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Arterial Vascular Abnormality Accompanying Cerebral Cortical Dysplasia

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Summary: We report a case of dysplastic arterial vascular abnormality in a 32-year-old man with overlying neuronal cell migration disorder. MR images showed a thickened left insular cortex adjacent to the abnormal vascular network. These findings suggest the possibility of leptomeningeal damage during neuronal cell migration as the cause of the overlying vasculopathy. The true pathogenesis of these seemingly associated abnormalities is unknown.

Index terms: Arteries, abnormalities and anomalies; Migration anomalies

Arterial vascular dysplasia associated with disorders of neuronal cell migration is a rare occurrence (1). We report a case of cortical dysplasia with an associated arterial abnormality and prominent venous drainage. Magnetic resonance (MR) imaging and cerebral angiography were helpful in demonstrating the lesion.

Case Report

A 32-year-old man was alert and oriented without neurologic deficit after a single-vehicle accident. His medical history was significant for medically controlled epilepsy since childhood. He reported only a few generalized seizures in the past. An electroencephalogram showed spike and wave complexes with sporadic slowing in the left frontal leads. MR images showed a small thickened cerebral cortex in the left insular and temporal regions with normal signal intensity (Fig 1A). The left sylvian fissure was shallow and vertical with an incomplete operculum. These findings suggested cerebral cortical dysplasia. Contrast-enhanced MR imaging did not show parenchymal enhancement; however, an enlarged vessel ascending over the parietal lobe cortex was noted (Fig 1B).

Cerebral angiography showed a dilated and ectatic arterial vascular network in the left sylvian fissure (Fig 1C and D). The arteries distal to this had a normal pattern with somewhat smaller distal cortical segments of the middle cerebral artery. There was no evidence of early venous drainage. A large superficial vein noted on the MR images drained into the superior sagittal sinus from the region of the dysplastic cortex (Fig 1E). Treatment with antiepileptic medication produced no change in the patient's clinical status.

Discussion

The phenomenon of neuronal migration begins in the eighth fetal week and ends around the 16th week, with smaller waves of cell migration continuing up to week 25 (2). Anomalies of neuronal migration may be divided into several categories depending on the timing and severity of the injury and the appearance of the resulting anomaly (3). Abnormal or prominent venous drainage has been described in association with cortical dysplasia (1). These anomalies do not appear to be arteriovenous shunts or fistulas. Our patient had a large superficial cortical vein draining into the superior sagittal sinus.

Previously, two cases of ectatic cerebral arteries with congenital disorders of cellular migration have been reported (4, 5). Doran et al (4) concluded that either congenital or acquired factors could produce the histologic outcome that displayed areas of smooth muscle atrophy replaced by fibrous tissue with hyaline changes

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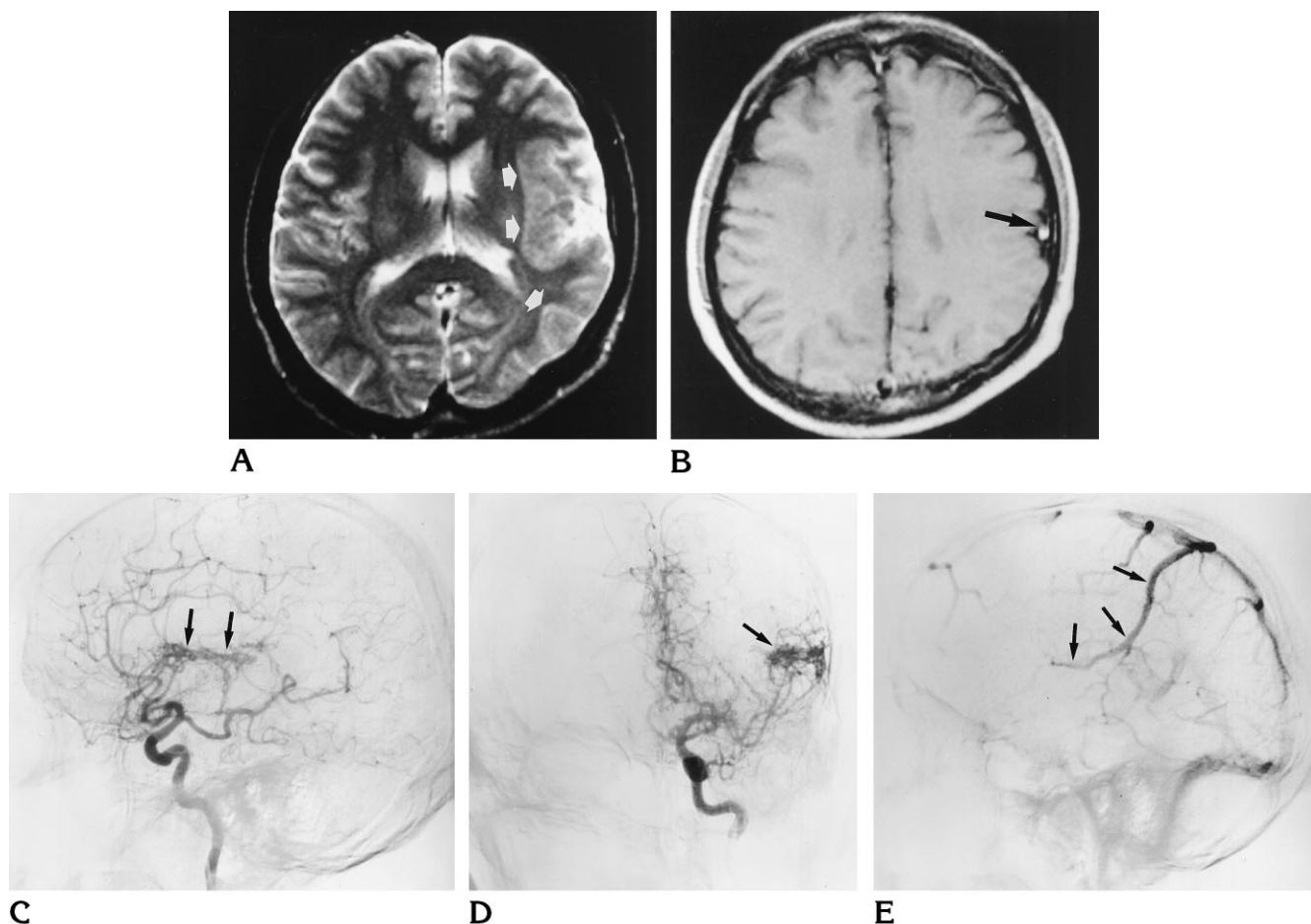


Fig 1. A 32-year-old man with history of medically controlled epilepsy since childhood who presented after a motor vehicle accident. A, T2-weighted (2000/120 [repetition time/echo time]) MR image shows thickened cerebral cortex of the left insula and temporal lobe (arrows). The signal of this lesion is similar to that of normal cortex. The sylvian fissure is shallow and wide.

B, Contrast-enhanced MR image (300/14; flip angle, 90°) shows enhancement in an enlarged vessel on the surface of the left frontoparietal region (arrow).

C and D, Anteroposterior and lateral views of left internal carotid angiogram show a tortuous arterial vascular network (arrows) involving the opercular segments of the left middle cerebral artery. The abnormal network is located in the left opercular cleft. No early venous filling is identified. Anterograde flow to the distal segments is noted.

E, Venous phase of the carotid angiogram shows an enlarged cortical vein (arrows) that was noted on an MR image (B) draining from the dysplastic cortex to the superior sagittal sinus.

in the media, while the intima is concentrically thickened.

According to Padget (6), the main arterial stems have an adult configuration by the 52nd day of gestation (± 1 day). Therefore, during the period of neuronal cell migration, cerebral arteries do not have a primitive vascular rete. The abnormal arterial network seen in the present case probably does not represent a persistent primitive vascular rete. Cortical dysplasia is a common manifestation of congenital cytomegalovirus infection (3). In an autopsy case of a 28-week-old fetus with disseminated cytomegalovirus infection, tortuous leptomeningeal vessels were noted along with a small

eruption of superficial cortical neurons, suggesting glial heterotopia (7). This suggests that leptomeningeal damage in the period of neuronal cell migration could induce a dysplastic arterial abnormality accompanying the cortical dysplasia. Possible causes of the disorder in the present case include a vascular reaction to prenatal infection or direct damage to the arterial media, resulting in the dysplasia encountered.

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