Slow-flow vascular malformation of the pons: congenital or acquired capillary telangiectasia.

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Slow-Flow Vascular Malformation of the Pons: Congenital or Acquired Capillary Telangiectasia?

We read with great interest “Slow-Flow Vascular Malformations of the Pons: Capillary Telangiectasias?” (1) in the January 1996 issue of AJNR. In this article, Barr et al postulate that telangiectasia are acquired lesions because of the late age of onset in their series (mean age, 48 years). Computed tomography (CT) of the brain and subsequent magnetic resonance (MR) imaging were recently performed at our institution on a 18-month-old infant with suspected child abuse syndrome. Plain films and T1-weighted spin-echo and T2-weighted turbo spin-echo MR images showed no abnormalities. Both contrast-enhanced CT (Fig 1A) and contrast-enhanced T1-weighted MR images (Fig 1B) showed a stippled enhancement with brushlike border in the midpons without any mass effect. MR also showed an abnormal draining vessel extending from the lesion to the right surface of the pons. These findings were consistent with a capillary telangiectasia of the pons. The neurologic examination did not reveal any posterior fossa symptoms.

Our case and case 9 of the series (age 12 years) are more consistent with a congenital than an acquired lesion, as suggested by Barr et al. We also hypothesize that, in case 9, the association between the headache presented by the patient and telangiectasia of the pons was fortuitous. In this case, telangiectasia of the pons could be considered an incidental finding. We completely agree with the authors on the issue of the follow-up of such a lesion, especially in young patients, as in our case. Because it remains unclear whether these patients are at increased risk of hemorrhage or development of cavernous angiomas (1), should we propose an MR follow-up exam or just consider this lesion an incidental finding?

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Reply

We appreciate the letter by Guibaud et al and are intrigued by their presentation of a case similar to those in our series. Our objective was to call attention to the distinctive features of these pontine lesions, and present the arguments that support a diagnosis of capillary telangiectasia based on MR imaging. We believe that recognition of the MR findings offers the promise of gaining new insight into the incidence, pathophysiology, and natural course of capillary telangiectasia.

In our discussion, we intended to emphasize the possibility that telangiectasia is an acquired lesion in patients with an underlying venous anomaly, as has been suggested by McCormick et al (1), rather than a primary developmental lesion. This would explain the frequent presence of an associated draining vein. Although the patients in our series were predominantly adults, the important issue is whether these patients were predisposed to such lesions, not at what age the lesions developed. Autopsy studies have long suggested that capillary telangiectasia is most prevalent in adults (2, 3). However, because the lesion is usually considered an incidental finding, the age of onset could be considerably earlier than the age of detection by autopsy. It is interesting that both the patient presented by Guibaud et al and our youngest patient had prominent anomalous veins. Perhaps telangiectasia occurs earlier in cases with more severe venous restriction, or possibly this is a consequence of anomalous venous drainage on a congenital basis.

We agree that the association between the presenting symptoms and the telangiectasia may be fortuitous, not just in case 9, but in all of our patients. As stated in our
discussion, we have insufficient evidence to determine whether the symptoms were caused by the telangiectasia or were coincidental. Regarding follow-up, we have chosen to study our patients, at least initially, at 1- to 2-year intervals, in the absence of new or progressive symptoms. Though likely benign, the natural course of pontine telangiectasia detected with MR remains unknown for both children and adults. Thus, the individual benefit of such surveillance cannot be determined. However, because there is no current therapy that would be directed at an asymptomatic pontine telangiectasia, it may be prudent to follow this patient clinically rather than with imaging.

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Editor’s Note.—Dr. Guibaud et al’s letter was also referred to Issam A. Awad. His comments follow.

Comment
The paper by Barr et al and the letter from Guibaud et al generate an interesting debate. Barr and colleagues present twelve cases of pontine lesion with little or no T2 prolongation on MR images, and with faint stippled enhancement on either MR or CT scan. The lesions appear limited to the pons, and are often associated with anomalous venous structure. The lesions present with nonspecific symptoms or with vague symptoms referable to the posterior fossa (although without distinct locating neurologic signs). The clinical course has remained largely benign, although one case reportedly had an associated symptomatic cavernous malformation adjacent to the lesion. The authors suggest that these might represent capillary telangiectasia. Except in the case of associated cavernous malformation, there is no histopathologic verification by either biopsy or autopsy. Guibaud et al agree with the authors, and contribute one additional case which was apparently discovered incidentally after traumatic clinical presentation. Again, no histopathologic verification is provided.

Do these lesions represent capillary telangiectasia, and what do we know about this entity? From the Greek, the term telangiectasia refers to telos, end; angeon, vessel; and ektasis, a stretching out: “dilation of small or terminal vessels.” In the neuropathologic literature, the term has been used to refer to capillary malformations, consisting of dilated capillary proliferation encountered incidentally at autopsy, and most frequently in the region of the pons. Similar capillary malformations are incidentally found at autopsy throughout the brain, without apparent clinical associations. In cases with solitary or multiple cavernous malformations, there is evidence of capillary telangiectasia adjacent to the hemorrhagic cavernous lesions and elsewhere in the same brain. These have been referred to as “satellite telangiectasia” beyond a cavernous malformation, and frequently include microhemorrhages and coalescence of the abnormal capillaries, forming “baby” hemorrhagic cavernous lesions. It has been suggested that hemorrhagic transformation of capillary telangiectasia leads to coalescence into frank cavernous malformations. The latter progress by further hemorrhagic angiogenic proliferation. Similar telangiectasia has been observed in regions of the brain with anomalous venous drainage (venous developmental anomaly or venous angioma), and it has been suggested that they can evolve into frank cavernous malformations in association with the venous anomalies.

Elsewhere in the body, the term telangiectasia has been used to refer to a wide variety of vascular malformations, including the typical true arteriovenous malformation of Osler-Weber-Rendu (hereditary hemorrhagic telangiectasia). This disease is in fact associated with multiple brain arteriovenous malformations, and not multiple telangiectasia as would be suggested by its nomenclature. Nor are the systemic lesions in this disease true telangiectasias, but rather arteriovenous malformations as well. Because of this confusion in semiology, I favor abandoning the term capillary telangiectasia, in favor of the term capillary malformation. I and others have proposed a simple but uniform classification of terminology for the various brain vascular malformations into capillary, venous, arteriovenous, cavernous and mixed lesions. Frequently, there would be mixed cavernous-capillary lesions as noted above, and also mixed venous-capillary and venous-cavernous malformations in the same host. This terminology is simple, and incorporates all types of lesions reported to date within their respective clinical-radiologic-pathologic contexts. The lesions reported by Barr et al and Guibaud et al would therefore be capillary malformations, associated in a few cases with venous malformations, and in one case with a cavernous malformation.

While I essentially agree with the likely interpretation proposed by the authors, we must remain cautious about diagnosing this entity in the absence of histologic verification. It is clear that a similar radiographic appearance might be mimicked by hamartomatous or benign intrinsic neoplastic entities. These also would be consistent with the nonspecific clinical presentations and benign clinical course. Rarely, such lesions could progress radiographi-
cally and/or clinically, warranting a more aggressive clinical stance, including biopsy. Even a capillary malformation, with or without associated venous malformation, might progress through microhemorrhages or other triggering factors into a symptomatic proliferative cavernous malformation that might require clinical attention.

I would therefore agree with the rhetorical question posed by Guibaud and colleagues, proposing MR follow-up of these cases. I would emphasize that the described MR and clinical features are neither fully sensitive nor specific for capillary malformation. For the many reasons mentioned previously, the radiologist cannot afford to place a benign clinical label, or even a specific histopathologic nomenclature, on an entity without clear verification, especially in view of potential clinical progression. When such a lesion is encountered, a cautious radiologic interpretation might suggest that it is “consistent with capillary vascular malformation,” but also propose differential diagnostic considerations, and urge close serial follow-up with MR imaging and clinical correlation.

Vertebral Artery Loop Causing Cervical Compression

Cervicobrachial neuralgia is most often related to compression of a nerve root. Such compression can be caused by various pathologic processes, including cervical spondylosis, degenerative disk disease, or, less commonly, a tumor. When a patient with cervicobrachial neuralgia is found to have enlargement of the neuroforamen, the cause is often a tumor (usually a neurinoma or meningioma). Enlargement of the foramen by a vessel compressing the nerve root is a rare finding, with few reports in the literature (1–4). We report here a case of cervical nerve root compression caused by a loop of the vertebral artery.

A 74-year-old woman was admitted to our hospital with a 4-week history of right cervicobrachial neuralgia. Neurologic examination revealed complete palsy of the right deltoid muscle and slight atrophy and palsy of the right biceps muscle without sensory deficits, leading to a diagnosis of right cervical (C-5) nerve root compression. Radiographic imaging showed a slight enlargement of the right C4-5 intervertebral foramen (Fig 2A). Myelography revealed a failure of filling the C-5 nerve root sheath. Postmyelogram CT showed a mass filling the enlarged right C4-5 foramen. On enhanced CT (section, 4 mm), the intraforaminal mass enhanced strongly, indicating a high vascularization of the space-occupying lesion. Digital sub-
traction angiography showed no tumor stain or tumor vessels; however, a loop of the right vertebral artery was identified at the C-5 level (Fig 2B), leading to the diagnosis that the vertebral artery was compressing the nerve root. This was confirmed on enhanced spiral CT (Fig 2C), MR imaging, and MR angiography (Figs 2D and E). At surgery, the vertebral artery canal was unroofed at the level of the vertebral bodies of C-4 and C-5 followed by ventral displacement of the loop. After surgery, the patient experienced immediate and lasting pain relief for the follow-up period of 2 years. The palsy of the deltoid muscle improved with rehabilitation.

Cervical nerve root compression by a loop of the vertebral artery is rare. The typical clinical presentation is radicular pain and palsy. The compressed nerve roots usually range from C-3 to C-6 and commonly occur on the left side (1–6), an observation consistent with the dominance of the left vertebral artery.

High-resolution enhanced CT can clearly show a vertebral artery loop (Fig 2C). The optimal visibility of the anatomy of the region, especially the exact course of the loop, is best obtained from MR, MR angiography, and CT angiography. These methods provide the information necessary for planning surgery. Microvascular decompression of the nerve root was successful in relieving the symptoms in the case we report and in others (2), thereby suggesting that this is the appropriate therapy for this condition.

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