

Acute Ataxia in an Infant

Leonard E. Swischuk, MD

Key Words: infant, acute ataxia, ADEM

Pediatric Radiology, Radiology Department, University of Texas Medical Branch, Galveston, TX.

Address correspondence and reprint requests to Leonard E. Swischuk, MD, Pediatric Radiology, University of Texas Medical Branch, Galveston, TX 77550-0365. E-mail: lswischu@utmb.edu.

Copyright © 2005 by Lippincott Williams & Wilkins

ISSN: 0749-5161/05/2106-0403

CASE

This 18-month-old boy, previously healthy, except for recurrent bouts of otitis media, suddenly, one afternoon, started to stagger and weave while walking. Very quickly, this became worse and he then developed a left facial droop with some drooling. In addition, the mother noticed that he could not focus on her with his eyes and then she became quite concerned. All of this happened within a few hours.

She took the infant to her local emergency room where the patient was evaluated. At that institution, a CT study of the brain without contrast was interpreted as being normal. The white blood cell count was 12,400 with 43% neutrophils, 45% lymphocytes, 1% monocytes, and 1% eosinophils. A urinalysis was negative and an attempted spinal tap revealed a bloody tap. Thereafter, the patient was referred to our emergency room (ER). In our ER, the previously

noted neurologic deficits were substantiated and no new ones were detected. The patient did indeed stumble while walking, and for certain, had a left facial droop. At the same time, the patient was relatively coherent and would respond to commands, questions, and so forth. Repeat laboratory investigation substantiated the results from the incoming hospital.

Prior to this episode, and as previously noted, the patient was healthy except for repeated bouts of otitis media. However, the patient fell at dayschool the day before and hit his right cheek. The patient also had another fall at home on a carpeted floor. None of these episodes were reflective of major trauma, and thus could be dismissed as significant factors.

On arrival to our ER, the patient's heart rate was 135 per minute and regular, the blood pressure 110/62, the temperature 36.5°C, and the respiratory rate was 38 and regular. Physical examination was normal except for those findings referable to the central nervous system. These findings were exactly the same as those elicited in the patient's home emergency room.

A repeat spinal tap revealed a glucose level of 65 mg%, a total protein level of 61 mg%, a white blood cell count of 165 with a differential of 8% segmented neutrophils, 29% lymphocytes, 6 atypical lymphocytes, and 56% monocytes. Various clinical consultations were obtained, blood was sent off for evaluation for any abnormal aminoacidurias or other metabolic abnormalities, and an MR study of the brain was obtained (Fig. 1). What do you think the diagnosis might be in this patient?

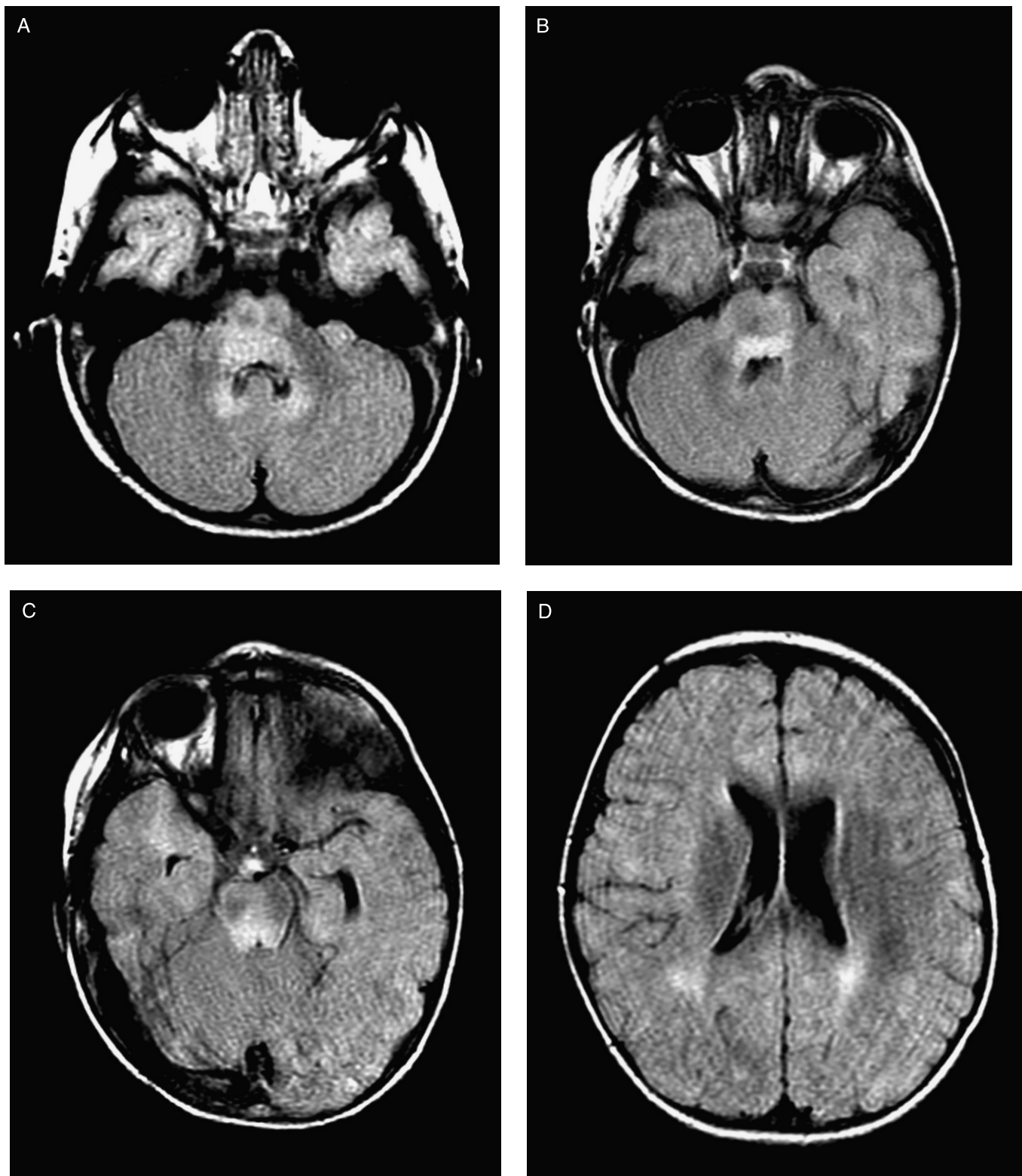


FIGURE 1. A–D, Four axial, MR FLAIR images of the brain.

DISCUSSION

Clearly, in our patient, some neurologic problem was present and had rapid onset. With such rapid onset one probably first should think of infection. With the patient having episodes of recurrent otitis media, one might consider bacterial-induced meningitis. However, this was not the case as the white blood cell count along with the cerebrospinal fluid findings did not substantiate a bacterial infection. Rather, they favored a viral infection.

Another cause of acute onset of neurologic symptoms would be some form of intracranial bleeding. In this regard, the patient did fall twice, once on the right cheek and once on the calvarium. However, these falls were not significant and it would be doubtful that they could have produced any significant intracranial injury. So the past history of trauma also seemed to be out of the picture.

Now what about the endless number of white matter demyelinating diseases? These are metabolic or genetic in origin and for the most part are difficult to initially identify. They may or may not have acute onset, and because of this the patient was tested for these conditions. However, none were identified. As a result, the investigation again returned to infection and, in this regard, the patient was assessed for Epstein-Barr, influenza, parainfluenza, and other viruses. The results were negative, but eventually it was determined that the patient did have a viral gastroenteritis some 10 days prior to onset of these most recent symptoms (an interesting finding).

The findings on the subsequent MR study did not fit well with a viral encephalitis, but were more in keeping with

acute disseminated encephalomyelitis. This condition, probably autoimmune in pathophysiologic mechanism, often has onset after vaccination or viral infections.¹ Most often changes occur supratentorially but occasionally the posterior fossa (pons-cerebellum) is primarily involved.²⁻⁴ This is what occurred in our patient. The supratentorial findings were not as profound as the infratentorial, pontine, and parapontine findings (Fig. 2).

The patient was placed on antibiotic and antiviral medication as soon as assessed in our ER. There was contemplation of adding steroid therapy but the patient began to rapidly improve on this therapeutic regimen. Therefore, steroids were not administered. After 3 or 4 days the patient was discharged with only minimal neurologic residua and was to be followed in the Neurology Clinic. Our patient was fortunate because often this disease is more devastating and results in death, or at least serious neurologic deficit.

REFERENCES

1. Barkovich AJ. *Pediatric Neuroimaging*. 3rd ed. Baltimore, MD: Lippincott Williams & Wilkins; 2000:102-106.
2. Baum P, Barkovich AJ, Koch T, et al. Deep gray matter involvement in children with acute disseminated encephalomyelitis. *AJNR Am J Neuroradiol*. 1994;15:1275-1283.
3. Gregorio L, Sutton CL, Lee DA. Central pontine myelinolysis in a previously healthy 4-year-old child with acute rotavirus gastroenteritis. *Pediatrics*. 1997;99:738-743.
4. Shian WJ, Chi CS. Fatal brainstem encephalitis caused by Epstein-Barr virus. *Pediatr Radiol*. 1994;24:596-597.

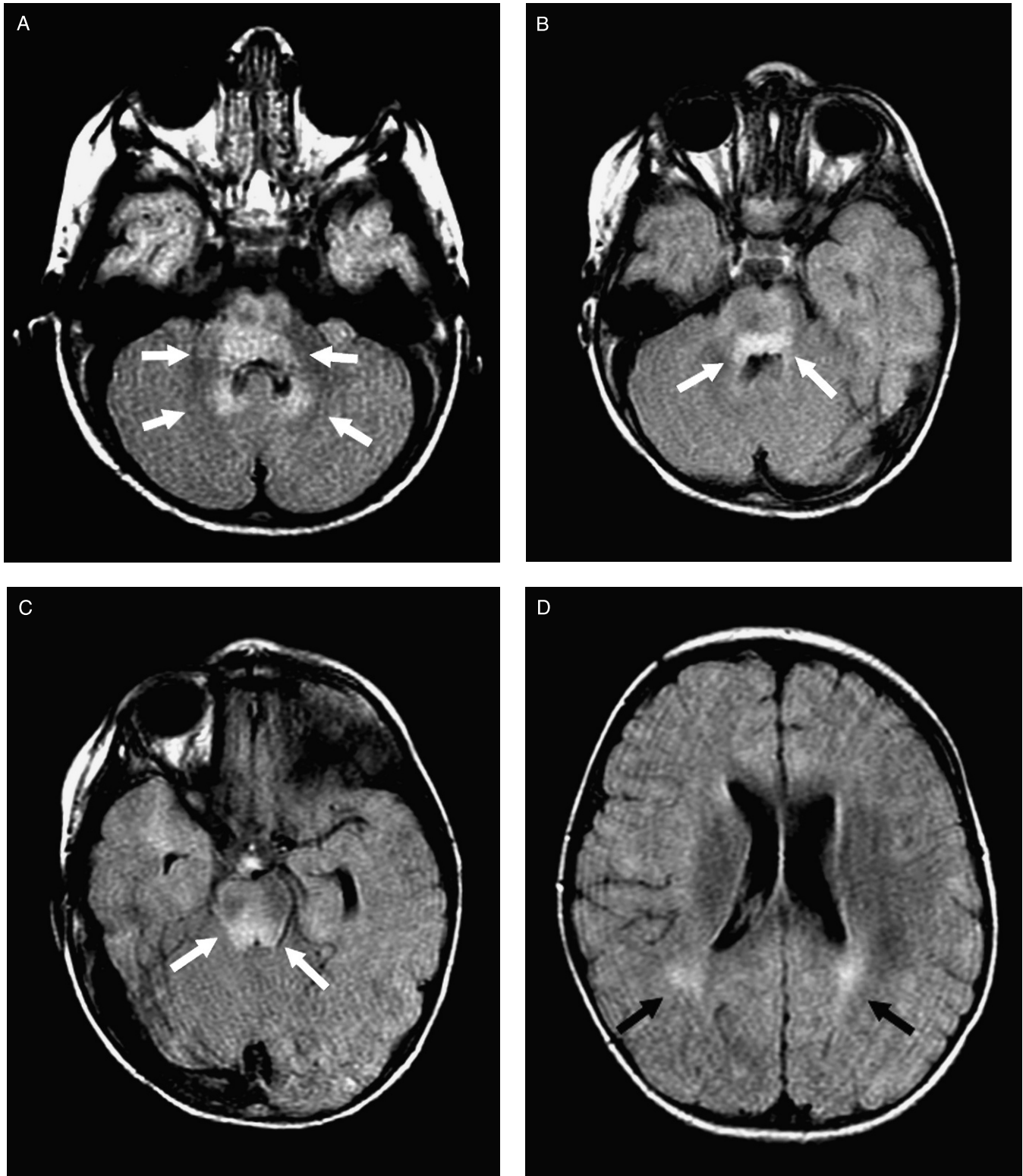


FIGURE 2. Four axial (FLAIR) images of the brain. A, Lowermost slice demonstrates increased signal along the superior aspect of the cerebellum (arrows). B, A slightly higher slice demonstrates increased signal in the pons (arrows), just adjacent to the fourth ventricle. C, A slightly higher image demonstrates more diffuse high signal in the brainstem (arrows). D, A supratentorial axial image demonstrates subtle, but nonetheless abnormal increased signal in the white matter of the posterior, paraventricular regions (arrows). All of these findings affect the white matter.