



# Large Dolichoectasia Concurrently Involving Both Anterior and Posterior Cerebral Circulation: A Case Report

전순환계와 후순환계를 동시에 침범한 긴머리확장증  
환자의 영상 소견: 증례 보고

Youngsoo Soh, MD , Soo Jung Kim, MD , Eun Ae Yoo, MD\*

Department of Radiology, Presbyterian Medical Center, Jeonju, Korea

## ORCID iDs

Youngsoo Soh <https://orcid.org/0000-0002-0670-8523>

Soo Jung Kim <https://orcid.org/0000-0001-7861-6217>

Eun Ae Yoo <https://orcid.org/0000-0001-7903-354X>

Received November 30, 2021

Revised February 28, 2022

Accepted April 14, 2022

## \*Corresponding author

Eun Ae Yoo, MD  
Department of Radiology,  
Presbyterian Medical Center,  
365 Seowon-ro, Wansan-gu,  
Jeonju 54987, Korea.

Tel 82-63-230-8006

Fax 82-63-230-8387

E-mail [silver-824@hanmail.net](mailto:silver-824@hanmail.net)

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<https://creativecommons.org/licenses/by-nc/4.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

Dolichoectasia is an uncommon disorder characterized by the presence of a dilated, elongated, and tortuous cerebral artery. Its main pathologic mechanism is the disruption of the internal elastic lamina. Risk factors for dolichoectasia are advanced age, chronic hypertension, and metabolic disease. It mainly involves the vertebrobasilar vasculature (or “posterior circulation”), but dolichoectasia can also be seen in the anterior circulation, particularly the anterior cerebral artery. There are no reported cases of dolichoectasia involving both anterior and posterior circulation in South Korea. Here we report an unusual case of dolichoectasia involving both anterior and posterior circulation in a young female without any underlying disease on the basis of prominent imaging findings.

**Index terms** Ectasia; Cerebral Circulation; Internal Carotid Artery; Middle Cerebral Artery; Posterior Cerebral Artery; Digital Subtraction Angiography

## INTRODUCTION

Dolichoectasia is an uncommon disorder characterized by dilatation (ectasia), elongation (dolichosis), tortuous morphology and occasionally aneurysmal change in the cerebral artery (1-3). It mainly involves the vertebrobasilar system of the posterior circulation, but it also can be affected in the anterior circulation of the brain (3, 4). It is mostly asymptomatic (3), but the diverse clinical presentations such as compressive symptoms on surrounding brain structure and vascular events (transient ischemic attacks [TIAs], ischemic or hemorrhagic stroke) can

occur (1). Although it can cause these kinds of various brain injuries, few radiologic researches for dolichoectasia have been reported in South Korea (4). Besides, existing articles on dolichoectasia are largely about cases occurring in the vertebrobasilar system and in elderly patients (1-3).

We report a rare case of dolichoectasia involving both anterior and posterior circulation simultaneously, showing more severe dolichoectasia in the anterior circulation with aneurysm in an adolescent female on the basis of prominent radiologic findings on CT, digital subtraction angiography (DSA) and MR angiography (MRA).

## CASE REPORT

An 18-year-old female visited our emergency department with syncope and headache. Neurologic examinations and laboratory results showed no remarkable findings. She didn't have any underlying disease. Contrast enhanced brain CT with CT angiography was taken.

CT showed a dense calcification along the vessel wall (Fig. 1A, left image) and dilatation of the left side circle of Willis (Fig. 1A, middle and right image) with no evidence of brain ischemia or hemorrhage. She was diagnosed with dolichoectasia in intracranial artery. For further evaluation, DSA was taken, and it demonstrated diffuse dilatation of the left distal internal carotid artery (ICA) and M1 branch of the middle cerebral artery (MCA) (Fig. 1B, left image), with aneurysm of the ectatic left distal ICA (Fig. 1B, middle image). There was also corkscrew-like prominent tortuosity with mild dilatation of the left proximal posterior cerebral artery (PCA) (Fig. 1B, right image, Fig. 1C). During admission, the patient was given conservative treatment for headache.

Four years later, a brain MRA for follow up was taken. It showed a midbrain compression by tortuous and mildly dilated left proximal PCA, a focal small outpouching aneurysm (3.5 mm sized, posterior direction) from the tortuously ectatic left distal ICA (Fig. 1D, left image), a markedly tortuous elongated dilatation of the left distal ICA (C7), diffuse dilatation of the left proximal MCA, and corkscrew-like prominent tortuosity with mild dilatation of the left proximal PCA (Fig. 1E). It revealed no change of the dolichoectasia in both anterior and posterior cerebral circulation. A dilated ICA could potentially compress cranial nerve, but in this patient, there was compression on upper brain stem only without compression of cranial nerve. She had no symptoms related to cranial nerve compression.

This study was approved by the Institutional Review Board of our institution and the requirement for informed consent was waived (IRB No. E2021-047).

## DISCUSSION

Dolichoectasia is a rare disorder characterized by dilated, elongated and tortuous cerebral artery (2, 3). Since ectasis is the key feature of dolichoectasia, it has been called dilatative arteriopathy (2). Although mostly affected in the vertebrobasilar system of the posterior cerebral circulation, it also can be seen in the anterior circulation, particularly the anterior cerebral artery (3). And it can involve both vertebrobasilar system and anterior cerebral circulation simultaneously (2-5). Risk factors for dolichoectasia are old age, chronic hypertension, metabolic dis-

ease and hereditary disease that affect vascular interstitial structure formation (1-3, 5).

Dolichoectasia can be considered as part of a systemic vasculopathy because of its association with enlarged descending thoracic aorta and coronary arteries, other systemic arterial diseases such as aortic aneurysm and cerebral microangiopathy (6). A possible genetic predis-

**Fig. 1.** An 18-year-old female with dolichoectasia.

**A.** Precontrast axial CT of the brain shows a dense calcification (arrow) along the vessel wall of the left side of the circle of Willis (left image). Postcontrast axial CT of the brain shows a dilated left side of the circle of Willis (arrows) (middle image). Three-dimensional volume rendering image of CTA shows a dilated left side of the circle of Willis (right image).

**B.** DSA lateral view of left ICA angiogram shows diffuse dilatation of the left distal ICA and MCA M1 branch (left image). DSA AP view of left ICA angiogram shows a small aneurysm (arrow within circle) of the ectatic left distal ICA (middle image). DSA AP view of left PCA angiogram shows corkscrew-like prominent tortuosity (arrow within circle) with mild dilatation of the left proximal PCA (right image).

**C.** Three-dimensional rotational angiography images of left PCA show corkscrew-like prominent tortuosity (arrows) with mild dilatation of the left proximal PCA.

AP = anteroposterior, CTA = CT angiography, DSA = digital subtraction angiography, ICA = internal carotid artery, MCA = middle cerebral artery, PCA = posterior cerebral artery

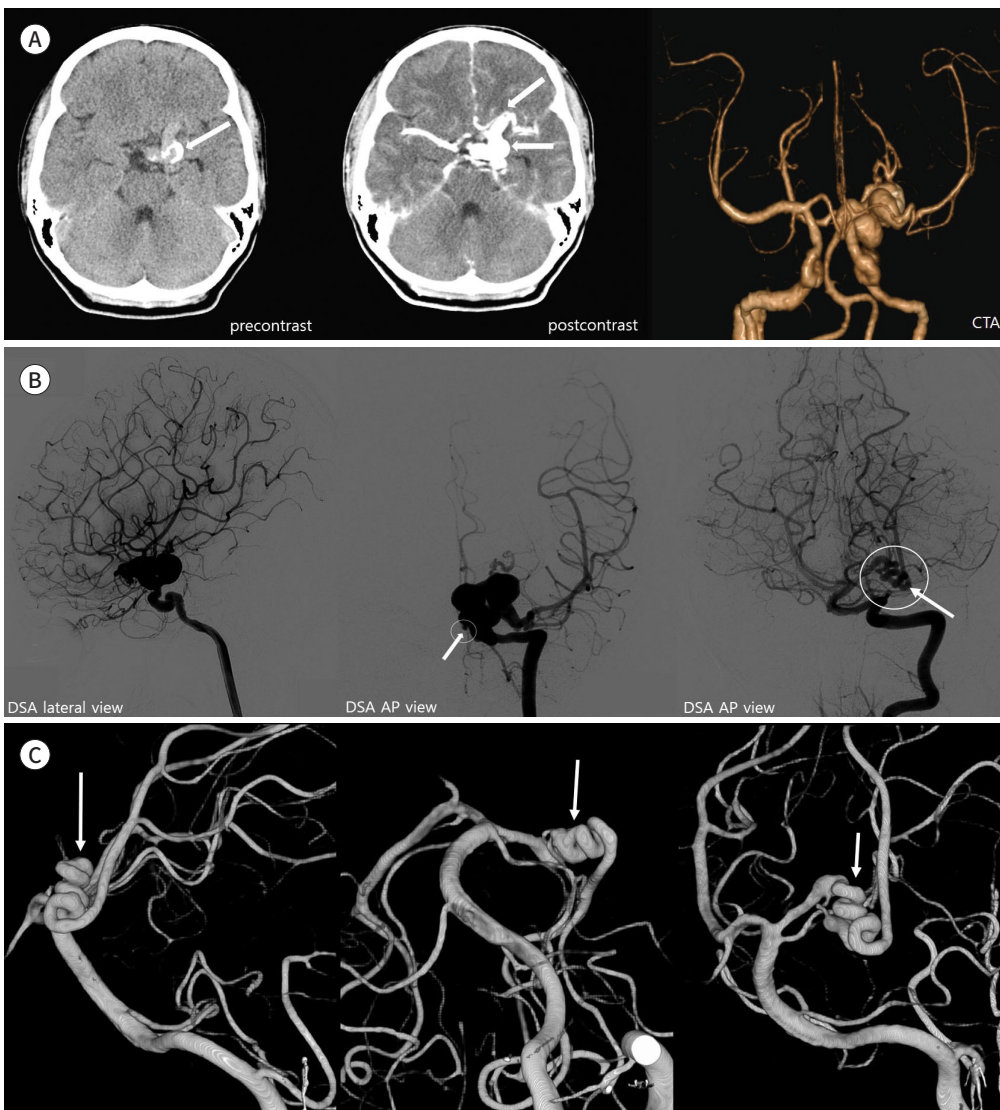
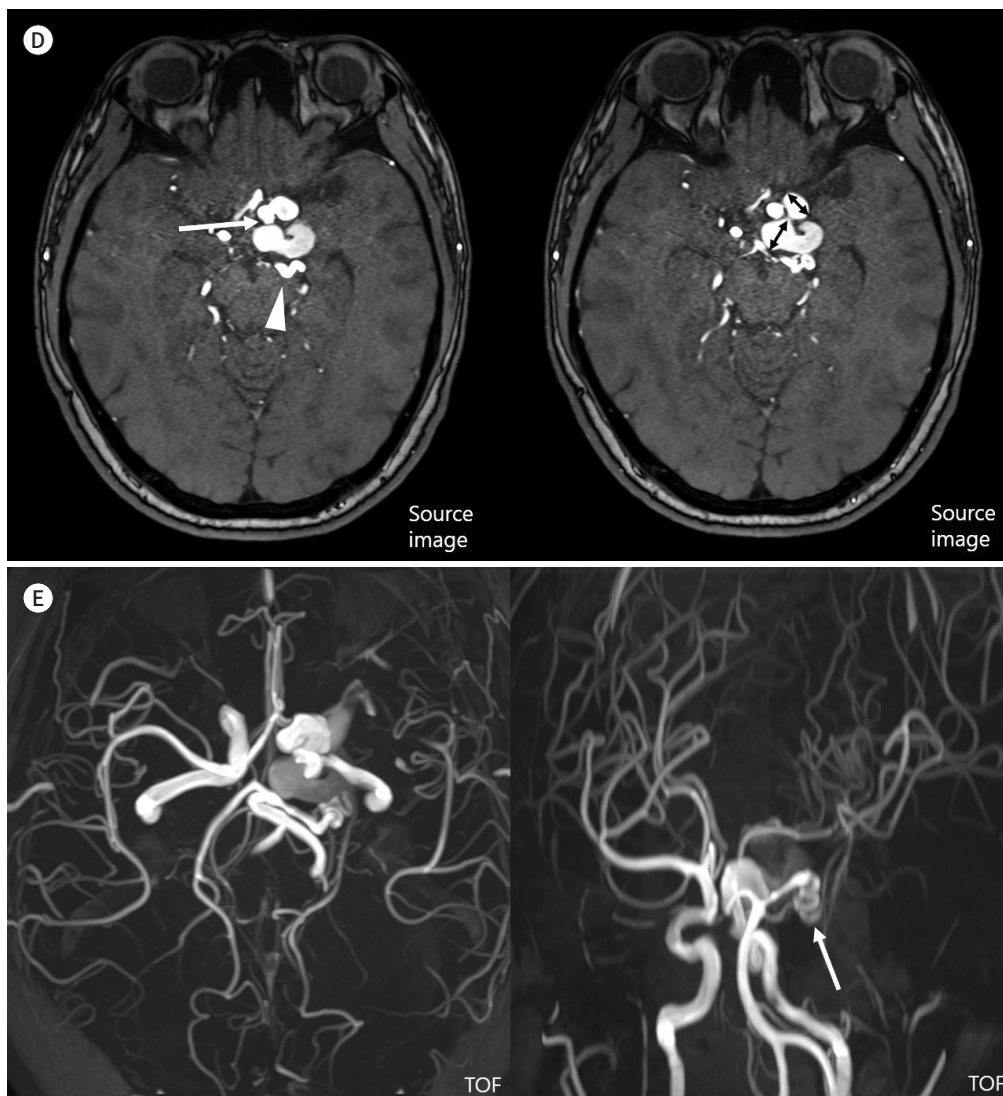


Fig. 1. An 18-year-old female with dolichoectasia.

**D.** Four-year follow-up MRA. Source image of TOF MRA of the brain shows midbrain compression (arrow-head) by tortuous and mildly dilated left proximal PCA. It also shows a focal small outpouching aneurysm (arrow) from tortuously ectatic left distal ICA (left image). Source image of TOF MRA of the brain shows dilated left distal ICA with a diameter of 9.9 mm (upper arrows at both ends) and a dilated left proximal MCA with a diameter of 11.2 mm (lower arrows at both ends) (right image).

**E.** Four-year follow-up MRA. TOF MRA of the brain shows markedly tortuous elongated dilatation of the left distal ICA, and diffuse dilatation of the left proximal MCA without interval change (left image). TOF MRA of the brain again shows corkscrew-like prominent tortuosity with mild dilatation of the left proximal PCA (arrow) (right image).

ICA = internal carotid artery, MCA = middle cerebral artery, MRA = MR angiography, PCA = posterior cerebral artery, TOF = time-of-flight



position for dolichoectasia has been proposed due to its association (6) with polycystic kidney disease, Ehlers-Danlos and Marfan syndromes, Pompe disease (1, 6), tuberous sclerosis, cavernous angioma, craniocervical malformations (1), neurofibromatosis type 1, Fabry disease, and sickle cell disease. Other associations were reported including infections such as syphilis, varicella-zoster virus (6), and the acquired immune deficiency syndrome (AIDS) (2). Immuno-

globulin G4-related disease is also known to be associated with dolichoectasia (6). Hence, the above-mentioned diseases must be ruled out when dolichoectasia is diagnosed.

Main pathologic mechanism of dolichoectasia is the disruption of the internal elastic lamina (1, 3) and thinning of the media by smooth muscle atrophy (3). Angiogenesis develops with intimal hyperplasia and co-exists with intramural hemorrhage and thrombus formation (1, 5).

When dolichoectasia occurs in the posterior circulation, it is more often asymptomatic than in the anterior circulation (1). Symptoms result from three mechanisms: compression, rupture and ischemia. According to these suggested mechanisms, diverse clinical presentations can appear (3, 7). For compression, there are compressive symptoms on brain stem or cranial nerve, cerebrospinal fluid flow obstruction by direct compression on the third ventricle and hydrocephalus (2, 7, 8). As for symptoms related to rupture, intracranial hemorrhage of the dilated vessel and resultant compressive symptom can occur (5). Ischemic symptoms are ischemic stroke due to thrombus formation or TIA (2, 5).

Smoker and colleagues developed an assessment criteria for dolichoectasia in basilar arteries (1). In this criteria, there are three quantitative measures on the morphology of the basilar artery. Each of them are 'laterality score for tortuosity, bifurcation height for elongation, and basilar artery diameter for the degree of dilatation' (1, 6). For giving a definition on ectasia in basilar artery, cutoff value of 4.5 mm in arterial diameter at mid pons level was suggested (2, 5). Smoker's criteria is helpful to diagnose dolichoectasia in the basilar artery. But it cannot be applied to other cerebral vessels (1, 2). As our case showed involvement of dolichoectasia in the anterior and posterior circulation without involving basilar artery, it is not fit to our case for assessing dolichoectasia by Smoker's criteria.

Passero and Rossi have proposed diameter cutoffs to demonstrate ectasia for vessels other than the basilar artery (2). Its suggested cutoff values are  $\geq 7$  mm for the ICA,  $\geq 4$  mm for the MCA, and  $\geq 4$  mm for the vertebral artery (2). Our case revealed dilated diameters of 9.9 mm in left distal ICA and 11.2 mm in left proximal MCA (Fig. 1D, right image) exceeding the cutoff values.

Gutierrez and colleagues have also made a proposal for determining dolichosis of vessels other than the basilar artery (2). It is a visual assessment of the compression on adjacent structures and the vascular tortuosity in comparison with the contralateral artery (2). In this patient's source image of time-of-flight MRA, it showed midbrain compression by tortuous and mildly dilated left proximal PCA (Fig. 1D, left image). Left proximal PCA showed corkscrew-like prominent tortuosity with mild dilatation compared to right PCA (Fig. 1C).

There is no established treatment for dolichoectasia except treatment for dolichoectasia-associated complications (1, 6). To prevent ischemic event, anticoagulants or antiplatelet agents can be used. But, antithrombotics may increase risk of intracerebral hemorrhage (9). In addition, ventriculoperitoneal shunting for hydrocephalus (9) and decompression surgery for compressive symptoms can be done (1). For asymptomatic patients like our case, conservative management is favored. But many other attempts for asymptomatic patients such as sling retraction, vascular clip grafting, and dolichoectasia vessel bypass and exclusion have been reported to be successful (1).

Passero and Rossi (10) conducted a prospective clinical and imaging follow-up of 156 verte-

probasilar dolichoectasia (VBD) patients for about 11.7 years. The results revealed that 93 patients (60%) experienced more than one event: 75 patients developed stroke (59 ischemic and 21 hemorrhagic), 31 patients developed compressive symptoms, and 2 patients developed hydrocephalus. The events were highly associated with the severity of VBD. Progression of VBD was seen in 43% of patients and it had an association with a higher morbidity and mortality. For follow-up period, 62 patients expired and most of them had stroke. The prognosis of VBD relied mostly on the severity of the state at diagnosis and on its progression. Our case presented dolichoectasia in the anterior circulation (left distal ICA and M1 branch of the MCA) and the posterior circulation (left proximal PCA) with no involvement of vertebrobasilar artery. Our patient's long-term prognosis on dolichoectasia, therefore, could be expected indirectly from Passero and Rossi's paper about VBD.

We report a rare case of dolichoectasia that has not been reported yet in South Korea. It is interesting that it involved both anterior and posterior cerebral circulation in an adolescent female without any risk factor or any underlying disease and any significant neurologic symptom.

#### Author Contributions

Conceptualization, Y.E.A.; data curation, S.Y.; investigation, S.Y.; methodology, Y.E.A.; project administration, K.S.J.; resources, K.S.J.; supervision, Y.E.A.; visualization, S.Y.; writing—original draft, S.Y.; and writing—review & editing, Y.E.A.

#### Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

#### Funding

None

## REFERENCES

1. Gutierrez J, Sacco RL, Wright CB. Dolichoectasia—an evolving arterial disease. *Nat Rev Neurol* 2011;7:41-50
2. Del Brutto VJ, Ortiz JG, Biller J. Intracranial arterial dolichoectasia. *Front Neurol* 2017;8:344
3. Uchino A, Kohyama S. Long segment dolichoectasia of the right internal carotid artery diagnosed by CT angiography. *Radiol Case Rep* 2019;14:1084-1087
4. Tien KL, Yu IK, Yoon SJ, Yoon YK. CT and MR imaging features in patients with intracranial dolichoectasia. *J Korean Radiol Soc* 2000;42:205-214
5. Kim JY, Kim JW, Yoon MJ, Suk SH. Acute subarachnoid hemorrhage due to giant vertebrobasilar dolichoectasia. *J Neurocrit Care* 2019;12:117-121
6. Samim M, Goldstein A, Schindler J, Johnson MH. Multimodality imaging of vertebrobasilar dolichoectasia: clinical presentations and imaging spectrum. *Radiographics* 2016;36:1129-1146
7. Yuh SJ, Alkherayf F, Lesiuk H. Dolichoectasia of the vertebral basilar and internal carotid arteries: a case report and literature review. *Surg Neurol Int* 2013;4:153
8. Kim BS, Lee JG, Ahn S, Park JH. Huge vertebrobasilar dolichoectasia. *J Korean Neurol Assoc* 2019;37:432-434
9. Nouh A, Remke J, Ruland S. Ischemic posterior circulation stroke: a review of anatomy, clinical presentations, diagnosis, and current management. *Front Neurol* 2014;5:30
10. Passero SG, Rossi S. Natural history of vertebrobasilar dolichoectasia. *Neurology* 2008;70:66-72

## 전순환계와 후순환계를 동시에 침범한 긴머리확장증 환자의 영상 소견: 증례 보고

소영수 · 김수정 · 유은애\*

긴머리확장증은 확장, 연장 및 구불구불한 형태의 대뇌동맥을 특징으로 하는 드문 질환이다. 주요 병리기전은 내탄력판의 파괴로 알려져 있으며, 위험요인으로 고령, 만성 고혈압, 그리고 대사성 질환 등이 있다. 긴머리확장증은 주로 후순환계의 척추뇌기저동맥을 침범하는 것으로 알려져 있으나, 전순환계, 특히 전대뇌동맥에 이환되는 경우도 있다. 긴머리확장증이 전순환계와 후순환계를 모두 침범한 사례는 아직 국내에 보고된 바가 없다. 이에 우리는 기저 질환이 없는 젊은 여자 환자에서 전순환계와 후순환계가 모두 이환된 매우 희귀한 형태의 긴머리확장증 사례를 현저한 영상 소견을 토대로 보고하고자 한다.

전주예수병원 영상의학과