tumors, and vestibular diseases.

A risk factor is identified in more than 70% of children afflicted with stroke, and many harbor multiple risks. Three large categories of associations are cardiac disease, arteriopathies, and prothrombotic disorders. Complex congenital heart lesions are associated commonly, with interventional or surgical procedures also increasing the risk. Arteriopathies include arterial dissection, Moyamoya disease, and sickle cell disease as well as inflammatory conditions related to infection (meningitis, varicella) or vasculitic conditions. Prothrombotic disorders include the factor V Leiden mutation, elevated lipoprotein (a) concentrations, and deficiencies of protein C or S. A wide variety of acute and chronic systemic illnesses may increase the risk of pediatric stroke.

Stroke is diagnosed by clinical and neuroimaging findings. CT can demonstrate AIS and rule out hemorrhage, but is insensitive in the acute phase. MRI is the investigation of choice; diffusion-weighted MRI has revolutionized the early diagnosis of cerebral ischemia. Angiography can be accomplished with MR or CT techniques, although conventional angiography may be required for diagnoses such as dissection. Additional investigations such as echocardiography and prothrombotic testing are used to eliminate other risk factors.

The Underlying Condition
Arterial dissection accounts for 7% to 20% of pediatric AIS and may occur in the anterior (carotid) or posterior (vertebrobasilar) circulations. Dissection occurs when blood extrudes into the medial layer of the arterial wall through a tear in the endothelial surface. This abnormal surface allows pathologic thrombus formation, with subsequent localized arterial occlusion or embolization to downstream arteries. The pathophysiology of the initial vessel injury is not completely understood. Tethering of the vertebral arteries between the C1 and C2 vertebrae is likely to be relevant because neck rotation stretches the local segment of the artery. A history of recent trauma often is obtained, but the trauma usually is trivial and within the normal experiences of childhood. A history of sports injuries or chiropractic neck manipulation should be sought. Adult evidence suggests that most patients experiencing dissection harbor ultrastructural connective tissue abnormalities, although few have recognizable clinical syndromes such as Ehlers-Danlos or Marfan. Specific clinical clues to dissection include the presence of Horner syndrome (ca-
rotid dissection) or neck pain (vertebral dissection).

Management
A child suspected of having had a stroke should be seen urgently by a pediatric neurologist. Evidence-based management is lacking, but recent consensus-based publications provide useful guidelines. The immediate thrombolytic (“clot-busting”) treatments proven in adult stroke remain unproven in children, but studies are underway. Acute anticoagulation therapy with heparin or LMWH appears safe and may decrease the early progression of stroke and multiple strokes observed in this patient. Published guidelines recommend either initial aspirin (ASA) therapy or acute anticoagulation until investigations have delineated the cause for the stroke, followed by 3 to 6 months of anticoagulation for arterial dissection or a presumed cardiac cause. Early management also should provide supportive care to minimize secondary brain injury and should include maintenance of normal blood pressure, blood glucose concentration, and temperature, along with aggressive treatment of infection and immediate treatment of seizures.

Patients whose strokes are due to other or unknown causes may be maintained on long-term ASA (3 to 5 mg/kg per day), which also is recommended for patients suffering dissection or cardiogenic stroke, following 3 to 6 months of anticoagulation. Such secondary stroke prevention is important because the risk of ongoing recurrence of AIS, particularly in the first 6 months, is 10% to 25%. Early and aggressive physical, occupational, and speech therapy is essential.

Only about 33% of children will be neurologically normal after a stroke, and the mortality rate is 5% to 10%. Most survivors live with moderate-to-severe disability. Motor deficits are most common, but other sequelae include language disorders, cognitive and behavioral problems, movement disorders, headaches, and epilepsy. The burden of illness is exacerbated because the morbidity of stroke in a child affects the entire family and lasts a lifetime.

Lessons for the Clinician
Acute onset of a focal neurologic deficit in a child is an emergency and should be considered a stroke until proven otherwise. A high degree of clinical suspicion is required to improve recognition of pediatric stroke and avoid delays in diagnosis. Because time-dependent treatments now are available and evolving, the importance of improving awareness and diagnostic sensitivity cannot be overemphasized. (Elizabeth Berger, MD, Adam Kirton, MD, Gabrielle deVeber, MD, Hospital for Sick Children, Toronto, Ontario, Canada)

Case 3 Discussion
Head CT performed to assess the boy’s developmental delay revealed four large cystic lesions with right uncal herniation and a mild midline shift to the left, subfalcine herniation, and a persistent anterior fontanelle (Fig. 3).

The differential diagnosis of cystic brain lesions includes neurocysticercosis, paragonimiasis, cerebral hydatid disease (echinococcosis), cerebral abscesses, and arachnoid cysts. Because of the patient’s country of origin, there was a high degree of suspicion for parasitic disease such as neurocysticercosis, paragonimiasis, and echinococcosis.

Neurocysticercosis, endemic to Central and South America, is caused by the larval form of the pork tapeworm, *Taenia solium*. The embryos of this parasite are carried by the bloodstream to the CNS, where they form cysts known as cysticerci. An acute inflammatory reaction occurs on the death of the larva, often complicated by basal meningitis and hydrocephalus. The characteristic CT findings are spherical calcifications 1 to 2 mm in diameter (the scolices), surrounded by a calcified rim 7 to 12 mm in diameter. Although stages are multiple in the evolution of the infection, each with its own imaging findings, the very large noncalcified lesions in this patient, which lacked a central scolex, did not resemble any of these stages.

Paragonimiasis, infection by the lung fluke *Paragonimus westermani,* is another condition endemic to Latin America. Brain involvement is reported in 2% to 27% of affected patients. The infection causes extensive cerebral inflammation and multiple interconnecting granulomas, often appearing as conglomerated, calcified rings that are unlike the discrete, well-separated, noncalcified lesions in this patient.

Cerebral abscess is an important consideration because such lesions often are multiple and are likely to occur in patients who have cyanotic heart disease (resulting from paradoxical emboli). Although variable in appearance on imaging, cerebral abscesses are described classically as having thin, regular-enhancing walls, with or without nodularity on the inner border. Arising most commonly at the corticomedullary junction, they incite abundant vasogenic edema because of the inflammation.

Arachnoid cyst, a developmental variant, occurs peripherally within the layers of the arachnoid membrane. Arachnoid cyst was excluded in this patient by the intracerebral location of the lesions and by their multiplicity, peripheral enhancement, and spherical shape.

Cerebral hydatid disease, an infec-
tion by the tapeworm *Echinococcus granulosus*, also is endemic to Latin America and typically involves the liver and lung and sometimes bone and the genitourinary system. In children, however, this infection may involve the brain. Approximately 50% to 75% of cerebral cases occur in childhood. On rare occasions, the cyst may become infected by bacteria and present as fever. CNS hydatid cysts occur most commonly in the middle cerebral territory of the cerebral hemispheres, as in this patient. The cysts may be single or multiple, usually fewer and larger than those of cysticercosis. They contain fluid similar to water in imaging characteristics. Contrast enhancement and edema of the surrounding brain tissue are uncommon. Wall calcification and enhancing nodules are rare features.

Clinicians should be aware that *E. granulosus* also is found in many parts of the world other than Latin America, including pockets of endemic infection in Arizona, California, New Mexico, and Utah. In addition to these areas, indigenous disease has occurred in Minnesota and among Native Americans in Western Alaska. Recently, concern has been expressed in many parts of Europe because the organism has been detected in many countries in Western Europe. China also has endemic disease. Clinicians must be aware of their patients’ travel histories. The intermediate hosts most relevant to human infection are dogs, sheep, and cattle.

The diagnostic clue in this case was the size of the cystic brain lesions. The four ring-enhancing lesions had thin, uniform walls and surrounding vasogenic edema. The largest lesion was in the right front parietotemporal lobe and measured 7.6 × 6.2 × 8.5 cm. A multilobulated lesion in the left parietal lobe measured 10 × 5 × 4 cm, a smaller lesion was evident in the left temporal parietal lobe, and the smallest was adjacent to the right lateral ventricle. Because they were large, ring-enhancing lesions, there was a low suspicion for both arachnoid cyst and neurocysticercosis. Serology for testing for echinococcosis, cysticercosis, and *Paragonimus* infection was sent to the Centers for Disease Control and Prevention.

**Making the Diagnosis**

The most likely diagnosis, based on radiologic findings and history, was hydatid disease caused by *E. granulosus*. CT of the chest, abdomen, and pelvis revealed no additional visceral cysts. Tests for malaria and dengue fever and blood cultures were negative. The fever eventually resolved following administration of broad-spectrum antibiotics and was presumed to be due to bacterial super-infection of the cysts.

The immunoblot assays for *Echinococcus*, cysticercosis, and *Paragonimus* were negative, but the enzyme immune assay was positive for *Echinococcus*. Based on these results, echinococcal cerebral hydatid disease was diagnosed.

Echinococcal infection is diagnosed via a combination of physical findings and imaging results, with confirmation sometimes by serologic studies, in a patient who has been in an area where the parasite is found.

Figure 3. CT scan showing hydatid (echinococcal) cysts.
**Treatment**

Dexamethasone was started to assist with reduction of the surrounding cerebral edema, which is uncommon in hydatid disease. Shortly thereafter, albendazole was administered to treat the presumed *E gransulosis* infection, which is the treatment of choice for this disease. The child was unable to undergo surgical drainage and resection of these cystic lesions because of his cardiac lesion.

Benzimidazoles (mebendazole and albendazole) are used commonly to treat echinococcosis. Benzimidazoles inhibit the assembly of tubulin into microtubules, thus impairing glucose absorption through the wall of the hydatid parasite, leading to cell death. Medical therapy such as albendazole is indicated for patients who have inoperable, widespread, or numerous cysts of *E gransulosis* and for patients who have complicated medical problems and are unsuitable candidates for surgery, such as this patient. The efficacy of medical therapy seems to correlate more with the duration of therapy than with the serum or cyst concentrations achieved.

Surgery remains the definitive treatment for this disease, but it is associated with the risks of operative morbidity, recurrence of cysts, and spillage of fluid from the cysts, which can lead to anaphylaxis or dissemination of the infection.

**Lessons for the Clinician**

Echinococcosis should be considered in the differential diagnosis of space-occupying cystic lesions. With increasing numbers of immigrations, international adoptions, and airplane travel, clinicians should be alert to the risks of parasitic diseases in children. (Latha Chandran, Adelaide W. During, Dvorah Balsam, State University of New York Medical Center at Stony Brook, Stony Brook, NY)

To view Suggested Reading lists for these cases, visit pedsinreview.org and click on Index of Suspicion.

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**Clarification**

In the article on Heart Murmurs in the April 2007 online issue of *Pediatrics in Review*, the caption for audio file #2 (atrial septal defect) should read: This is a systolic ejection murmur and widely split S2 of a secundum atrial septal defect (ASD). The murmur follows S1 and is crescendo-decrescendo followed by a split S2.
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