

Abnormal Vascular Drainage in Anomalies of Neuronal Migration

A. James Barkovich^{1,2}

Four patients are presented who had apparently anomalous vascular channels that were initially erroneously interpreted as vascular malformations. These vascular channels represent the normal venous drainage of a pachygyric cortex. The recognition of this finding as a manifestation of the underlying abnormal neuronal migration is important in order to avoid further intervention in these patients.

Anomalies of neuronal migration are relatively common malformations of the brain. Crome and Stern [1] observed migration anomalies in more than 5% of 500 consecutive autopsies of mentally deficient patients. These anomalies are being recognized with increasing frequency in patients who have developmental delay and seizure disorders [2, 3]. We recently encountered four patients who had focal migration anomalies and associated large vascular channels, an observation that has not previously been reported. The purpose of this report is to promote awareness of this association and thereby avoid unnecessary intervention.

Subjects and Methods

The ages of the four patients studied were 9 months, 7 years, 35 years, and 36 years. The three oldest patients had seizure disorders and the youngest was hemiparetic; all were developmentally delayed. None of these patients is included in the author's prior study of neuronal migration anomalies [2].

The three oldest patients had only MR imaging. One was scanned on a GE unit and two on a Disonics MT/S scanner. Sagittal spin-echo (SE) images were obtained by using a repetition time (TR) of 600 msec and an echo time (TE) of 20 msec. Axial images were obtained by using an SE 2500/35-70 sequence. Additional coronal SE 600/20 images were obtained in one patient. The youngest patient was evaluated with CT; axial 10-mm scans were obtained with and without contrast. The 9-month-old infant, the first in our series, underwent standard transfemoral cerebral arteriography.

Results

All patients had an abnormally deep sulcus that was lined by thickened cortex. The abnormal cortex was seen to involve a variable amount of the hemisphere beyond the cleft in the different patients (Figs. 1-4). In each case, a large vascular structure was seen running within the sulcus, enlarging in diameter in the more rostral sections. The diameter of the vessel was roughly proportional to the amount of affected cortex. On both the contrast-enhanced CT and the MR studies, smaller vessels were seen to run from the thickened cortex into the large draining vessel, raising the possibility of a vascular malformation (Fig. 1). The lateral ventricle was enlarged and the thickness of the centrum semiovale diminished in size in the affected hemispheres in all patients. One patient had a region of high signal intensity lining the ventricle on the long-TR images (Fig. 3).

Received October 14, 1987; accepted after revision January 27, 1988.

The views expressed in this article are those of the author and do not reflect the official policy or position of the Department of the Army, Department of Defense, or the U.S. Government.

¹ Department of Radiology, Letterman Army Medical Center, Presidio of San Francisco, CA 94129-6700. Address reprint requests to Technical Publications Editor, HSHH-CI-ME.

² Department of Radiology, Section of Neuroradiology, University of California School of Medicine, San Francisco, CA 94143.

AJNR 9:939-942, September/October 1988
0195-6108/88/0905-0939

© American Society of Neuroradiology

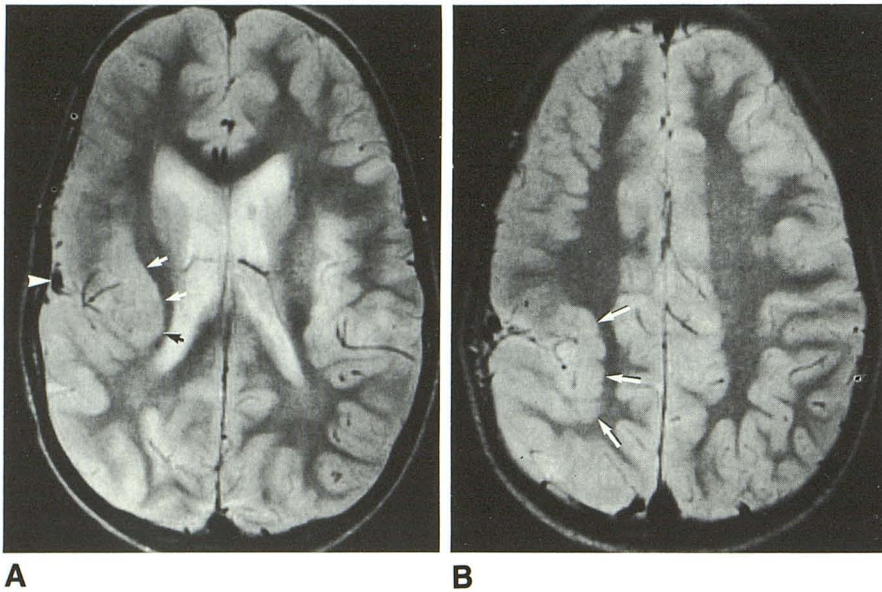


Fig. 1.—Axial SE 2500/35 images of 7-year-old child.

A, Image at level of bodies of lateral ventricles. Cortex is abnormally thick in right opercular region (arrows) with thinning of underlying white matter and a single large, deep sulcus in posterior sylvian region. Several small vessels run centrifugally within cleft from a large vascular structure that lies at orifice of the sulcus (arrowhead).

B, At a higher level, abnormal cortex and single large sulcus (arrows) are more pronounced. More small vessels are seen within sulcus, draining into larger, superficial venous structure.

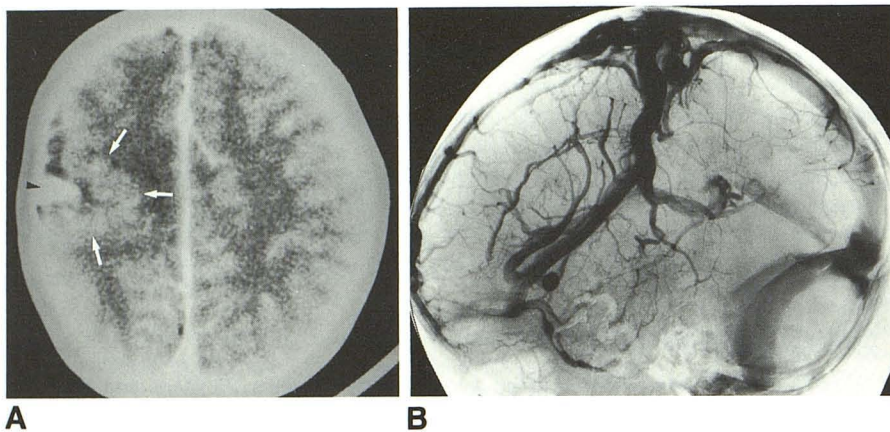


Fig. 2.—9-month-old infant.

A, Axial, contrast-enhanced CT scan shows abnormally thickened cortex and a single deep sulcus (arrows) in right cerebral hemisphere. A large, contrast-enhancing vessel is seen within sulcus (arrowhead). (On other images, smaller, enhancing vascular structures were seen radiating toward the large vessel.)

B, Lateral view from venous phase of right internal carotid arteriogram. Large vessel seen on CT scan is shown to be a large superficial vein, which drains majority of superficial veins of the hemisphere. (No arteriovenous shunting and no abnormal tangles of vessels were noted on previous films in the sequence.)

Left carotid and vertebral angiography was performed on the 9-month-old infant by a transfemoral approach. Films obtained in the venous phase of the left internal carotid artery injection showed a large vein originating in the left anterior sylvian region and draining superficially into the superior sagittal sinus (Fig. 2). The majority of the superficial venous drainage in the hemisphere appeared to course through this venous channel. Films in the arterial phase showed diminished tortuosity of the opercular vessels, a finding previously described in pachygyric cortex [4]. There was no arteriovenous shunting, and no abnormal tangles of vessels were identified. The remainder of the venous drainage in the brain was unremarkable.

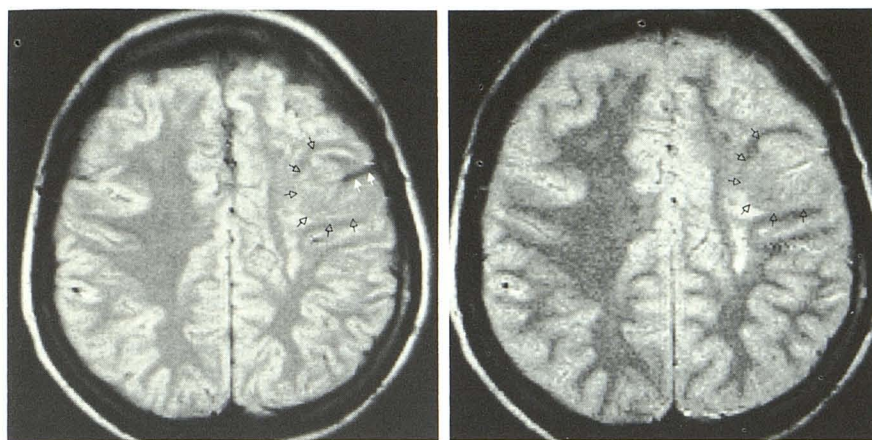
Discussion

The phenomenon of neuronal migration was first described near the turn of the century [5]. The neurons that constitute the mammalian brain are generated in proliferative zones

situated along the ventricular surface of the developing brain [5–8]. At the end of the second gestational month, the neurons migrate from their sites of origin along radially aligned glial cells to relatively distant final positions [7, 8].

The major cell migration activity lasts about 2 months, beginning in the eighth fetal week and ending at about week 16. Smaller waves of cell migration continue up to week 25. An insult to the brain during this period results in a migration anomaly. A more detailed description of the migration anomalies and their radiographic appearance has recently been published [2].

There is evidence [9, 10] that migration anomalies are vascular in origin and that episodes of hypoperfusion cause a laminar necrosis in the developing telencephalon. It has been proposed that the damaged tissue in this necrotic layer includes the radial glial cells; damage to these cells prevents further neuronal migration and leads to a thickened, disorganized cortex [9]. The affected area retains the fetal appearance of a smooth cortex with a few deep convolutions. Although



A

B

Fig. 3.—35-year-old patient.

A, Axial SE 2000/40 image shows large infolding of gray matter in left frontal region (open black arrows). A large vessel (white arrows) is seen with sulcus.

B, Axial SE 2000/80 image shows spatial mismapping of signal from vessel (seen as increased signal posterior to vessel), indicating low (venous) flow within vessel. High signal intensity within left hemispheric white matter is presumably a result of gliosis. Open black arrows show large infolding of gray matter.

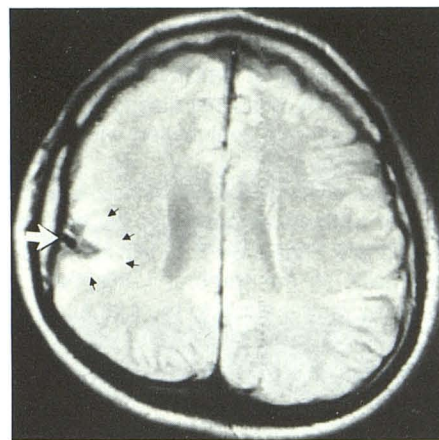
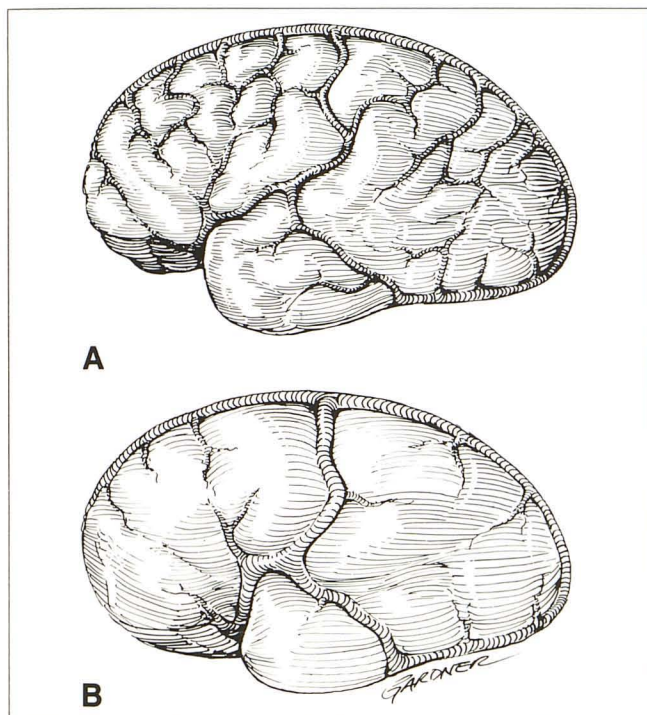


Fig. 4.—36-year-old patient. Axial SE 2000/35 image reveals an infolding of gray matter in right posterior sylvian region (small arrows). A large vessel is seen coursing within this enlarged sulcus (large arrow). Patient was referred for embolization of this vessel, presumed to be an arteriovenous malformation.



A

B

Fig. 5.—Comparison of venous drainage in normal and pachygyric brains.

A, Normal superficial venous drainage from brain. Veins form within cerebral sulci from coalescence of venules and venous channels running within cortex. These veins form cortical venous sinuses that drain into superior sagittal sinus.

B, Venous drainage in a pachygyric cortex. Cortex is thick with a flattened surface and a few, very large and deep convolutions. Venous blood draining the cortex is transported within small venous channels to larger vessels within the few large convolutions that are present. (In our patients, size of the large venous channel was roughly proportional to extent of the affected area.)

evidence has been presented for a genetic susceptibility to these anomalies [11, 12], the large majority of cases reported have been sporadic [13], lending further support to a vascular or infectious origin.

Larroche [14] has previously reported the presence of an angiomatous network of veins draining areas of polymicrogyric cortex. The explanation for this phenomenon can perhaps be found by examination of the underlying pathology in the pachygyric and polymicrogyric cortex. In the normal brain, veins form within the cerebral sulci from coalescence of venules and venous channels running within the cortex. These veins then form cortical venous sinuses that drain into the superior sagittal sinus (Fig. 5A). In the pachygyric cortices, normal convolutions of the brain do not form; instead, the cortex is thick with a flattened surface and a few very large and deep convolutions. In polymicrogyria there are many abnormal, very shallow sulci, which can disappear as the microgyri fuse [2].

In our patients, a single large vein appeared to drain the entire affected area of cortex. The size of the vessel, moreover, seemed to be proportional to the extent of the affected area. It seems, therefore, that the venous blood draining the cortex is transported within small venous channels to larger vessels within the few large convolutions that are present (Fig. 5B). In three of the patients the appearance corresponded to the primitive sylvian vein coursing in a fetal sylvian fissure similar to that seen in a normal fetus at 20–26 weeks gestation (Fig. 6) [7]. In the other patient, the deep sulcus was more anterior in location (Fig. 3). This finding implies that as normal cortical development ceases in the anomalous region, normal venous development may cease as well. The venous outflow from the abnormal cortex then drains via a network of small veins into a large venous channel running within the deep, primitive sulcus.

Although angiography was performed in order to better identify the nature of the large blood vessel in the primitive sulcus in our first patient (the 9-month-old), we no longer perform such additional diagnostic procedures when such a vessel is seen in conjunction with a thickened cortex and a deep sulcus.

Occasionally [15, 16], illustrations of deep convolutions similar to those in our series are thought to represent schizencephaly. These "clefts" are in fact simply deep convolutions. Normally, as the cerebral hemisphere matures, infolding of the gray matter leads to the formation of the normal gyri and sulci (Fig. 6). In hemispheres with impaired neuronal migration, the primitive sylvian fissure retains its shallow, vertical, fetal configuration. These sulci, however, do not communicate with the lateral ventricle, differentiating them from schizencephaly [17]. It is these primitive sylvian fissures in poorly developed cerebral hemispheres that were identified in three of our

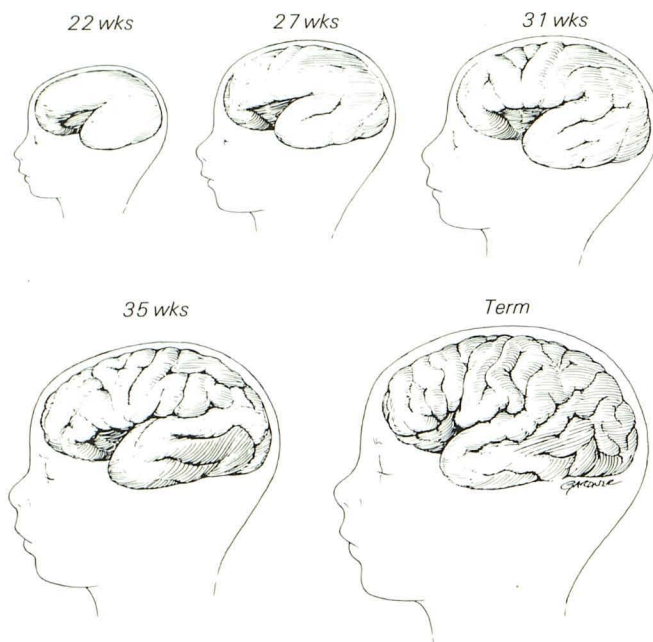


Fig. 6.—Gyral development in human brain. Notice how brain at 22 weeks gestation has a single, vertically oriented convolution, the primitive sylvian fissure. This appearance is similar to that of pachygyric or agyric hemisphere. As brain matures, secondary and tertiary gyri form, sylvian fissure becomes more horizontal in orientation, and lateral opening narrows. As normal cortical development ceases within the anomalous region, normal venous development may cease as well. Venous outflow from the abnormal cortex then drains via a network of small veins into a large venous channel running within the deep, primitive sulcus.

patients and in those patients described in the literature; these are not the holohemispheric clefts of schizencephaly.

In summary, we identified several patients who had migration anomalies with apparently anomalous vascular channels. It is important to recognize this anomaly as a manifestation of the underlying abnormal neuronal migration. Further intervention is not warranted in these patients.

REFERENCES

1. Crome L, Stern J. *The pathology of mental retardation*. London: Churchill, 1967
2. Barkovich AJ, Chuang SH, Norman D. MR of neuronal migration anomalies. *AJNR* 1987;8:1009-1017, *AJR* 1988;150:179-187
3. Byrd SE, Osborn RE, Bohan T, Naidich TP. MR evaluation of migration disorders of the brain in children. Presented at the 25th annual meeting of the American Society of Neuroradiology, New York, 1987
4. Daube JR, Chou SM. Lissencephaly: two cases. *Neurology* 1966;16:179-191
5. Larroche J-C. Malformations of the nervous system. In: Adams JH, Corsellis JAN, Duchon LW, eds. *Greenfield's neuropathology*. New York: Wiley, 1984:411-421
6. Ludwin SK, Malamud N. Pathology of congenital anomalies of the brain. In: Newton TH, Potts DG, eds. *Radiology of the skull and brain*, vol. 3. *Anatomy and pathology*. St Louis: Mosby, 1977:2979-3015
7. Lemire RJ, Loeser JD, Leech RW, Alvord EC. *Normal and abnormal development of the human nervous system*. Hagerstown, MD: Harper & Row, 1975:231-259
8. Rakic P. Neuronal migration and contact guidance in the primate telencephalon. *Postgrad Med J [Suppl 1]* 1978;54:25-37
9. Stewart RM, Richman DP, Caviness VS Jr. Lissencephaly and pachygyria. An architectonic and topographical analysis. *Acta Neuropathol (Berl)* 1975;31:1-12
10. Richman DP, Stewart RM, Caviness VS Jr. Cerebral microgyria in a 27-week fetus: an architectonic and topographic analysis. *J Neuropathol Exp Neurol* 1974;33:374-384
11. Dobyns WB, Stratton RF, Greenberg F. Syndrome with lissencephaly. I: Miller-Dieker and Norman-Roberts syndromes and isolated lissencephaly. *Am J Med Genet* 1984;18:509-526
12. Dobyns WB, Kirkpatrick JB, Hittner HM, Roberts RM, Kietzer FL. Syndromes with lissencephaly. II: Walter-Warburg and cerebro-oculo-muscular syndromes and a new syndrome with type II lissencephaly. *Am J Med Genet* 1985;22:157-195
13. Gastaut H, Pinsard N, Raybaud Ch, Aicardi J, Zifkin B. Lissencephaly (agyria-pachygyria): clinical findings and serial EEG studies. *Dev Med Child Neurol* 1987;29:167-180
14. Larroche J-C. Cytoarchitectonic abnormalities (abnormalities of cell migration). In: Vinken PJ and Bruyn GW, eds. *Handbook of clinical neurology*, vol. 30. *Congenital malformations of the brain and skull. Part I*. Amsterdam: North-Holland, 1977:479-506
15. Miller GM, Stears JC, Guggenheim MA, Wilkening GN. Schizencephaly: a clinical and CT study. *Neurology* 1984;34:997-1001
16. Bird CR, Gilles FH. Type I schizencephaly: CT and neuropathologic findings. *AJNR* 1987;8:451-454
17. Barkovich AJ, Norman D. MR imaging of schizencephaly. *AJNR* 1988;9:297-302, *AJR* 1988;150:1391-1396