

Lacunar Infarction of the Basal Ganglia as a Complication of Hemolytic-uremic Syndrome

MRI and Clinical Correlations

Francis J. DiMario, Jr., MD,* Helen Brönte-Stewart, MD,‡
Joseph Sherbotie, MD,† Mary Ellen Turner, MD†

Central nervous system (CNS) complications of hemolytic-uremic syndrome (HUS) commonly consist of alterations in mental status, seizures, and rarely hemiparesis.^{1,2} The authors report the clinical evolution of left hemiparesis and later choreo-athetoid movements in a patient who sustained a right lacunar infarction as a complication of HUS. The infarction is demonstrated on magnetic resonance imaging (MRI).

CEREBRAL INFARCTION as a complication of hemolytic-uremic syndrome (HUS), is uncommon.¹ Although the pathophysiology of this syndrome involves small vessel occlusions, cerebral occlusive syndromes have been reported infrequently. We report the clinical and magnetic resonance imaging (MRI) findings of a patient who developed a lacunar infarction of the basal ganglia as a complication of HUS.

Case Report

A previously healthy 5.5-year-old right-handed girl was admitted after a 7-day evolution of dehydration and bloody diarrhea. On the day of admission she complained of headache and light headedness. Family history was significant for a maternal first cousin who suffered a grand-mal seizure during an episode of hemolytic-uremic syndrome (HUS). Her general examination was notable for parched mucous membranes. She had a normal neurological exam except for mild encephalopathy manifested by

inattention and disorientation. Laboratory data upon admission revealed thrombocytopenia, hemolytic anemia, and azotemia consistent with a diagnosis of HUS (Table 1). Urinalysis was positive for proteinuria, hematuria, and granular casts. No stool culture was done. She remained hospitalized for 7 days after receiving an infusion of fresh frozen plasma (FFP) and washed, packed, red blood cells (PRBC). Her mental status gradually returned to normal with improvement of her metabolic and hematologic abnormalities. She was discharged home on day 15 of illness.

Two days after discharge she was noted to have difficulty climbing stairs and noticeable clumsiness of her left hand. There were no complaints of vertigo, visual changes, or difficulty swallowing. Examination on readmission revealed a left hemiparesis, predominantly involving the distal anti-gravity musculature of the leg more than the arm and not involving the face. Mental status, cranial nerve, and sensory exams were normal. She was treated with infusions of FFP as well as receiving physical and occupational therapy. A head computerized tomography scan with and without contrast 3 days after the onset of weakness was normal. She was discharged home 27 days later still demonstrating a mild left hemiparesis, from a probable right hemispheric lacunar infarction.

Three days following her second discharge the patient was noted to be "speaking out of the left side of her mouth." Examination subsequently revealed an increased left hemiparesis with new findings of left facial droop, left palatal paresis and tongue deviation

From the Divisions of *Pediatric Neurology, †Nephrology and Pediatrics, The Children's Hospital of Philadelphia; and ‡The Department of Neurology, The Hospital of the University of Pennsylvania, Philadelphia, Pennsylvania.

Correspondence to: Francis J. DiMario, Jr., MD, Division of Neurology, The Children's Hospital of Philadelphia, 34th Street & Civic Center Blvd, Philadelphia, PA 19104.

Received for publication May 1987, revised July 1987, and accepted August 1987.

TABLE 1. Clinical and Laboratory Data

Number of Days After Onset of HUS When Neurological Deficit Appeared	Neurologic Exam	Serum Na ⁺ (mEq/dl)	Hgb (g/dl)	Hct (%)	Platelets	Retics	Smear	Great/BUN (mg/dl)/(mg/dl)	Urine Output	Radiologic Studies	Treatment
8 days	Mild encephalopathy; BP 110/66	134	8.3	23.7	44K	8.2%	+Fragmented cells	3.9/112	2.5 cc/kg/hr	None	PRBC* 5 cc/kg/day for 1 day FFP* 5 cc/kg/day for 1 day
17 days	Pure motor left hemiparesis not involving face; BP 110/70	135	7.7	21.7	378K	26%	+Fragmented cells	0.6/11	2.0 cc/kg/hr	Head CT with and without contrast; normal 3 days after ictus	FFP† 10 cc/kg/day for 5 days
30 days	Increased left hemiparesis with onset of left facial droop; tongue deviation to left and transient left palatal paresis; BP 116/76	138	10.7	30.9	301K	12%	+Few fragmented cells	0.7/15	1.0 cc/kg/hr	Head CT with and without contrast; normal 3 days after ictus MRI: subacute hemorrhage right caudate and lenticular nucleus 30 days after ictus	FFP 20 cc/kg/day for 11 days

Factor VIII, Protein C, Antithrombin III; Bleeding time, PT, PTT; all normal.

† FFP = Fresh Frozen Plasma.

* PRBC = Packed Red Blood Cells.

to the left. Mental status and the remainder of her cranial nerves and sensory exams were normal. She again received infusions of FFP with continued physical and occupational therapy. An MRI of the brain and brainstem 30 days after the initial onset of weakness revealed a subacute hemorrhage of the right caudate and lentiform nuclei without demonstrable brain stem lesions (Fig. 1). She was discharged home with a complete resolution of palatal weakness and partial resolution of her left hemiparesis, facial droop, and tongue deviation. Subsequently, she has developed an involuntary movement disorder. This is predominantly characterized by choreo-athetoid movements of the left arm.

Discussion

The CNS manifestations of HUS have been reviewed recently.^{1,2} These complications range from minor alterations in mental status such as lethargy and irritability, to major neurologic signs including coma, decerebrate posturing, seizures, and hemiparesis. Children with CNS dysfunction in association with HUS tend to have lower minimum serum sodium concentrations, more severe azotemia, and are more likely to require dialysis than children without CNS signs.¹ Coma and elevated CSF protein have been features associated with poor outcome.¹

Cerebral infarction as a complication of HUS has been documented infrequently. Neuropathologic reports of patients with HUS who suffered from CNS symptoms have had small vessel thromboses demonstrated within the CNS.^{2,3,4,5} A single patient at autopsy had focal areas of infarction with surrounding edema and necrosis in both cerebral cortex and basal ganglia.⁴ Crisp *et al.* described two patients with severe neurological disturbances and CT abnormalities.⁶ Each patient initially had peripheral wedge-shaped hypodensities that later evolved into faint hemorrhagic infarctions. They emphasized the utility of the CT scan in distinguishing those patients whose CNS dysfunction may be on a structural basis from those with a metabolic etiology.

Our patient presented with a common CNS manifestation of HUS, mental status changes. This mild encephalopathy cleared with resolution of the underlying metabolic disturbances. A recurrence of CNS dysfunction manifested by a progressive pure motor hemiparesis fits well with a thrombotic lacunar type cerebral infarction. These are typically small vessel, subcortical occlusions producing highly focal clinical syndromes.⁷ Pure motor hemiparesis can be seen with vascular lesions involving the internal capsule and basal ganglia, the lower basis pontis, and the midportion of the cerebral peduncles.⁷ These lesions result from vascular occlusions of penetrating branch arteries from the middle cerebral, basilar, and upper vertebral arteries. The CT scan may demonstrate a discrete infarction of limited dimension (several millimeters), confined to these subcortical structures. This is distinct from the wedge shaped lesions seen

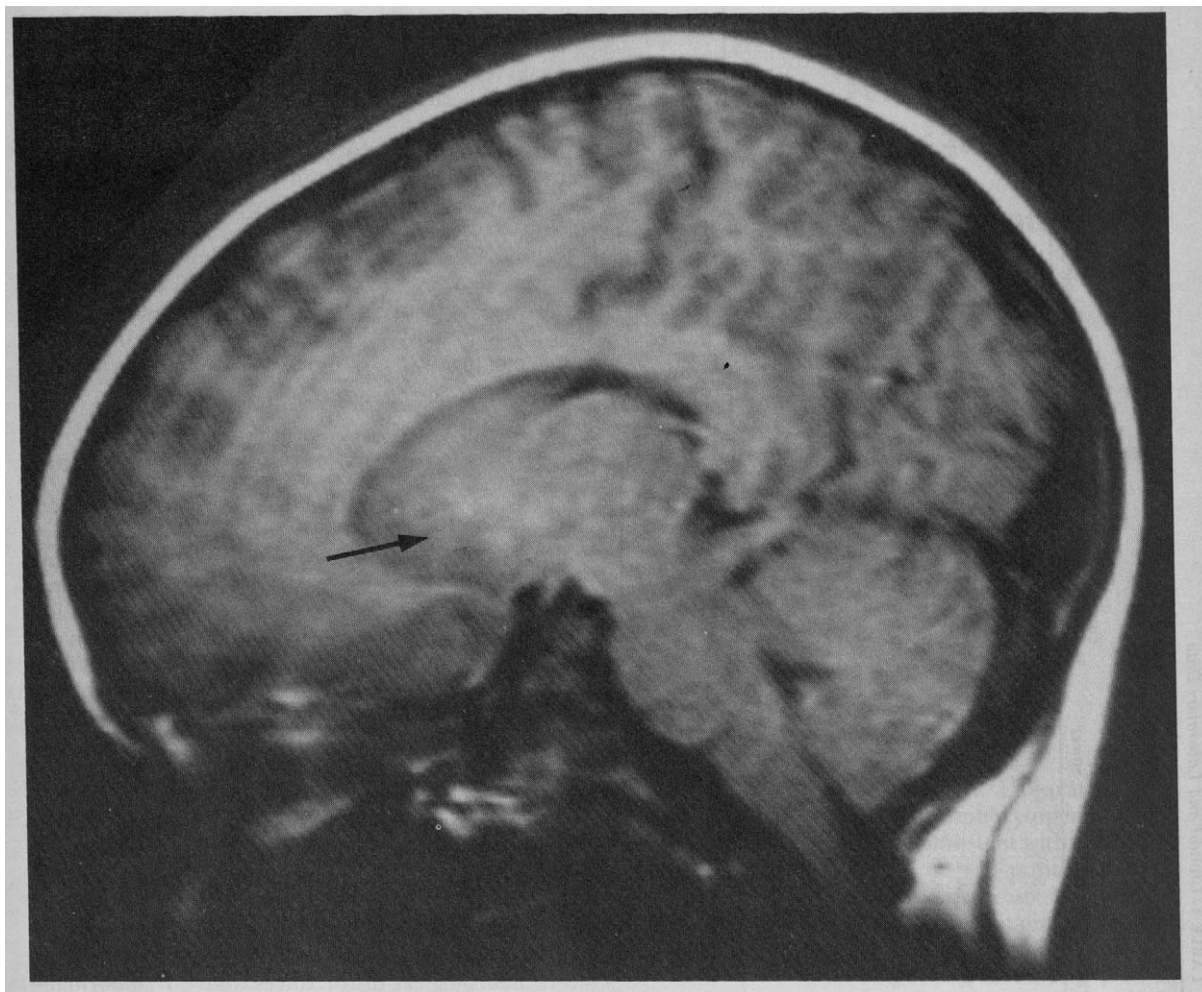


FIG. 1. A. T₁ weighted sagittal image; B. T₂ weighted axial image. Both depict punctate foci of increased intensity seen in the region of the right caudate and lentiform nuclei (arrow, arrowheads).

with infarctions of major arteries.⁸ Lacunae, if sufficiently small, may not be well visualized by CT within the first several days. This was the case in our patient, where the CT scan was normal 3 days after her infarction. Nonetheless, even small lacunae are generally demonstrated on CT 2 to 3 weeks after onset.⁸ The study was helpful in ruling out a large infarction, major hemorrhage, and a mass lesion. Each of these entities would have had its own therapeutic implications. Our patient's evolution of increased motor deficits probably represented the clinical expression of microscopic hemorrhage into a previously ischemic area. The MRI demonstrated this (Fig. 1). The facial droop, palatal paresis, and tongue deviation were manifestations of an upper motor neuron dysfunction (*i.e.*, corticobulbar fibers).

Intracranial hematomas can be placed into various stages of evolution. Based upon their characteristic intensity patterns as seen on MRI, a lesion can be

staged as acute (<1 week old), subacute (>1 week and <1 month old), or chronic (>1 month old).⁹ The MRI in our patient is a subacute lesion because of its hyperintensity on both the T₁ and T₂ weighted images. A lesion 1 week to 1 month of age conformed well to the clinical history.

This case provides both clinical and neuroimaging evidence for the microvascular thrombotic complications of HUS within the central nervous system. Neither lacunar infarction in HUS nor MRI of this lesion in HUS, to our knowledge, has been reported previously.

Acknowledgments

We express our appreciation to Dr. Larissa T. Bilaniuk for neuro-imaging interpretation, Dr. Samuel Tucker for reviewing the manuscript, and Linda Cella for preparing the manuscript.

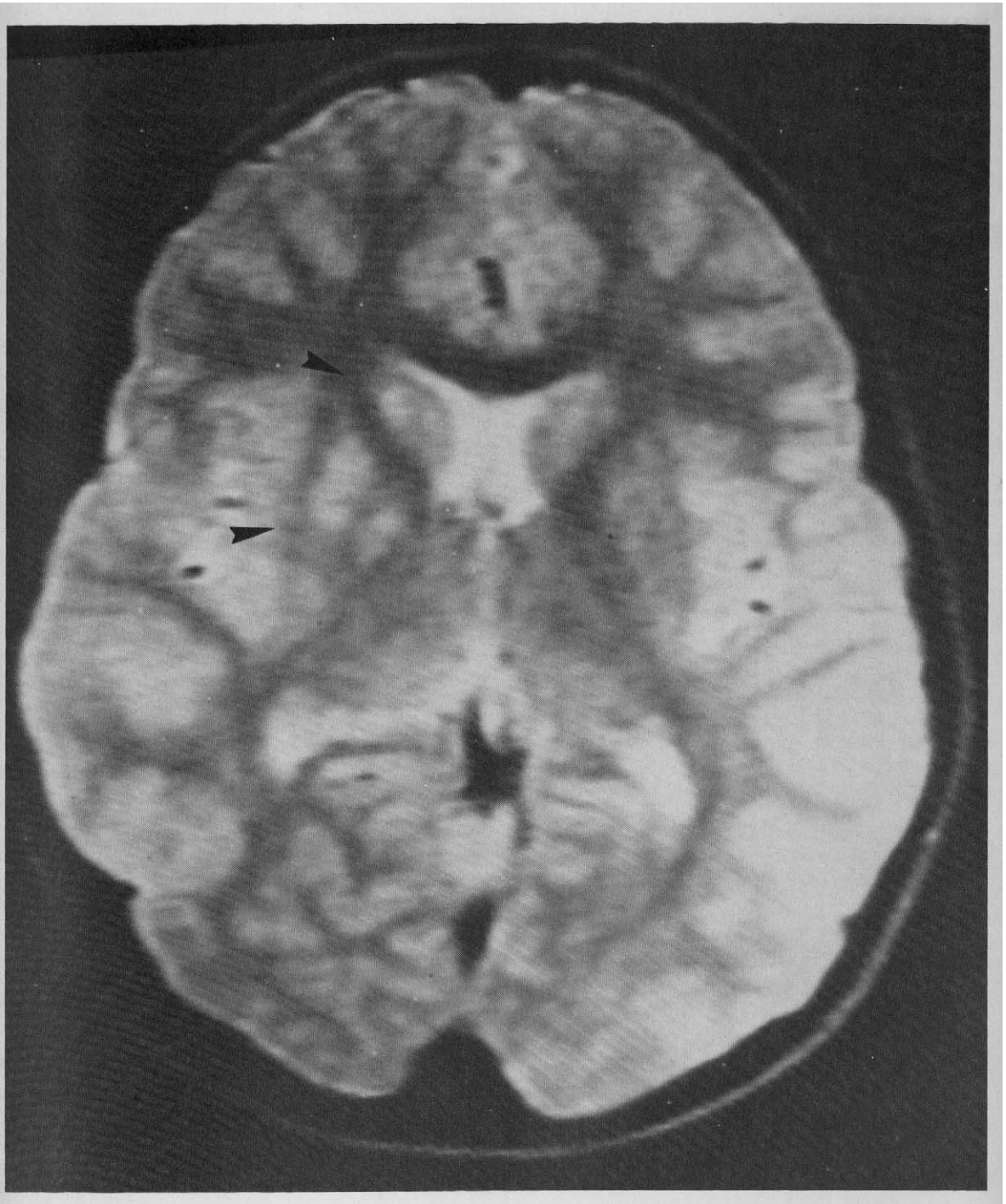


FIG. 1. (*Continued*)

References

1. Bale JR, Brasher C, Siegler RL. CNS manifestations of the hemolytic-uremic syndrome. *Am J Dis Child* 1980;134:869-72.
2. Gianantonio CA, Vitacco M, Mendilaharsu F, Gallo GE, Sojo ET. The hemolytic-uremic syndrome. *Nephron* 1973;11:174-92.
3. Gianantonio CA, Vitacco M, Mendilaharsu F, Rutty A, Mendilaharsu J. The hemolytic-uremic syndrome. *J Pediatr* 1964;64:478-91.
4. Upadhyaya K, Barwick K, Fishaut M, Kashgarian M, Siegel N. The importance of non-renal involvement in hemolytic-uremic syndrome. *Pediatrics* 1980;65:115-20.
5. Rodney JC, Anderson R McD, Hopkins IJ. Clinical and pathological aspects of central nervous system involvement in the hemolytic-uremic syndrome. *Aust Paediatr J* 1971;7:28.
6. Crisp DE, Siegler RL, Bale JF, Thompson JA. Hemorrhagic cerebral infarction in the hemolytic-uremic syndrome. *J Peds* 1981;99:273-6.
7. Fischer CM. Lacunar strokes and infarcts: A review. *Neurology* 1982;32:871-6.
8. Lee SH, Rao KCVG. In *Cranial Computed Tomography*. New York: McGraw-Hill, 1983;583-658.
9. Gomori JM, Grossman RI, Goldberg HI, Zimmerman RA, Bilaniuk LT. Intracranial hematomas; Imaging by high-field MR. *Radiology* 1985;157:87-93.