

Diffuse Capillary Telangiectasia of the Brain Manifested as a Slowly Progressive Course

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Key Words

Capillary telangiectasia · Cerebral blood flow · Xenon CT

Abstract

Brain capillary telangiectasia (BCT) are usually small, solitary, benign in clinical manifestation, or found incidentally at autopsy. However, diffuse BCT are rarely reported. A 39-year-old woman had the first generalized seizure 10 years previously. Thereafter, she had sustained progressive spastic paraparesis and blurred vision. Computed tomography (CT) showed diffuse brain atrophy with numerous calcified spots. Contrast T1-weighted magnetic resonance images showed diffuse faintly enhancing lesions with stippled appearance in the whole brain. Cerebral angiography revealed multiple small nets of dilated capillaries and Xenon CT showed diffuse low cerebral blood flow in the resting and good cerebral reserve capacity in the acetazolamide challenge test. Therefore, diffuse BCT was diagnosed clinically. Although BCT are not rare, diffuse BCT with slowly progressive neurological symptoms had never been reported before. It causes global cerebral ischemia that lead to brain atrophy and a degenerative course.

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Brain capillary telangiectasias (BCT) are composed of multiple dilated capillaries surrounded by normal tissue [1]. The natural course is usually benign, and clinically aggressive BCT has rarely been reported [2]. Here, we report a case of diffuse BCT with presentation as late-onset seizure, progressive vision impairment and spastic paraparesis.

Case Report

A 39-year-old woman had experienced the first generalized seizure 10 years previously, without preceding head trauma, infection or family history. Two years later, insidious-onset, slowly progressive spastic paraparesis occurred. Slurred speech and blurred vision of both eyes were also found.

At the first admission at the age of 34 years, serum and cerebrospinal fluid biochemical studies were unremarkable, and head magnetic resonance imaging (MRI) showed multiple small enhanced lesions on gadolinium-enhanced T₁-weighted images. No conclusive diagnosis was made at that time. However, her symptoms progressed in the following years. So she was admitted again 5 years later for re-evaluation.

On admission, the patient's appearance was normal without visible lesions on the face, trunk or limbs. Cranial nerve examinations showed poor visual acuity in both eyes (20/200 OU), with normal pupil sizes and light responses. Her muscle power showed mild weakness with hyperreflexia of the four limbs and extensor plantar responses bilaterally. Sensation and coordination functions were intact. Her gait was unsteady and affected by marked spasticity and circumduction of both legs.

Laboratory studies of ordinary hematology and blood biochemistry, erythrocyte sedimentation rate, C-reactive protein, endocrine

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and immune profiles were within normal limits. Head computed tomography (CT) showed diffuse brain atrophy with numerous small calcified spots in the whole cerebrum and cerebellum. Head MRI revealed no abnormality on T₁- and T₂-weighted images but gadolinium-enhanced T₁-weighted images showed diffuse faintly enhanced lesions with stippled or brush-like appearance in the bilateral cerebrum, cerebellum and brainstem involving both gray and white matters (fig. 1a–c). The size and location of enhanced lesions were similar to those seen on MRI 5 years earlier. Cervical spine MRI revealed no definite abnormality. Cerebral angiography showed cerebral arteries with a normal main trunk but multiple small nests of dilated capillaries either in the anterior or posterior circulation (fig. 1d). Xenon CT revealed diffuse, low cerebral blood flow in the resting and good cerebral reserve capacity in the acetazolamide challenge test (fig. 2).

Combining the clinical course and imaging findings, symptomatic diffuse BCT was diagnosed. A further pathological study was suggested but was not accepted by both the patient and her family.

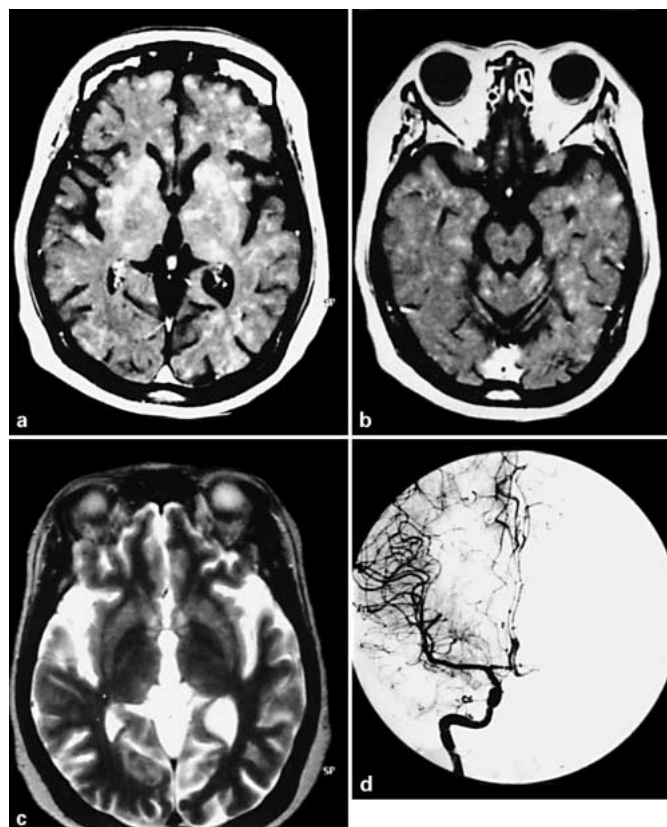


Fig. 1. Gadolinium-enhanced T₁-weighted MRI of the brain shows marked brain atrophy and diffuse faintly enhanced lesions with stippled or brush-like appearance in bilateral cerebral and cerebellar hemispheres and brainstem involving both gray and white matters (a, b) and a normal appearance of T₂-weighted MRI of the brain (c). Cerebral angiography shows multiple small nets of dilated capillaries mainly in the subcortical area by the internal cerebral artery injection (d).

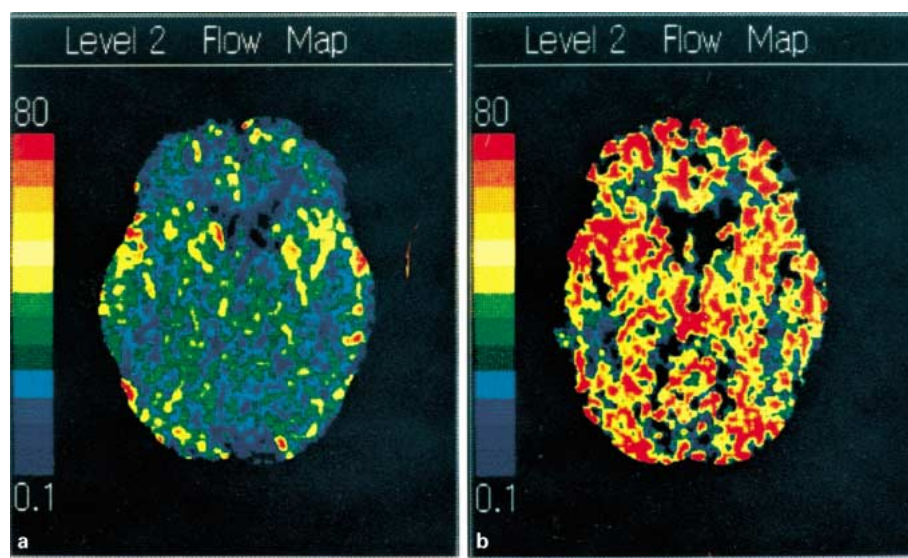


Fig. 2. A xenon CT study reveals diffuse low cerebral blood flow in the resting state (a) and good cerebral reserve capacity in the acetazolamide challenge state (b).

Discussion

Vascular malformations of the brain include arteriovenous malformations, venous angiomas, cavernous angiomas and BCT [1]. Characteristic MRI findings of different vascular malformations have been well known [3]. Angiography of BCT usually appears normal but occasionally shows multiple nests of dilated capillaries with prolonged vascular staining in some instances such as patients with Osler-Weber-Rendu disease [4].

In one study of 18 patients with BCT, their clinical course was usually benign, most had a single lesion localized in the pons, temporal lobe, medulla or caudate nucleus except for one who had 2 lesions in the pons [5]. The relationship between BCT and clinical symptoms is still uncertain. Based on previous studies, symptomatic BCT could be due to hemorrhage into the adjacent brain parenchyma or to direct compression by a local mass [6, 7]. The dilated capillaries – like blood pools and cerebral blood – may become static which would lead to cerebral hypoperfusion and hypoxic insult.

Our patient had late-onset seizure, progressive vision impairment and spastic paraparesis. BCT could have been diagnosed based on characteristic findings from the MRI study, even though a pathological study was lacking. Unchanged size and location in a 5-year follow-up study could rule out other diagnoses, such as neoplasm, demye-

lination disorder or subcortical infarction. Multiple small nests of dilated capillaries on cerebral angiography further confirmed this particular vascular malformation. Although mixed vascular malformations are not uncommon (especially mixed BCT and cavernous malformation) [8], the lack of hallmarks on T₂-weighted MRI such as a surrounding hypointense hemosiderin ring with an internal popcorn appearance made coexistent cavernous malformations less likely.

In our case, xenon CT revealed diffuse, low cerebral blood flow in the resting study and good cerebral reserve capacity in the challenge test. These findings could be interpreted as diffuse BCT decreasing cerebral blood flow because of a decreased demand for flow due to either cerebral neuronal loss or decreased metabolic activity. In this case, chronic insufficiency-related extensive neuronal loss could be demonstrated indirectly by the findings of numerous small calcifications and diffuse brain atrophy in the head CT study.

Conclusion

Although BCT is not rare, as far as we know, diffuse BCT has never been reported. A clinical course suggesting neurological degenerative disease should evaluate the possibility of BCT.

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