Case Reports

Detection of Cerebral Vasculopathy in Sickle Cell Disease Using Transcranial Doppler Ultrasonography and Magnetic Resonance Imaging

Case Report

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We describe a case of homozygous sickle cell anemia in which noninvasive transcranial Doppler ultrasonography and magnetic resonance imaging were used to detect angiographically documented occlusive lesions of intracranial arteries. (*Stroke* 1988;19:518–520)

Cerebral infarction is recognized as an important complication of sickle cell anemia (HbSS).[1-4] Extensive angiographic and neuropathologic data have demonstrated occlusive lesions involving the basal intracranial arteries, particularly the distal internal carotid (ICA), proximal middle cerebral (MCA), and anterior cerebral (ACA) arteries.[5-8]

Although angiography clearly demonstrates the predisposing lesions, it is invasive and is reserved for patients being considered for long-term transfusion therapy who have already developed symptoms or suffered infarction. We present a case with sickle cell anemia that demonstrates the ability of transcranial Doppler ultrasonography (TCD)[9-11] and magnetic resonance imaging (MRI)[4] to detect angiographically documented occlusive lesions of intracranial arteries.

Case Report

A six-year-old black girl with homozygous HbSS was admitted with the acute onset of right hemiparesis and decreased speech. Ten months before admission, she was diagnosed as having absence seizures by electroencephalography but had no other neurologic history. She had not been on a program of regular exchange transfusion.

She was alert but had diminished motor power in her right arm (2/5) and right leg (4/5). Computed tomography (CT scan) (GE 9800, Schenectedy, New York) performed 3 days after the onset of symptoms showed a large area of decreased attenuation encompassing the MCA distribution on the left.

MRI (GE 1.5 Tesla Signa Unit) was performed 15 days after admission according to a research protocol designed to enhance visualization of the basal vessels most often involved in HbSS. Five scanning series were performed: 1) sagittal localization views using the partial saturation technique [resonance time (TR), 800; echo time (TE), 20 msec] with 3-mm slices separated by 1 mm, 2) coronal partial saturation views using the parameters as in 1, 3) axial partial saturation views using the parameters as in 1, 4) oblique 30° sagittal partial saturation localization views (TR, 600; TE, 20 msec) with 5-mm slices separated by 0 mm, and 5) oblique coronal partial saturation views using the parameters as in 1.

MRI revealed an attenuated distal ICA on the left with no visualization of the proximal left MCA in any view. In both coronal series, the proximal segment of the right ACA appeared markedly stenotic (Figure 1, left and right).

Four-vessel angiography was performed after hydration and reduction of hemoglobin S to <30% by partial exchange transfusion. Angiography demonstrated several abnormalities (Figure 2): 1) moderate stenosis of the left supraclinoid ICA, 2) occlusion of the left MCA with reconstitution of distal MCA branches by leptomeningeal collaterals, and 3) severe stenosis of the right ACA in its proximal segment. No abnormalities were noted in the cervical carotid arteries or on posterior circulation views.

TCD (TC2-64B, EME, Uberlingen, FRG) was performed with 2-MHz pulsed Doppler using the transtemporal approach (transtemporal diameter 140 mm). Hematocrit at the time of TCD examination was

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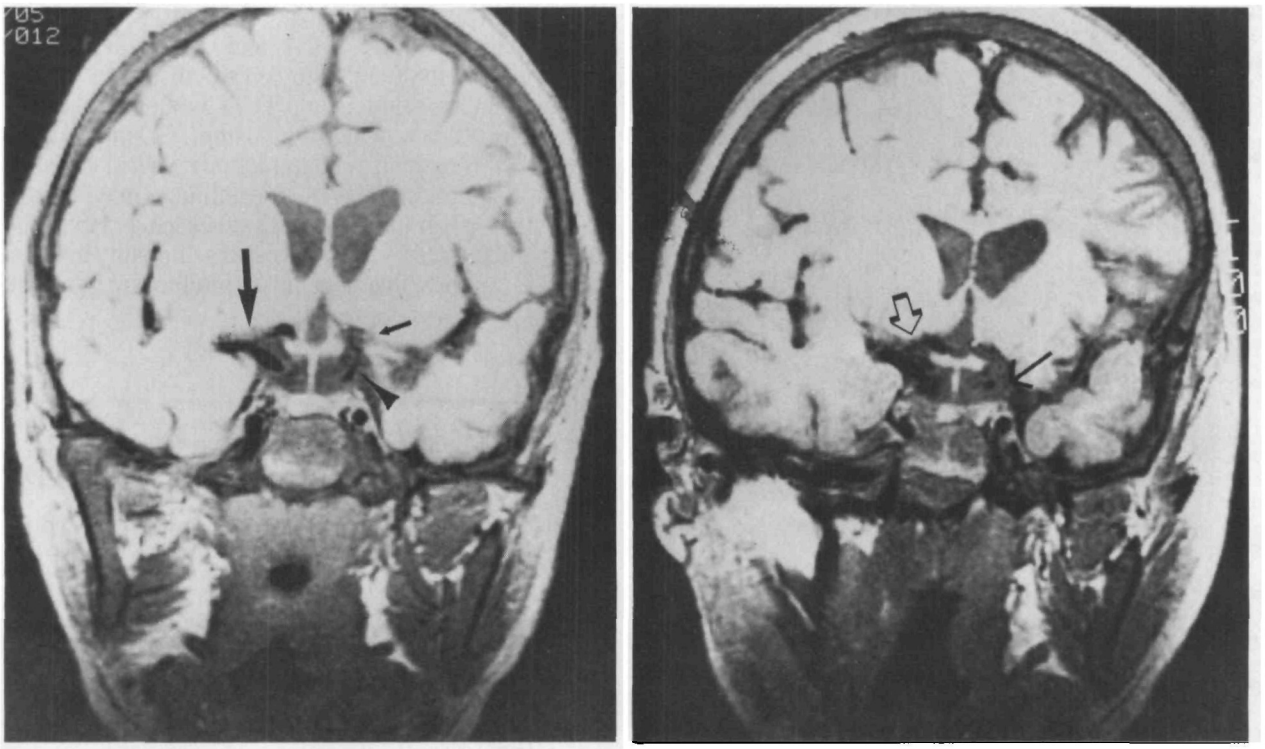
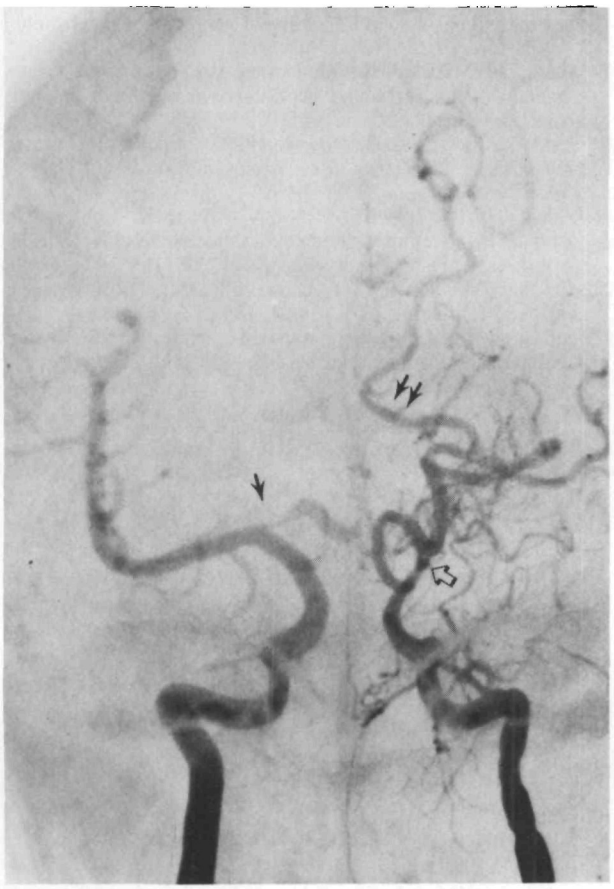


FIGURE 1. Left: *Coronal T1-weighted magnetic resonance image (MRI) showing apparent marked stenosis of proximal right anterior cerebral artery (ACA) (large arrow). Distal internal carotid (ICA) and proximal middle cerebral (MCA) arteries on right appear normal. On left, distal ICA is stenotic (arrowhead) and proximal segment of MCA is not visualized (small arrow).* Right: *Oblique (left side 30° forward) coronal T1-weighted MRI again showing proximal right ACA stenosis (open arrow). In this section, stenotic distal segment of left ICA is cut axially, demonstrating reduced luminal diameter (arrow).*



26%. No left MCA blood flow could be demonstrated by TCD; at a depth of 70 mm, left ACA blood flow toward the probe (reversed flow) was observed. On the right, normally directed MCA blood flow was detected at depths of 45 and 50 mm (Figure 3, top), with velocities higher than those seen in normal adults but consistent with this patient's age and hematocrit.[11] High-velocity blood flow away from the probe in the left ACA was recorded at a depth of 60 mm (Figure 3, bottom). Peak systolic and mean velocities at this depth were estimated to be 300 and 220 cm/sec, respectively, compared with 200 and 128 cm/sec in the right MCA. The distance between the temporal soft tissue and the ACA stenosis on MRI coronal views was estimated to be 57.5 mm.

Discussion

The ability of MRI to image vascular structures based on "signal void" is well recognized,[12] but its potential for detecting focal stenosis or occlusion of

FIGURE 2. *Cerebral angiogram with both internal carotid artery (ICA) injections superimposed. Proximal segment of left anterior cerebral artery is markedly stenotic (single arrow). On right, supraclinoid ICA is stenotic and proximal segment of middle cerebral artery (MCA) is not filled with contrast. Posterior cerebral artery (double arrows) is prominent and provides collaterals that reconstitute distal branches of left MCA in later views.*

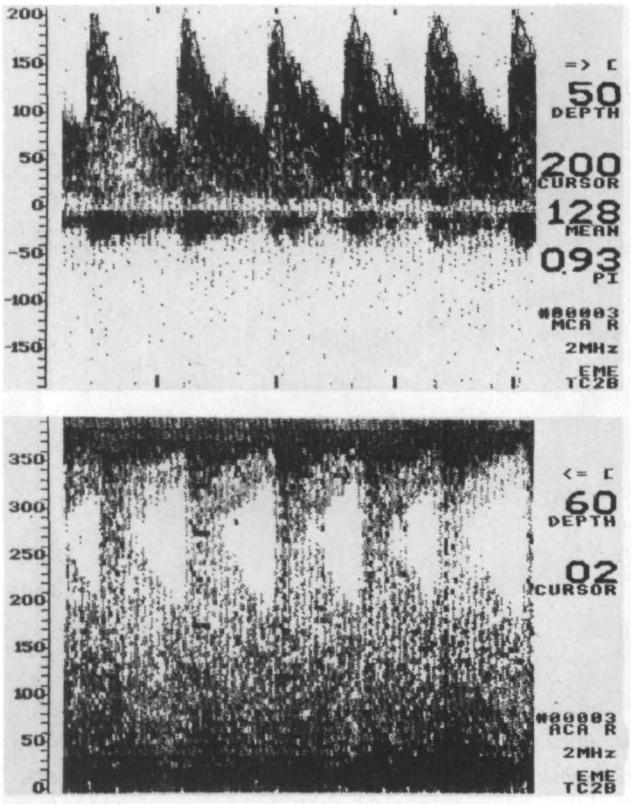


FIGURE 3. Top: *Transcranial Doppler display from right middle cerebral artery (MCA) at depth of 50 mm from right temporal window;* y *axis, velocity in cm/sec;* x *axis, time in 1-second intervals. Flow toward transducer (physiologic direction) is indicated by arrow in upper right corner.* Bottom: *Transcranial Doppler display at 60 mm depth showing markedly elevated velocity in right anterior cerebral artery (ACA). Zero line has been moved to bottom of display to show complete velocity profile of ACA. Although systolic peaks are partly obscured by representation of blood flow from MCA in opposite direction (spectra coming down from top of display), it is apparent that peak velocity is >300 and mean velocity is >>200 cm/sec in proximal ACA. Flow is away from transducer, in the physiologic direction.*

intracranial arteries has received little attention. This case demonstrates the ability of MRI to detect focal abnormalities of intracranial arteries. To improve vessel visualization, we recommend that the MRI protocol be modified to provide thin, closely spaced sections and supplemental oblique coronal views. Additional evidence for stenosis can be obtained from TCD. The proximal MCA and ACA, areas often involved in HbSS and moyamoya disease,[13] can be reliably interrogated using TCD without hazard or discomfort. It is now feasible, using TCD and MRI, to detect HbSS patients with intracranial vascular disease before disabling stroke. These techniques may limit the need for angiography in conditions such as HbSS that are characterized by intracranial vasculopathy and may provide noninvasive means to monitor response to therapy.

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KEY WORDS • anemia, sickle cell • magnetic resonance • ultrasonic