Brain capillary telangiectasia (BCT) is a rare vascular malformation with a prevalence of 0.4%–0.7% based on autopsy and MRI diagnosis. Most BCTs are small in size and clinically silent with a predilection of pons and basal ganglia. Clinical data of large and symptomatic BCTs are rare. Only 10 symptomatic cases with 3 diffuse BCTs have been reported in the English-language literature of more than 200 BCTs. Large BCTs have a much higher risk of causing uncontrolled bleeding and severe neurological defects, and they can be fatal if left untreated. Therefore, large BCTs should be managed with special caution. Because of the lack of reports, diagnosis of large BCTs has been difficult. Strategies of management are undefined for large or giant BCTs.

The current study presents 5 cases of giant and large BCTs. To the authors’ knowledge, this is the largest series of this disease ever reported. Radiological findings, histopathological characteristics, clinical presentations, and surgical management were analyzed in 5 symptomatic, unusually large BCTs (mean diameter 5.06 cm, range 1.8–8 cm).

Four patients presented with focal or generalized seizures, and 1 patient presented with transient vision loss attributed to the lesions. Gross-total resection of the lesion was achieved in all patients. After surgery, the 4 patients with seizures were symptom free for follow-up periods varying from more than 1 to 5 years with no additional neurological deficits. The unique location, radiological characteristics, and clinical course suggest that giant BCTs could be a different entity from small BCTs. Surgery might be a good option for treatment of patients with intractable neurological symptoms, especially in those with surgically accessible locations. Complete removal would be anticipated to provide relief of the symptoms without causing new neurological deficits.

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**KEY WORDS** capillary telangiectasia; vascular malformation; resection; vascular disorders

**ABBREVIATIONS** BCT = brain capillary telangiectasia; CM = cavernous malformation; DSA = digital subtraction angiography; ECoG = electrocorticography; EEG = electroencephalography.


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Methods

This study was approved by the local ethics board. Informed consent was obtained from all patients. All 5 cases were histologically confirmed as BCTs according to WHO classification. The review of these specimens was performed by 2 neuropathologists from Peking Union Medical College Hospital. There were 4 males and 1 female with a median age of 18 years (range 3–35 years) who presented with symptoms including seizures (4 cases) and recurrent neurological event (1 case). The time delay between the first symptoms and diagnosis was 1 month to 3 years; 1.5-T or 3-T T1-weighted and T2-weighted images were obtained in all cases. The maximum diameters of the lesions varied from 1.8 to 8 cm (Table 1). T1-weighted MRI after gadolinium contrast administration was performed in 2 cases (Cases 1 and 4). Digital subtraction angiography (DSA) was performed in 3 cases (Cases 1, 3, and 5), and MR angiography was performed in 1 case (Case 2). Preoperative interictal electroencephalography (EEG) and intraoperative electrocorticography (ECoG) were performed to localize and monitor the epileptic foci before and during the surgeries. The mean follow-up time was 23 ± 15 months (± SD).

Results

Case 1

An 8-year-old boy presented with 3 years of recurrent status epilepticus with binocular right-sided lateral gaze accompanied by nausea and vomiting. Management of seizures with oral valproate sodium reduced seizure frequency. However, side effects of valproate sodium involving gastrointestinal distress progressed, and patient compliance was poor. The patient still experienced 1 seizure episode each week while receiving medication. Left frontal focal sharp waves were found on EEG, indicating potential seizure focus, which was consistent with the location of the lesion. Brain MRI showed a cone-shaped lesion measuring 6.5 × 5.0 cm in the left frontal lobe. The lesion was slightly hypointense on T1-weighted MRI and hyperintense on T2-weighted MRI. After contrast administration, the lesion demonstrated linear enhancement on MR angiography. Brain MR angiography study did not show any vascular malformation. The MR angiography study did not show any vascular malformation. EEG revealed a slow wave at the right temporooccipital region. Within the lesion, there were small hypointense and hyperintense nodules indicating hemorrhagic foci (Fig. 2). The MR angiography study did not show any vascular malformation. EEG revealed a slow wave at the right temporooccipital region, although no typical epileptic wave was detected. Resection was scheduled for seizure control.

The lesion was totally removed in a piecemeal fashion. The amount of blood loss was 200 ml. Histopathological examination showed cortical neural tissue with significantly increased numbers of dilated capillary-type blood vessels, consistent with capillary telangiectasias (Fig. 2). The patient recovered well from the surgery and was discharged on antiepileptic drugs. She stopped taking medications 5 months before the last follow-up (12 months after surgery) and did not experience any further seizure episodes.

Case 2

A 3-year-old girl presented with generalized tonic-clonic seizures precipitated by diffuse abdominal pain, nausea, and vomiting. The initial seizure onset occurred 1 month prior to presentation. The patient experienced 4 episodes of seizure within that month, which could not be controlled by antiepileptic agents. Preoperative brain T1- and T2-weighted images showed a cone-shaped lesion with a brushlike vascular structure that measured 8.0 × 3.0 cm in the right temporooccipital region. Within the lesion, there were small hypointense and hyperintense nodules indicating hemorrhagic foci (Fig. 2). The MR angiography study did not show any vascular malformation. EEG revealed a slow wave at the right temporooccipital region, although no typical epileptic wave was detected. Resection was scheduled for seizure control. Resection was scheduled for seizure control.

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Case 3

A 17-year-old man presented with right-sided complex partial seizures. During the seizure episode, the patient experienced right-sided facial spasm, myoclonic movement of the right extremities with his head turning to the right, and bilateral upward eye deviation. Antiepileptic medications including valproic acid, lamotrigine, and carbamazepine were administered each for 4–6 months as initial

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Size (cm)</th>
<th>Location</th>
<th>Symptoms</th>
<th>DSA</th>
<th>Draining Vein</th>
<th>Pathology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8, M</td>
<td>6.5 × 5.0</td>
<td>Lt frontal lobe</td>
<td>Seizure</td>
<td>No positive finding</td>
<td>Yes</td>
<td>Mixed BCT &amp; CM</td>
</tr>
<tr>
<td>2</td>
<td>3, F</td>
<td>8.0 × 3.0</td>
<td>Rt temporooccipital region</td>
<td>Seizure</td>
<td>Not performed</td>
<td>Yes</td>
<td>BCT</td>
</tr>
<tr>
<td>3</td>
<td>17, M</td>
<td>5.0 × 4.0</td>
<td>Lt frontal lobe</td>
<td>Seizure</td>
<td>No positive finding</td>
<td>Yes</td>
<td>Mixed BCT &amp; CM</td>
</tr>
<tr>
<td>4</td>
<td>35, M</td>
<td>1.8 × 1.3</td>
<td>Lt temporal lobe</td>
<td>Lt eye transient vision loss</td>
<td>Not performed</td>
<td>No</td>
<td>Intermittent form of BCT &amp; CM</td>
</tr>
<tr>
<td>5</td>
<td>19, M</td>
<td>4.0 × 3.0</td>
<td>Rt temporal lobe</td>
<td>Seizure</td>
<td>No positive finding</td>
<td>Yes</td>
<td>Mixed BCT &amp; CM</td>
</tr>
</tbody>
</table>
therapy. Although the frequency of the seizure episodes was reduced, none of the drugs was able to completely eliminate the seizure. The left frontal lesion measured 5.0 × 4.0 cm (Fig. 3) and was diagnosed as being either a vascular malformation or low-grade glioma. EEG detected bilateral frontoparietal epileptic waves with a left-sided dominance, which is consistent with the location of the vascular lesion. Surgery was scheduled for resection of a suspected vascular malformation/neoplastic lesion associated with intractable seizure.

During the operation, the preoperative visible ECoG spike wave at the left frontal lobe disappeared after resection of the mass. The histopathological evaluation of the lesion was consistent with capillary telangiectasias with coexisting venous malformation. The patient recovered well and has been seizure free since the most recent follow-up (2 years after surgery).

**Case 4**

A 35-year-old woman complaining of transient visual loss accompanied by dizziness for 3 weeks presented to our clinic. Ophthalmological examination findings were normal for both eyes. Brain MRI showed a lesion in the left mesial temporal lobe adjacent to the hippocampal head. The 1.8 × 1.3–cm lesion was mildly hypointense on T1-weighted images, isointense on T2-weighted images, and slightly enhanced after contrast administration (Fig. 4). Low-grade glioma was suggested as the initial diagnosis, and surgical removal was scheduled.

A gray-reddish mass with enriched blood supply located adjacent to the head of the hippocampus was completely removed in the surgery. The histopathological evaluation of the lesion demonstrated clusters of dilated capillaries (Fig. 4). This patient underwent follow-up for 5 years and 6 months; her vision improved gradually, and no additional neurological disability was observed.

**Case 5**

A 19-year-old man presented with recurrent generalized seizures induced by exhaustion. Preoperative brain MRI showed a vascular lesion measuring 4.0 × 3.0 cm in the right temporal lobe; findings on DSA appeared normal (Fig. 5). Preoperative EEG demonstrated spike form discharge at the location of the lesion. The seizure was intractable with long-term anticonvulsant medication. A lesionectomy was performed. Postoperative imaging studies confirmed a gross-total resection. The histopathological examination of the lesion was consistent with capillary telangiectasias with a coexisting venous malformation (Fig. 5). The patient recovered well and at the most recent follow-up (1 year and 6 months after surgery) had not experienced any seizures.
symptomatic large or giant capillary telangiectasias

Fig. 2. Case 2. a and b: Axial T1-weighted (A) and T2-weighted (B) images showing heterogeneous vascular lesions of the right temporocipital region. Isointense to hypointense brush-like patterns of the lesion indicate malformation of the capillaries. Hypointense and hyperintense nodules are suggestive of the presence of small hemorrhagic foci. c: Light microscopy of a surgically acquired sample demonstrating the dilated endothelial walls of the capillaries with intervening neuronal tissue. H & E, original magnification ×100; bar = 100 µm. Postoperative T1-weighted (D) and T2-weighted (E) images showing no residual tumor. Figure is available in color online only.

Fig. 3. Case 3. a and b: Sagittal T1-weighted (A) and axial T2-weighted (B) images showing vascular structure with no mass effect in the left frontal parietal lobe. The lesion is isointense on the T1-weighted image and heterogeneous on the T2-weighted image. Surrounding hypointensity of the lesion on the T1-weighted image suggests previous hemorrhage. Intervening brain parenchyma can be seen on the T2-weighted image. c: Light microscopic study revealing dilated capillary structure with intervening brain tissue. H & E, original magnification ×100; bar = 100 µm. d and e: Postoperative CT scans showing no bleeding. F: Intraoperative photograph showing the vascular lesion (arrows). Figure is available in color online only.
Most of the reported BCTs are small and clinically silent.\textsuperscript{12} Traditionally, invasive management had been avoided in small asymptomatic BCTs because of the high risk of hemorrhage. Nevertheless, recent studies revealed that untreated large or giant BCTs tend to have an aggressive course with hemorrhage, which might lead to permanent neurological deficits or even death.\textsuperscript{12,13,27,29} Conservative management might be insufficient to either prevent the progression or control the symptoms. In the previous literature, surgical management of giant and large BCTs has been reported to have favorable outcomes.\textsuperscript{4,6,17,18,22,24,25,27} In our case series, all lesions were symptomatic and their surgical removal lesions resulted in significant symptom relief.

\textbf{Diagnosis of BCTs}  
Diagnosis of capillary telangiectasia is mainly based on location and radiological characteristics.\textsuperscript{3,9,10,12,16} Differentiating BCTs from other brain lesions such as infarction, neoplasm, and hemorrhage is challenging.\textsuperscript{19} Large BCTs have been frequently misdiagnosed. Precise identification of the lesion is critical for both diagnosis and management.\textsuperscript{1,6,8,19,25,26}  
Mild or moderate enhancement with irregular borders on contrast-administered T1-weighted imaging seems to be a common finding for both small and large BCTs.\textsuperscript{10,27} For small BCTs, T1- and T2-weighted images alone are oftentimes insufficient to establish a diagnosis, so that susceptibility-weighted images and diffusion-weighted images are often required.\textsuperscript{5,9,10} In contrast, large BCTs can be visualized on T1- and T2-weighted images as the vessel abnormalities are quite prominent.\textsuperscript{9} After contrast administration, larger BCTs often demonstrate weblike linear enhancement with heterogeneously intense nodules.\textsuperscript{9} In our series, the lesions were visible on T1- and T2-weighted images in Cases 1 and 2, presenting as prominent brush-like cone-shaped lesions with linear enhancement accompanied by interspersed hypo- and hyperintense nodules. On T2-weighted images, a hypointense margin around the proximal part of the lesions was observed in 3 cases, suggestive of old hemorrhage or a coexisting CM. Dilated draining veins were observed in 4 of the 5 cases (80%) in our series.\textsuperscript{7,12}  
Gross et al. proposed that there might be a stage in the formation between BCT and CM, which could be defined as a new entity.\textsuperscript{12} The unique characteristics of the giant BCTs in our series add more support to this theory.\textsuperscript{6,8,11} Small BCT and CM might be the two ends of the same pathological entity, with giant or large BCT as the bridging developmental stage. Moreover, it has been suggested that either BCT or CM could develop as a consequence of dynamic change in the blood flow and then develop into the other type of lesion (i.e., CM or BCT).\textsuperscript{12,23}

\textbf{Histopathology of Large BCTs}  
The histopathological characteristics of BCTs have been well described in previous studies.\textsuperscript{10,15,16,20} Micro-

![Fig. 4. Case 4. A and B: Axial T1-weighted (A) and T2-weighted (B) images showing a subtle isointense lesion within the left temporal lobe. C: Postcontrast T1-weighted image showing mild enhancement of the lesion with linear enhancement. D and E: Postoperative T1- and T2-weighted images. No residual lesion can be seen. F: Light microscopy showing the typical histopathological appearance of a BCT. H & E, original magnification \(\times100\); bar = 100 \(\mu m\). Figure is available in color online only.](image-url)
symptomatic large or giant capillary telangiectasias

In all cases, BCTs are composed of numerous thin-walled, well-formed capillary vessels with interspersed normal neuronal tissue. The pathogenic relationships among BCT, CM, and developmental venous anomaly have been increasingly discussed. Both BCT and CM have been reported to have formed de novo with the presence of developmental venous anomaly. Restricted venous outflow may cause capillary hypertension and induce dilation of the vessel wall. Also, increased arterial perfusion caused by capillary dilation might lead to development of a venous anomaly. The presence of histopathological characteristics of BCT with radiological signs of CM observed in our series indicate BCT and CM might have the same pathological origin. In previous studies, it has been suggested that CM is a late stage of evolution of the BCT. In recent years, there have been increasing reports of mixed BCTs and CMs.

Treatment of Large or Giant BCTs

For small asymptomatic lesions, conservative treatment is the first choice. However, unlike small BCTs, hypoperfusion of the involved region is more severe in large BCTs. Ischemic accident has been reported in previous studies. In addition, development of an aggressive form with increased risk of bleeding from giant or diffuse BCTs has also been reported. In the existing reports of 10 cases of symptomatic large BCTs, 8 underwent resection. Favorable outcomes without significant neurological deficits were documented in most of these cases. In our series, patients with intractable seizures were seizure free, and the patient with episodic visual disturbances regained her vision gradually. Conservative therapy should be recommended after surgery to reach optimal control of the symptoms.

Conclusions

Large BCTs present with radiological, histopathological, demographic, and clinical features that are distinct from small BCTs. The frequently observed coexistence of CM indicates the existence of a form occurring between the stages of capillary and venous malformation. This entity should be managed in time to avoid severe sequelae as reported in previous studies. Surgery might offer a good option for treatment of patients with intractable neurological symptoms, especially when the location of the lesions is surgically accessible, and complete removal is expected to provide effective relief of the symptoms without causing new neurological deficits.

References


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Disclosure
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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