

Slow-Flow Vascular Malformations of the Pons: Capillary Telangiectasias?

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PURPOSE: To report clinical and MR features that suggest telangiectatic vascular malformations of the pons. **METHODS:** The MR scans and clinical data of 12 patients demonstrating an enhancing pontine lesion with minimal or no signal abnormality on T2-weighted images were reviewed. None of the patients underwent angiography or biopsy. Follow-up scans, available for all patients between 3 weeks and 40 months (range, 11.5 months), were reviewed. **RESULTS:** The patients presented with a variety of symptoms including headache (n = 4), vertigo (n = 3), gait abnormality (n = 3), and hearing loss (n = 2). Two were referred for biopsy or treatment of presumed pontine glioma. On precontrast MR, 3 of 12 lesions were isointense on both T1- and T2-weighted images. Three of 12 lesions were slightly hypointense on T1-weighted images and 8 of 12 were slightly hyperintense on T2-weighted images. Postgadolinium images showed a discrete focus of enhancement with irregular or brushlike borders. Eight of 12 had an anomalous draining vessel from the lesion to the surface of the pons. None demonstrated mass effect or hemorrhage. Gradient-echo sequences in 7 patients all showed marked T2 shortening, despite the absence of hemorrhage on either T1- or T2-weighted images. None of the follow-up scans showed radiographic or clinical progression. **CONCLUSION:** The benign clinical course, lack of mass effect, and minimal or no T2 prolongation argue against neoplasm and instead indicate a vascular cause. We suspect the decreased signal on gradient-echo sequences represents elevated intravascular deoxyhemoglobin from stagnant blood flow. The findings are atypical for cavernous angioma or classic venous malformation. Although pathologic confirmation is lacking, the radiographic features are most consistent with capillary telangiectasia or a transitional capillary-venous malformation. Despite the absence of progression or hemorrhage in any of the patients to date, the long-term prognosis currently is unknown. We emphasize the importance of recognizing the nonneoplastic nature of these lesions.

Index terms: Telangiectasia; Pons; Brain, magnetic resonance

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Cerebral vascular malformations frequently are classified as arteriovenous malformations, venous malformations (also called developmental venous anomalies), cavernous angiomas, or capillary telangiectasia. The imaging features of the first three types have been well described in the literature, and these lesions can be reliably distinguished on the basis of magnetic resonance (MR) (1-5). Capillary telangiectasia has been considered radiographically

occult, although the postmortem MR appearance recently has been described in a single whole-brain autopsy specimen (6).

We have encountered 12 patients in whom MR revealed a distinctive enhancing lesion of the pons that was clearly different from previous MR descriptions of vascular malformations. Though we are lacking surgical confirmation, the benign clinical course, imaging findings, and lack of progression are consistent with capillary telangiectasia, which may at least in some cases be accompanied by anomalous venous drainage.

Materials and Methods

The MR scans and clinical data of 12 patients found to have an enhancing pontine lesion with little or no T2 pro-

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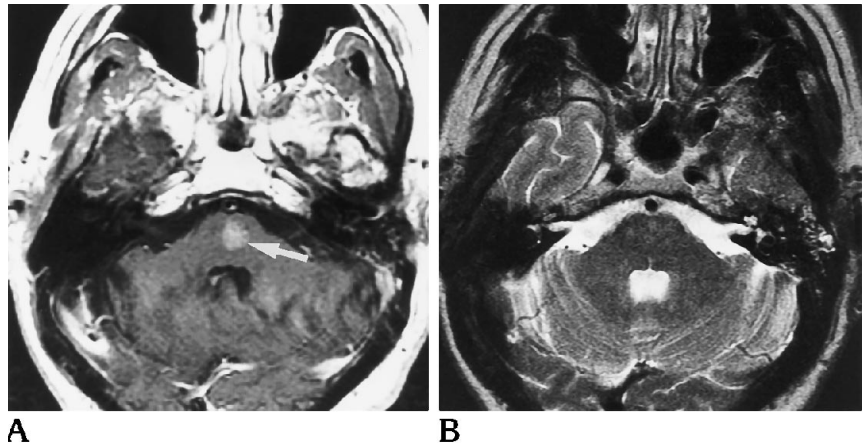
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Fig 1. Patient 4. A, Axial contrast-enhanced T1-weighted image (500/11/4) shows stippled enhancement in the pons (arrow) without mass effect.

B, Axial fast spin-echo T2-weighted image (4000/90 effective/1) is normal.



longation and no mass effect were retrospectively reviewed. Patients whose pontine lesion consisted solely of a tangle of flow voids, a linear enhancing vessel, or an enhancing lesion surrounded by a low-signal rim were excluded. The patients presented over a 3-year period from December 1990 to December 1993. Seven of the patients were scanned elsewhere and were referred to our institution for diagnosis or treatment.

All patients had T1-weighted scans with parameters of 500–710/11–29/2–4 (repetition time/echo time/excitations) performed before and after the administration of gadopentetate dimeglumine, as well as conventional (2300–2900/25–30, 80–100/0.75–1) or fast spin-echo (4000/18, 90–108/1–2) T2-weighted scans. Gradient-echo sequences with parameters of 500–600/25–30/2 and a 20° flip angle were available in seven patients and were evaluated for the presence of magnetic susceptibility effect. One patient was scanned at 1.0 T and the remaining patients were scanned at 1.5 T.

Follow-up MR scans, which were available in all patients between 3 weeks and 40 months from the initial study, also were retrospectively reviewed to detect interval change. Clinical information available at the time of the most recent MR was obtained through medical records or contact with referring physicians. None of the 12 patients underwent cerebral angiography or biopsy. One patient underwent resection of a cavernous angioma near the pontine lesion, and another underwent resection of a subfrontal meningioma.

Results

MR Features

The typical MR features of these lesions are seen in Figures 1 through 3. Three of 12 lesions were slightly hypointense on T1-weighted images, and 8 of 12 were slightly hyperintense on T2-weighted images. The remainder were isointense, including 3 of 12 lesions that could not be

detected on either the T1 or T2 precontrast sequences. Postgadolinium T1-weighted images showed a discrete focus of enhancement in the pons with irregular or brushlike borders, ranging in size from 0.3 to 1.7 cm in diameter. In at least 4 patients, the enhancement pattern was “stippled”; the lesion appeared composed of an aggregate of numerous, discrete, punctate foci of enhancement. In other cases, the enhancement pattern was homogeneous. In 8 of the patients, a small enhancing vessel could be seen extending from the lesion to the surface of the pons or fourth ventricle (Figs 2 and 3). None of the lesions exhibited mass effect.

Gradient-echo sequences were performed in seven cases, either on the initial or follow-up scans. All seven demonstrated low signal throughout the lesion, despite the absence of signal characteristic of parenchymal hemorrhage on T1- or T2-weighted images (Fig 3). Noncontrast computed tomography was performed in one patient with this finding to detect calcification and was normal.

Two of the patients were found to have additional lesions detected with MR that were thought to account for their clinical presentation (see below). No additional abnormalities were detected in the remaining 10 patients.

Follow-up scans were available in all patients, though in one case the follow-up duration was only 3 weeks. The mean follow-up duration was 11.5 months, including four patients who had follow-up scans more than 1 year after the initial presentation (13, 19, 34, and 40 months). In none of the follow-up scans has there been evidence of interval growth or hemorrhage.

Clinical Features

The patients ranged in age from 12 to 69 years at the time of presentation, with a mean age of 48 years. Seven were male and five female. Presenting symptoms included headache, vertigo, dizziness, tinnitus, hearing loss, gait abnormality, visual disturbance, memory loss, and confusion (Table). Of note, 6 of 12 presented with posterior fossa symptoms of vertigo, gait abnormalities, or hearing loss.

One patient who presented with monocular decreased vision was found to have a subfrontal meningioma, which was thought to account for his clinical symptoms. He underwent craniotomy and resection of the meningioma after his initial scan and has been free of symptoms since. His pontine lesion has remained stable on 19-month follow-up MR.

Another patient presented with transient dizziness and slurred speech and was found to have a cavernous angioma at the pontomedullary junction, inferior to the lesion in her striate pons. Postcontrast images showed an anomalous vessel that extended between the two separate lesions (Fig 4). She underwent craniotomy and resection of only the cavernous angioma, which was pathologically confirmed.

No remaining patients underwent craniotomy or biopsy, and none experienced progression of symptoms at the time of the most recent MR.

Discussion

The location, size, enhancement pattern, and lack of mass effect initially suggested that these lesions might represent capillary telangiectasia of the pons. The similarity between the pathologic features of capillary telangiectasia (Fig 5) and the lesions in our series is compelling. Capillary telangiectasia, although known to occur throughout the brain and spine, is most frequently found within the striate pons and is the most frequent incidental vascular malformation of the pons at autopsy (7, 8). Microscopically, it is composed of numerous thin-walled vessels without smooth muscle or elastic fibers that are separated by neural tissue. There is little or no gliosis (9).

The frequent presence of a visible vessel raises the possibility that these lesions represent venous malformations, rather than capillary telangiectasia. However, the confluent, nonlinear enhancement is not a feature of the classic de-

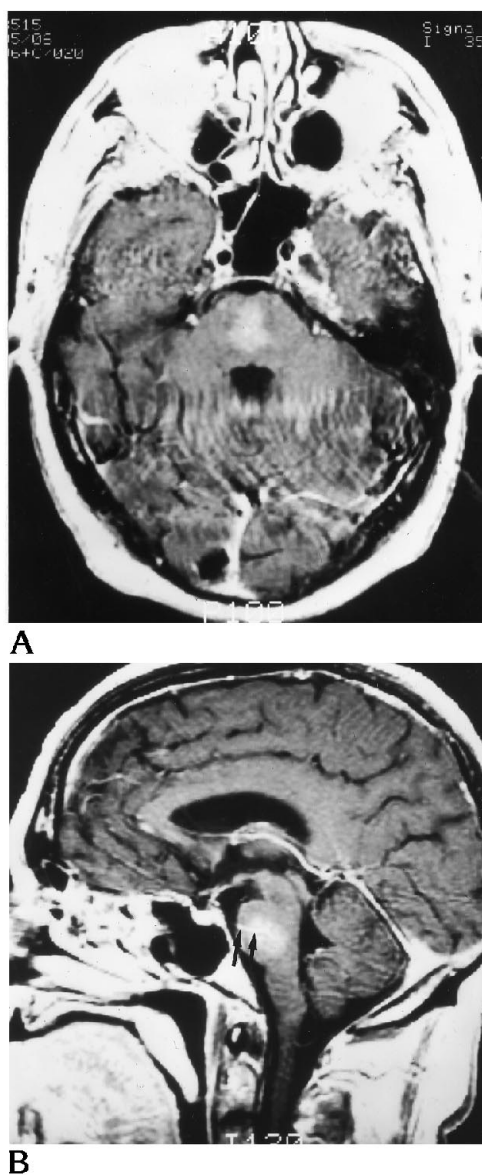


Fig 2. Patient 10. Axial (A) and sagittal (B) contrast-enhanced T1-weighted images (650/16/2) show an enhancing lesion with brushlike borders in the midpons. Sagittal image reveals an enlarged central vessel extending to the pial surface (arrows).

scription. In addition, many investigators report anomalous venous drainage of capillary telangiectasia (6, 10–14). In his 1941 description of the pathologic findings in “cerebral telangiectasis,” Blackwood noted a decrease in the total number of draining veins with dilatation of the central draining veins (14). This finding may be underestimated pathologically, because small vessels can be lost in preparation or overlooked unless the entire specimen is carefully examined (1, 15, 16). Pathologic studies of venous malformations also reveal associated capillary

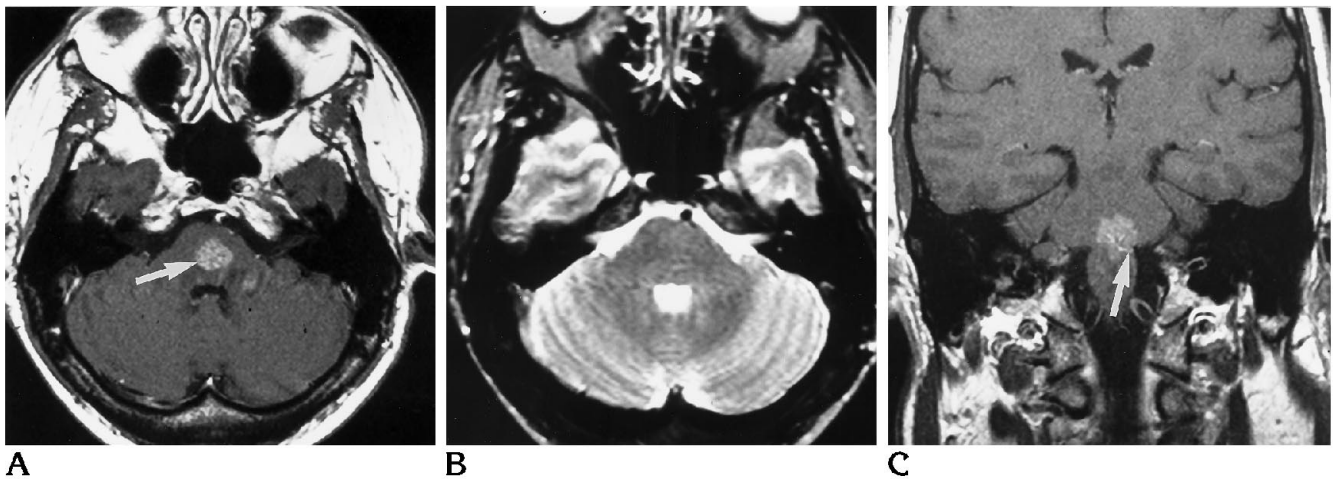


Fig 3. Patient 7. A, Axial contrast-enhanced T1-weighted image (650/16/2) shows stippled enhancement in the pons (*arrow*).

B, Axial T2-weighted image (2600/80/1) is normal.

C, Coronal contrast-enhanced T1-weighted image (650/16/2) reveals a central vessel extending laterally to the pial surface (*arrow*).

D, Coronal gradient-echo image (600/25/2; flip angle, 20°) shows marked low signal throughout the lesion (*arrow*).



abnormalities (7, 9, 17, 18). Because of the capillary proliferation associated with venous malformations, Cabanes concludes “venous angiomas seem to be capillary-venous malformations” (17). Vascular malformations traditionally have been treated as distinct entities, with diagnosis based on the predominant vessel present (17). In this regard, we consider these lesions to represent most closely the classic description of capillary telangiectasia. Whether they truly represent capillary telangiectasia or a transitional capillary-venous malformation may depend on the classification scheme.

Recent investigations have focused on the frequent occurrence of mixed or transitional forms of cerebral vascular malformations, especially the frequent association of venous malformations and cavernous angiomas (1, 6, 10, 11, 19–24). When these occur together, it is thought that the cavernous angioma accounts for the clinical symptoms, and resection of the

cavernous angioma alone is the recommended treatment (1). Focal hemorrhage associated with venous outflow obstruction has been postulated as an explanation for the development of cavernous angiomas, which are felt to be acquired lesions and which occur frequently in association with venous malformations (1). One patient with a venous malformation who later developed an adjacent cavernous angioma has previously been reported (Dillon WP, Hieshima GB, Halbach VV, Dowd CF, “A New Observation on the Association of Venous Angioma, Hemorrhage and ‘Cryptic Vascular Malformations,’” presented at the 29th Annual Meeting of the American Society of Neuroradiology, Washington, DC, June 9–14, 1991).

The development of capillary telangiectasia also might relate to venous outflow obstruction. Capillary and cavernous malformations have identical component vessel walls and are distinguished primarily by the presence or absence of

Summary of clinical and imaging features of 12 enhancing pontine lesions

Patient	Age, y	Sex	Presenting Symptoms	Location	Visible Vessel?	Follow-up
1	47	F	Right hearing loss, headache, vertigo	Right mid pons	Yes	6 mo
2	44	F	Intermittent gait abnormality	Right rostral pons	Yes	3 mo
3	53	M	Vertigo, tinnitus	Left rostral pons	Yes	40 mo
4	69	M	Abrupt onset right hearing loss	Left mid pons	No	3 wk
5	65	M	Left visual loss; subfrontal meningioma	Left rostral pons	No	19 mo
6	30	F	Recurrent vertigo, unsteadiness	Right mid pons	No	7 mo
7	43	M	Ocular migraine	Mid pons	Yes	3 mo
8	54	M	Headache	Mid pons	Yes	13 mo
9	12	M	Headache	Right mid pons	Yes	34 mo
10	57	M	Acute confusional state; ataxia on exam	Mid pons	Yes	6 mo
11	42	F	Memory loss, headache	Right caudal pons	No	6 mo
12	63	F	Transient dizziness, diplopia	Midrostral pons	Yes	1 mo

intervening neural tissue (7, 9). Capillary telangiectases and transitional lesions can be found at the periphery of cavernous angiomas in autopsy series (6). Because of these similarities, Rigamonti et al conclude that the two lesions represent "a spectrum within a single pathological entity" (6). We agree, and argue further that capillary telangiectasia and cavernous angiomas might represent the spectrum of changes that can occur as a result of venous restriction. One of the cases in our series illustrates this attractive but unproved notion. Both a pathologically confirmed cavernous angioma and separate area of presumed telangiectasia were found along the course of an anomalous vein (Fig 4). McCormick et al reached a similar conclusion in their discussion of a case of capillary telangiectasia associated with a venous malformation. They suggest that "elevated venous pressure in a venous angioma leads to ectatic dilated microvasculature, which represents an acquired telangiectasia that evolves toward a cavernous malformation" (20). The late age of onset in our series (mean age, 48 years) is consistent with the notion that telangiectasia is an acquired lesion.

The exact nature of these distinctive pontine abnormalities remains speculative in the absence of pathologic confirmation. Aside from vascular malformations, the differential diagnosis of an enhancing pontine lesion might include neoplasm, demyelinating disease, infection, infarction, or rarely, central pontine myelinolysis. The absence of mass effect or significant T2 prolongation argues against each of these entities (25–29). In particular, the distinction from neoplasm must be reinforced to avoid unneces-

sary biopsy in these cases. Two of the patients in our series, as well as numerous cases of pontine vascular malformations cited in the literature, initially presented with a presumptive diagnosis of pontine glioma (11, 30, 31). Though only short-term follow-up is available in several cases, the lack of radiographic or clinical progression in any of our patients supports our assertion that neoplasm can be excluded based on the imaging findings.

Capillary telangiectasia generally is considered a benign, incidental finding at autopsy, although there are reports of associated hemorrhage (8, 11, 20, 21). Although most spontaneous pontine hemorrhages are associated with hypertension, it is conceivable that underlying capillary abnormalities could be a predisposing factor in a proportion of cases. Long-term surveillance of these lesions, preferably with pathologic confirmation, is necessary before the true risk of hemorrhage in an incidentally discovered lesion is known.

Because of the study design and nonspecific nature of many of the presenting complaints in our series, we are unable to draw firm conclusions about the relationship of these lesions to any clinical signs or symptoms. It is interesting to note the frequency of posterior fossa symptoms in our series, including vertigo, gait difficulty, and hearing loss. One could argue, however, that the prevalence of these symptoms reflects selection bias. Patients with posterior fossa symptoms, especially hearing loss, are more likely to undergo contrast-enhanced MR with thin sections through the brain stem, which could allow detection of an otherwise incidental lesion.

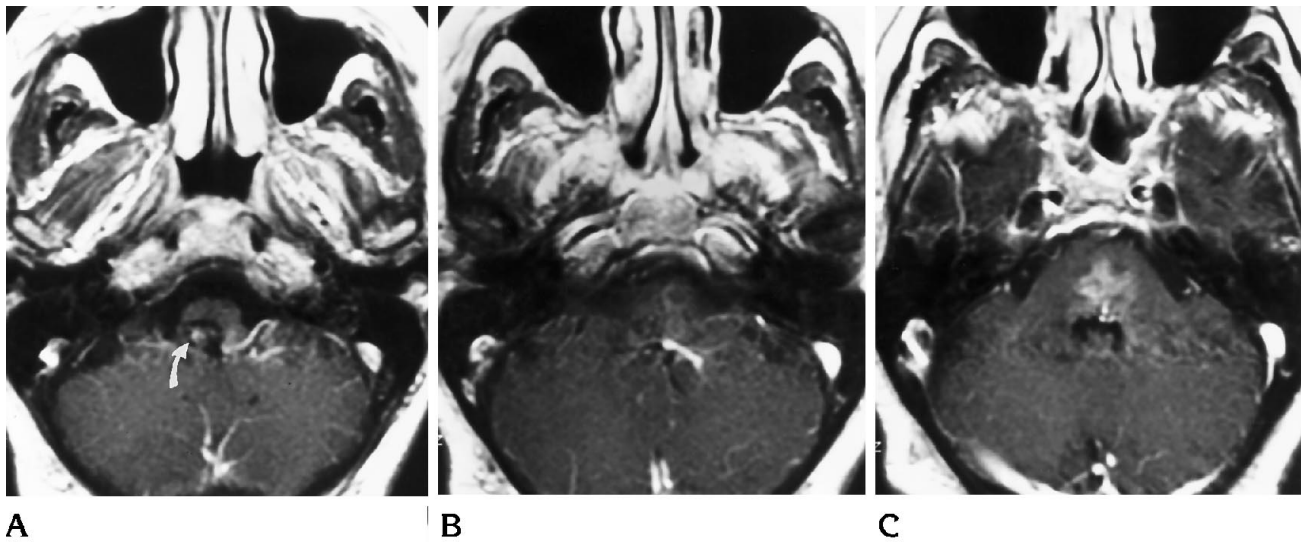


Fig 4. Patient 12. A through D, Consecutive axial contrast-enhanced T1-weighted images (500/29/2) demonstrate an abnormal vessel or vessels extending between the pontine lesion (*curved arrows*) and a surgically confirmed cavernous angioma at the pontomedullary junction (*arrow*).



Interestingly, there are reports of symptomatic pontine telangiectasia without hemorrhage (19, 32, 33). In 1928, Cushing and Bailey described a patient with unruptured pontomedullary capillary telangiectasia who demonstrated "peculiar attacks of prolonged unconsciousness" (32). In 1970, Farrell and Forno described a second case of symptomatic but non-hemorrhagic telangiectasis of the brain stem in a patient with a slowly progressive gait disturbance and speculated that "abnormal vessels caused a local deficiency of oxygen which led to the symptoms" (33).

We have chosen to follow our patients with MR at 1- to 2-year intervals or more frequently, depending on clinical status. Because of the uncertain significance of these lesions and the limited treatment options, the benefit of such

surveillance may be small, especially in patients without progressive symptoms. As for the initial detection of these lesions, most were barely visible on the precontrast T1- and T2-weighted images, and 3 of 12 could not be seen even in retrospect on either sequence. All were well seen on postcontrast T1-weighted images. None of the lesions would have been missed without contrast, however, in the seven patients who had gradient-echo sequences performed. Thus we feel that gradient-echo sequences might help increase the sensitivity of noncontrast MR in the detection of these lesions.

The finding of low signal on gradient-echo sequences also might add specificity, in that magnetic susceptibility is not a typical feature of pontine glioma or demyelinating disease. Although focal deposits of calcium or hemosiderin

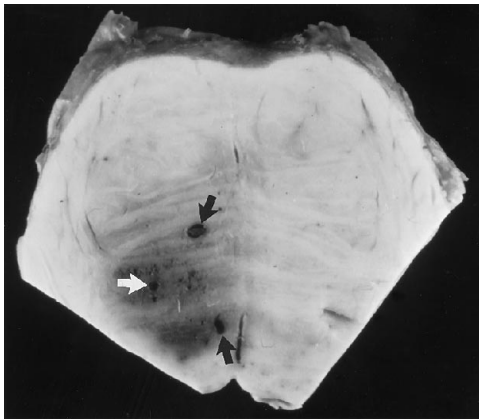


Fig 5. Gross pathologic appearance of pontine capillary telangiectasia, oriented similarly to axial MR (not from the present series). An irregular region of parenchymal discoloration without mass effect is seen in the right aspect of the pons. Note the presence of several macroscopically enlarged vascular spaces within the lesion (arrows) (case courtesy of Peter C. Burger, MD).

not detectable on T2-weighted images could potentially cause low signal on gradient-echo images, these usually are punctate or heterogeneous in appearance, unlike the homogeneous low signal in our series. Further, no calcification was detected in the one patient who underwent computed tomography. The low signal in these cases most likely reflects magnetic susceptibility from elevated intravascular deoxyhemoglobin in a region of stagnant blood flow. Increased local susceptibility can be detected as a result of increased intravascular deoxyhemoglobin and forms the basis for tissue contrast in functional MR (34–37). We have not yet performed blood flow single-photon emission computed tomography studies or dynamic perfusion MR studies in these patients but would expect to find evidence of vascular pooling.

In conclusion, the size, location, signal characteristics, enhancement pattern, and lack of progression are consistent with the pathologic description of capillary telangiectasia or a transitional capillary-venous malformation. A conservative approach is warranted when the described constellation of MR findings occurs in a patient without progressive neurologic complaints. It is unclear whether these patients are at increased risk for hemorrhage or the development of cavernous angiomas.

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