Capillary telangiectasias: clinical, radiographic, and histopathological features

Clinical article

CHRISTINA M. SAYAMA, M.D.,1 ANNE G. OSBORN, M.D.,2 STEVEN S. CHIN, M.D., PH.D.,3 AND WILLIAM T. COULDWELL, M.D., PH.D.1

Departments of 1Neurosurgery, 2Radiology, and 3Pathology, University of Utah, Salt Lake City, Utah

Object. Brain capillary telangiectasias (BCTs) are small, clinically benign, angiographically occult lesions that are usually incidental findings. Large capillary telangiectasias have not been reported previously as most BCTs are very small. Symptomatic BCTs are also rare, with few reports in the literature. The authors review the clinical manifestations, imaging, and histopathological characteristics of BCTs to further elucidate the diagnostic and clinical features of these vascular malformations.

Methods. The authors completed a retrospective radiological review of all cases of BCTs in the neuroradiology database at the University of Utah involving patients treated between January 1993 and December 2007. The MR imaging scans were reviewed, and the BCT was measured in 2 dimensions. They arbitrarily chose > 1 cm to define a large BCT as a majority of these lesions were smaller than that. The medical chart and the electronic database were used to gather each patient’s clinical information.

Results. One hundred thirty patients were identified in the archived neuroradiology database of capillary telangiectasias. Cases involving 105 patients with definite capillary telangiectasias were reviewed, and from these, 7 patients were identified to have a large capillary telangiectasia measuring > 1 cm. Upon further review, 2 of these patients were identified as having symptoms likely related to their capillary telangiectasia. These 2 cases are reported in the article. No patients with smaller BCTs were found to have symptoms related to their lesion.

Conclusions. Brain capillary telangiectasias are small vascular malformations that rarely cause symptoms. They are often overlooked on imaging because of their clinically benign nature; however, they have been misdiagnosed as glial tumors in the past. Specific MR imaging sequences (T1-weighted postcontrast and gradient refocused echo) are valuable in aiding diagnosis, as histopathological diagnosis is often not possible. These cases highlight that BCTs can cause symptoms, a finding that may actually be related to the size of the lesion (28.6% of large BCTs in this series were symptomatic, whereas none of the small ones were). (DOI: 10.3171/2009.9.JNS09282)

Key Words • capillary telangiectasia • seizure • symptomatic • vascular malformation

Abbreviations used in this paper: BCT = brain capillary telangiectasia; PACS = picture archiving and communication system.
dimension, considering that the vast majority of these lesions are very small. Although BCTs themselves are not rare (4–12% of vascular malformations), it is rare to encounter symptomatic capillary telangiectasias. Remarkably few reports have described symptoms due to BCTs, such as seizures, blurred vision, cranial nerve dysfunction, and progressive spastic paraparesis.\cite{3,6,8}

Brain capillary telangiectasias cannot be identified easily on angiography or on most imaging modalities. Their diagnosis is often made incidentally at necropsy. Premortem diagnosis using imaging and resection with histopathological diagnosis is essentially nonexistent. Thus, a more comprehensive understanding of the clinical, imaging, and pathological characteristics of these lesions is warranted to elucidate their diagnostic features. We reviewed our radiological database of BCTs to determine the percentage of large capillary telangiectasias and furthermore to define the incidence of symptomatic BCTs within this study population.

**Methods**

We undertook a retrospective radiological review of the neuroradiology database at the University of Utah to identify all patients noted to have radiological evidence of capillary telangiectasias between January 1993 and December 2007. The study was approved by the University of Utah Institutional Review Board.

Patient MR imaging scans were reviewed, and the largest diameter of the capillary telangiectasia was measured in 2 dimensions using our digitized PACS. When the MR imaging study was not available on PACS, measurements were made using a ruler and the sidebar available on the MR image itself. The lesions were measured at high magnification when available on the PACS (there were only 3 cases in which the images were no longer available on the PACS). We referred to documented MR imaging reports when it was difficult to identify the lesion. New measurements of each lesion were obtained, even if the size of the capillary telangiectasia was noted in the report. The clinical information was reviewed, and we further researched the presentation and symptoms of those patients identified as having large capillary telangiectasias.

**Results**

In our review of the neuroradiology database from the period between January 1993 and December 2007, we identified 130 patients with radiological evidence of a capillary telangiectasia (mean age 47.5 years, range 18–93 years; 75 women, 55 men). In 25 of these cases, review of the MR images demonstrated that another vascular malformation was more likely or the imaging studies did not include the sequences required to truly identify the lesion; thus, a total of 105 patients with radiologically definite capillary telangiectasia were identified (mean age 47.4 years; 63 women, 42 men).

Of these 105 patients, 7 (6.7%) were found to have large capillary telangiectasias measuring > 1 cm. The mean age of this subgroup of patients was 37 years (range 19–58 years); 3 patients were women, 4 were men. These large telangiectasias were located in the temporal lobe (uncus and insular cortex), basal ganglia, pons, midbrain, and parafalcine frontal lobe (Table 1).

Of the 7 patients with large capillary telangiectasias, 2 patients (28.6%) were identified as having symptoms likely related to their capillary malformations. The remaining 5 patients were asymptomatic. There were no patients with smaller capillary telangiectasias noted to have symptoms secondary to their lesion.

**Illustrative Cases**

**Case 6**

This 19-year-old woman presented to another facil-

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>BCT Size &amp; Location</th>
<th>Reason Imaged</th>
<th>Symptoms</th>
<th>Histopathological Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>19, F</td>
<td>1.5 cm × 2 cm, lt insular cortex</td>
<td>unknown</td>
<td>asymptomatic</td>
<td>NA</td>
</tr>
<tr>
<td>2</td>
<td>33, M</td>
<td>9.5 mm × 1.1 cm, pons</td>
<td>numbness in lt arm, Horner syndrome</td>
<td>asymptomatic</td>
<td>NA</td>
</tr>
<tr>
<td>3</td>
<td>49, M</td>
<td>1.5 cm × 4 mm, rt basal ganglia</td>
<td>cerebellar astrocytoma</td>
<td>asymptomatic</td>
<td>NA</td>
</tr>
<tr>
<td>4</td>
<td>58, F</td>
<td>1.3 cm × 1.2 cm, rt frontal parafalcine</td>
<td>MVA w/ occipital headache &amp; neck pain</td>
<td>asymptomatic</td>
<td>NA</td>
</tr>
<tr>
<td>5</td>
<td>48, M</td>
<td>1.2 cm × 8 mm, lt midbrain</td>
<td>progressive frontal headache</td>
<td>asymptomatic</td>
<td>NA</td>
</tr>
<tr>
<td>6</td>
<td>19, F</td>
<td>1.2 cm × 1.5 cm, rt uncus</td>
<td>seizures</td>
<td>seizures</td>
<td>cortical gray matter w/ increased numbers of dilated capillary-type blood vessels</td>
</tr>
<tr>
<td>7</td>
<td>33, M</td>
<td>1.4 cm × 1.2 cm, rt basal ganglia</td>
<td>seizures, headache</td>
<td>seizures, headache</td>
<td>NA</td>
</tr>
</tbody>
</table>

* MVA = motor vehicle accident; NA = not available.
Capillary telangiectasia resulting in seizures

ity in January 2005 with partial complex seizures. She had been having “déjà vu”–like events for several years but could not remember a specific onset. At the time of presentation, she was having approximately 2 events per week. She did not have any overt motor manifestations with her seizures. The results of her neurological examination were entirely unremarkable. Treatment with oral lamotrigine was initiated; although it did reduce the frequency of her seizures, she was inconsistent in taking the medication.

Magnetic resonance imaging studies obtained in June 2005 demonstrated an enhancing lesion in the right mesiotemporal region. The lesion measured 1.2 × 1.5 cm and was thought to be consistent with a glial tumor (Fig. 1). The patient underwent seizure monitoring, which localized her seizures to the right side of the brain. She also underwent Wada testing, which demonstrated language dominance on the left side of the brain and memory on both sides of the brain, with memory performance using the left hemisphere superior to that using the right hemisphere. The patient presented to the neurosurgery clinic in August 2005 for management of the suspected neoplastic lesion and associated seizures.

The patient subsequently underwent resection of the lesion with the dual goal of helping with her seizure disorder as well as providing a specific diagnosis of the lesion. The surgeon encountered a dark bluish area of tissue in the anterior temporal lobe that appeared abnormal. A 0.6-cm biopsy specimen was sent for final pathological analysis. A subpial resection of the lesion was completed with gross-total resection. Selective lesionectomy with resection of the amygdala and the amygdala-hippocampal junction was performed at the time of surgery. The histopathological evaluation of the lesion showed cortical gray matter with increased numbers of dilated capillary-type blood vessels, consistent with capillary telangiectasias (Fig. 2).

A postoperative CT scan in November 2005 showed no evidence of complication with normal postsurgical changes in the right frontotemporal region. The patient continued on oral lamotrigine therapy, and her mother did not report any further events or staring spells.

At her follow-up examination in neurology clinic 2 years later, the patient reported that no clear seizure activity had been witnessed while she had been off all medication. Thereafter, she was lost to follow-up.

Case 7

This 33-year-old man presented to our neurosurgery clinic in September 1993 with a 6- to 8-month history of brief trance-like spells and headaches. Previous CT and MR imaging scans from another hospital showed an area of contrast enhancement in the right anterior lateral hypothalamus that was suggestive of a possible glial tumor.

The patient and his wife reported that he had experienced 2 different kinds of spells. The first type of spell was more severe and had only occurred 5 or 6 times in the previous 6 months. These spells typically lasted 30 minutes; during this time, the patient felt an aura and impending sense of doom followed by a trance-like state in
which the patient wandered around staring and appeared confused. The second, less severe type of spell occurred several times a week and consisted of a few brief seconds of blank staring or a mind lapse. The patient also complained of a constant daily headache, usually in the right temporal area. The results of his neurological examination was unremarkable.

A baseline MR imaging study performed at our institution in September 1993 showed a patchy, poorly delineated enhancing area in the inferomedial basal ganglia and inferior lenticular limb of the internal capsule on the right side seen only on enhanced images. This lesion was found to measure 1.4 × 1.2 cm, and it was unclear whether it represented a vascular malformation such as a capillary telangiectasia or a glial tumor because there was no abnormal T2 signal within the region (Fig. 3).

Treatment with carbamazepine was initiated for control of what were thought to be partial complex seizures. Five months later, in February 1994, the patient was seen again in clinic and was noted to have had only 3 spells lasting 10–15 seconds each since his previous examination. A follow-up MR imaging study showed no change in the lesion in the right inferomedial basal ganglia, which again was seen only after enhancement with gadolinium. The lack of change considerably decreased the likelihood that this lesion was a glial tumor. Because the patient reported chronic fatigue and muscle pain in association with the carbamazepine therapy, it was replaced with phenytoin therapy with follow-up scheduled for 1 year.

One year later, the patient was again seen in the neurosurgery clinic and was clinically doing well on a combination of valproic acid and imipramine. (Phenytoin therapy had been discontinued due to adverse side effects.) He reported no more spells of confusion. Follow-up MR images were again obtained, and there appeared to be no change in the size of the lesion in comparison with pre-
Capillary telangiectasia resulting in seizures

Previous studies. Magnetic resonance angiography revealed a lesion involving the right infralenticular region with a large draining vein that emptied into the right basal vein of Rosenthal. When a repeat MR imaging study 3 years after the initial presentation again showed no change in the area of enhancement in the infralenticular portion of the anterior limb of the internal capsule measuring 1.4 × 1.2 cm, the patient was informed that he likely had a capillary telangiectasia associated with a draining vein. He was subsequently lost to follow-up.

Discussion

Capillary telangiectasias are difficult to diagnose clinically for several reasons. Patients rarely present with symptoms caused by the lesions, and the lesions are hard to diagnose without the proper imaging studies. Finally, histopathological specimens are rarely used to diagnose BCTs in living patients. Our cases demonstrate the difficulty of accurately diagnosing BCTs. The patient in our first illustrative case was thought to have a low-grade glial tumor on MR imaging, but on histopathological analysis the resected lesion was discovered to be a capillary telangiectasia. The patient in our second illustrative case was also referred to us with seizures and a suspected glial tumor based on MR imaging studies from another facility. He was treated medically for his seizures, and further imaging supported the diagnosis of capillary telangiectasia. His seizures were adequately controlled with antiepileptic medication until he was lost to follow-up.

Because of the difficulty of accurately diagnosing capillary telangiectasias, we have identified clinical manifestations (albeit rare), diagnostic radiological features, and histopathological characteristics of BCTs to help physicians more easily identify these lesions and guide therapeutic decisions.

Clinical Manifestations

Capillary telangiectasias are commonly located in the pons, temporal lobe, medulla, or caudate and typically have a benign clinical course. Although these lesions are usually small, we found 7 (6.7%) of 105 patients with BCTs had lesions that qualified as large capillary telangiectasias. It is very unusual for patients to experience symptoms caused by a BCT; however, when a lesion is symptomatic, it can manifest as seizures, cranial nerve dysfunction, confusion, dizziness, visual changes, vertigo, tinnitus, or progressive spastic paraparesis. Previous studies have indicated that symptoms of BCTs may be secondary to hemorrhage into or direct compression onto the adjacent normal brain tissue. Symptoms may also be due to hypoperfusion and hypoxic injury since dilated capillaries may lead to blood stasis. Two of our patients presented with seizures, which were thought likely to be due to their capillary telangiectasias. Of the patients with smaller capillary telangiectasias, many were referred for follow-up MR imaging for tumors, multiple sclerosis, vascular malformations, Parkinson disease, strokes, headaches, or seizures. In those patients referred for follow-up imaging for seizures, the seizures themselves were not noted to be in epileptogenic areas in the brain and did not correlate with location of the capillary telangiectasia (there were 4: 2 in the pons, 1 in the right cerebellar peduncle, and 1 in the right occipital lobe).

Although a majority of capillary telangiectasias will probably never cause symptoms within an individual’s lifetime, our cases show that symptomatic capillary telangiectasias are possible and should be considered in the differential diagnosis. The series also demonstrates that large capillary telangiectasias are more likely to be symptomatic, as none of the smaller ones were thought to be symptomatic. In addition, the cases also demonstrate the ease of mistaking a capillary telangiectasia, which is essentially benign, with the more concerning diagnosis of a glial brain tumor. The early and proper diagnosis of a capillary telangiectasia could put a family at ease, as these lesions have no neoplastic potential.

Radiological Features

Brain capillary telangiectasias are a type of “angio graphically occult vascular malformation” that is not visualized by serial cerebral angiography and also does not appear on most other imaging modalities. It is thought that BCTs cannot be seen on angiography because 1) they have sluggish flow; 2) the lesion may have hemorrhaged (the bleeding or clot obliterates the lesion); 3) the abnormal vessels are very small in size; or 4) they may only be visualized on very late angiographic sequences.

These lesions are not seen on CT and have been noted to be otherwise undetectable on conventional MR imaging, including T1- and T2-weighted MR imaging, fluid-attenuated inversion-recovery (FLAIR) imaging, or diffusion-weighted MR imaging. With gadolinium-enhanced T1-weighted MR imaging, however, BCTs are usually visible as small, faintly enhancing lesions with a stippled or brush-like appearance. An enlarged vessel, thought to most likely represent a draining vein, is seen in approximately two-thirds of capillary telangiectasias.

As shown in Figs. 1 and 3, the lesion appears to be a “dot in the spot” when visualized on postcontrast T1-weighted MR images; BCTs also demonstrate marked signal intensity loss on gradient-echo images. Therefore, both postgadolinium T1-weighted imaging and gradient-echo imaging are valuable in making the diagnosis of capillary telangiectasia. It is important to obtain thin-section (3-mm with a 0.5-mm gap or contiguous 3-mm sections) gradient-echo sequence images to avoid missing these small lesions, which vary in size from a few millimeters to as large as 2 cm. Not all of our patients had gradient-echo sequencing available. It was noted that 10% (11/105) of the patients had corresponding hypointensity on diffusion-weighted images, as seen in Fig. 3E.

Histopathological Characteristics

Brain capillary telangiectasias are composed of multiple dilated capillaries surrounded by normal brain parenchymal tissue. The capillaries appear microscopically as a single layer of endothelial cells that form ectatic vessels. Calcification, hemorrhage, gliosis, and hemosiderin-laden macrophages are usually not typical features that occur with BCTs but can be found in other types of vas-
cular malformations such as arteriovenous or cavernous malformations.

The patient in our first illustrative case had a lesion removed from her right mesiotemporal region that showed cortical gray matter with increased numbers of dilated capillary-type blood vessels. These dilated vessels were found among normal brain tissue without evidence of gliosis, hemorrhage, calcification, or hemosiderin-laden macrophages. The histopathological analysis confirmed the finding of capillary telangiectasia despite the preoperative possible diagnosis of a glial tumor based on previous MR imaging. On retrospective review of more diagnostic sequences, however, it is clear that the MR images were indeed consistent with a capillary telangiectasia.

Although histopathological analysis is indeed very helpful in the diagnosis of capillary telangiectasia, it is often not performed because these lesions are characteristically benign. It is beneficial to identify these lesions early using imaging characteristics to determine the best therapeutic decisions and avoid unnecessary surgical procedures.

Conclusions

Brain capillary telangiectasias are fairly common vascular malformations that rarely cause symptoms. Despite their benign nature, they are often misdiagnosed as glial tumors or other more serious disease processes. Most BCTs are small and can be monitored over time with serial imaging. Because they do not usually cause clinical symptoms, these lesions are often overlooked when a patient presents with symptoms such as a seizure disorder. Our cases demonstrate, however, that symptoms secondary to BCTs do occur and may be related to the size of the lesion. We discovered that 28.6% of the large capillary telangiectasias in our case series were symptomatic, whereas none of the small capillary telangiectasias were symptomatic.

The gold standard for diagnosis of these lesions, histopathological analysis after biopsy or resection, is often not possible. We highlight the importance of recognizing that BCTs can cause symptoms and that specific MR imaging sequences are valuable in their diagnosis. The proper diagnosis of a BCT can help define therapeutic decision-making (medical vs surgical management) and can also assuage anxiety for the patient and the patient’s family.

Disclosure

The authors do not report any conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Acknowledgment

The authors thank Kristin Kraus, M.Sc., for editorial assistance in preparing this paper.

References


Manuscript submitted February 20, 2009. Accepted September 10, 2009. Please include this information when citing this paper: published online October 9, 2009; DOI: 10.3171/2009.9.JNS09282.

Address correspondence to: William T. Couldwell, M.D., Ph.D., Department of Neurosurgery, University of Utah, 175 North Medical Drive East, Salt Lake City, Utah 84132, email: neuropub@hsc.utah.edu.