## CASE REPORT

# Dolichoectasia of the Anterior Cerebral Circulation: a Case Report

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## INTRODUCTION

Dolichoectasia is a cerebral arterial disorder characterised by elongation, dilatation, and tortuosity of the diseased artery.[1] Half of the cases of dolichoectasia are asymptomatic.[2] However, when symptoms are present, they can manifest as headache and be complicated by ischaemic changes, haemorrhage, and compressive effects, particularly for cranial nerves.[3]

Dolichoectasia affects the posterior cerebral circulation more commonly than the anterior.[4] For posterior cerebral circulation, the diagnosis criteria for dolichoectasia of the basilar and vertebral arteries are Smoker's and Ubogu's criteria, respectively.[2,5,6] For dolichoectasia of the anterior cerebral circulation, quantifications have been made in the literature to aid diagnosis.[7-9]

We report a case of dolichoectasia of the anterior cerebral circulation, first suspected on computed tomography (CT) to be arteriovenous malformation (AVM) or dural arteriovenous fistula (DAVF), and was confirmed to be dolichoectasia using digital subtraction angiogram (DSA). This case highlights the importance of considering dolichoectasia as a differential diagnosis, since its differentiation from AVM or DAVF has important clinical implications in terms of subsequent treatment options. The case also highlights the important role of DSA in differentiating AVM or DAVF from dolichoectasia.

## CASE REPORT

A 15-year-old girl with good past health presented to the accident and emergency department with sudden onset of headache with syncope. Physical examination was unremarkable; the patient had stable vital signs and no deficit on neurological assessment. A non-contrast CT scan was initially performed, which prompted subsequent contrast CT scans for further investigation.

The CT scans of the whole brain, from skull base to vertex, were performed on a 64-slice CT scanner (LightSpeed VCT, GE Healthcare, United States). The non-contrast scan was obtained in axial mode with tilting along the occipitomental line, and the CT angiogram (CTA) and CT venogram (CTV) were obtained in helical mode with no tilting. Intravenous contrast (70 mL Omnipaque 300, at 4 mL/s) was injected using a power injector via an 18-gauge catheter positioned in the antecubital vein. CTA was carried out with the assistance of bolus tracking with monitoring of the petrous internal carotid artery (ICA). Thereafter, CTV was acquired at 60 s after contrast injection.

Non-contrast CT brain revealed a serpiginous structure along the superior surface of the corpus callosum without associated mass effect (Figure 1). This was suspected to be an abnormal dilated cerebral vein related to a vascular anomaly such as AVM or DAVF. There was no intracranial haematoma or significant mass effect.

A subsequent CTA and CTV revealed normal calibre of the right proximal anterior cerebral artery (ACA) that abruptly transitioned to dilated and tortuous callosomarginal and pericallosal branches which corresponded to the aforementioned serpiginous structure. These branches appeared connected to the superficial cerebral veins that eventually drained to the superior sagittal sinus (Figure 2). The right A1 branch measured 2.3 mm while the right A2-A3 branch measured 3 mm. The calibre of the left ACA was normal. The left A1 branch and the left A2-A3 branch both measured 1.5 mm. Differential diagnoses included dolichoectasia and AVM or DAVF involving the callosomarginal and pericallosal arteries.

To exclude AVM or DAVF, an urgent DSA was performed and confirmed dilated and tortuous branches of the right ACA A2/A3 segment, corresponding to the pericallosal and callosomarginal artery. There was no early venous drainage to suggest AVM or DAVF (Figure 3). The final diagnosis was dolichoectasia of the ACA.

The patient was subsequently discharged after no further episodes of headache. Follow-up non-contrast magnetic resonance imaging 4 months later was performed on a 3T clinical scanner (Achieva TX, Philips Medical Systems, Best, The Netherlands). This revealed no associated infarcts or other abnormal signal related to the right ACA dolichoectasia (Figure 4). The dilated and tortuous callosomarginal and pericallosal branches showed normal flow void on T2-weighted sequence, and normal flow signal on time-of-flight magnetic resonance angiography (MRA) time flight, with no evidence of thrombosis. The branches deviated laterally and compressed on the right medial frontal lobe.

## DISCUSSION

In this case, the patient was initially suspected of having AVM or DAVF with the final diagnosis of dolichoectasia of the anterior cerebral circulation. Dolichoectasia of the anterior circulation is less common than posterior circulation involvement, and to the best of our knowledge distal branch dolichoectasia has only been specifically described in one other study. Doran et
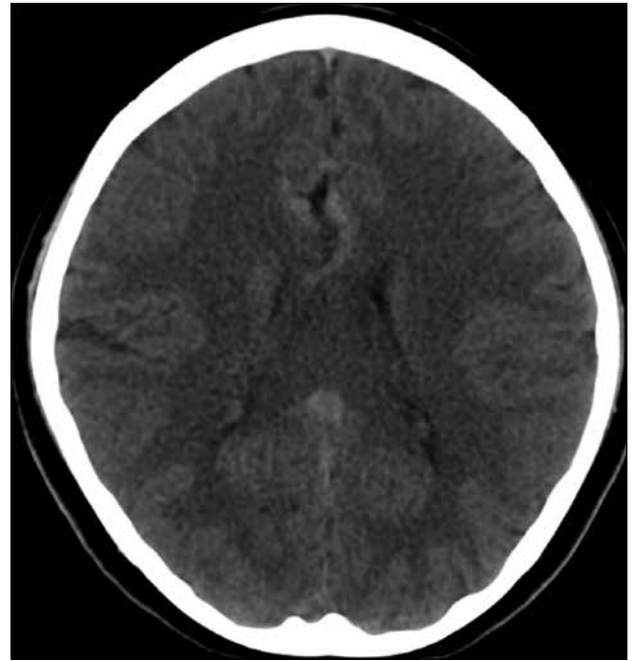


**Figure 1.** Plain computed tomography image of the brain showing slightly hyperdense serpiginous structure along corpus callosum.
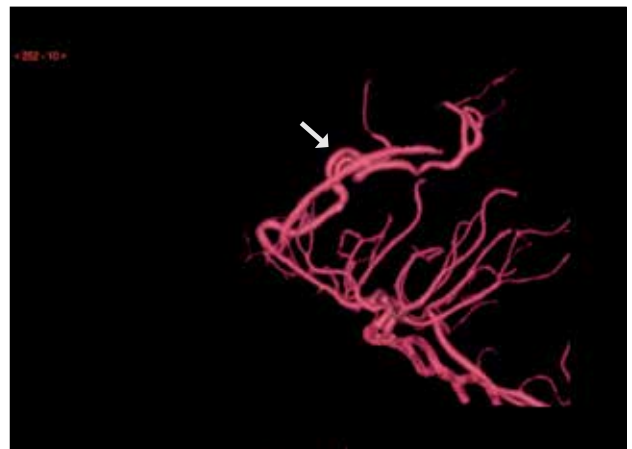


**Figure 2.** Computed tomography angiogram of the brain showing dilated and tortuous callosomarginal and pericallosal branches of the right anterior cerebral artery (arrow). This was reported to be dolichoectasia or arteriovenous fistula.

al[10] reported an adolescent who presented with seizures and was found to have bilateral ACA A2 dolichoectasia and cortical thickening of the medial frontal lobes, presumed to be a migration abnormality. Our patient was also an adolescent but presented with headache and had isolated and unilateral involvement of the distal ACA A2/A3 segment with no associated brain parenchymal

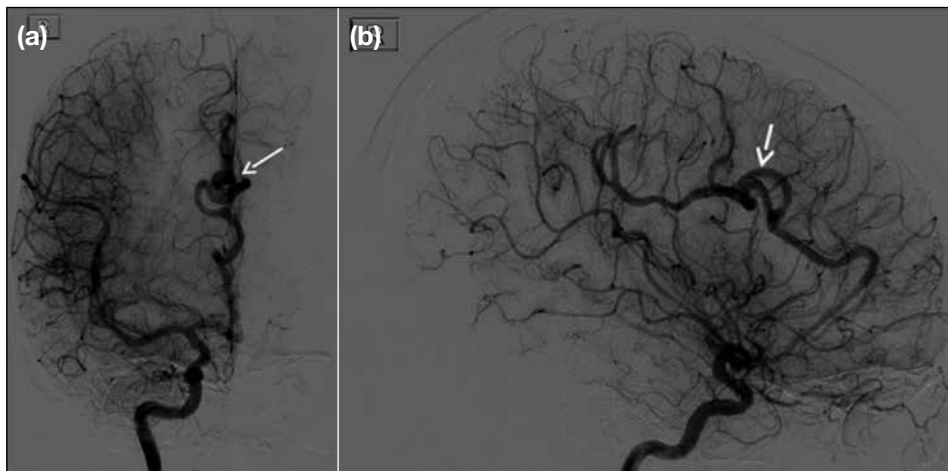**Figure 3.** Anteroposterior (a) and lateral (b) digital subtraction angiogram of the right internal carotid artery of the same patient showing dilated and tortuous callosomarginal and pericallosal branches of the right anterior cerebral artery (white arrows). No earlier draining vein is evident. Features are compatible with dolichoectasia.
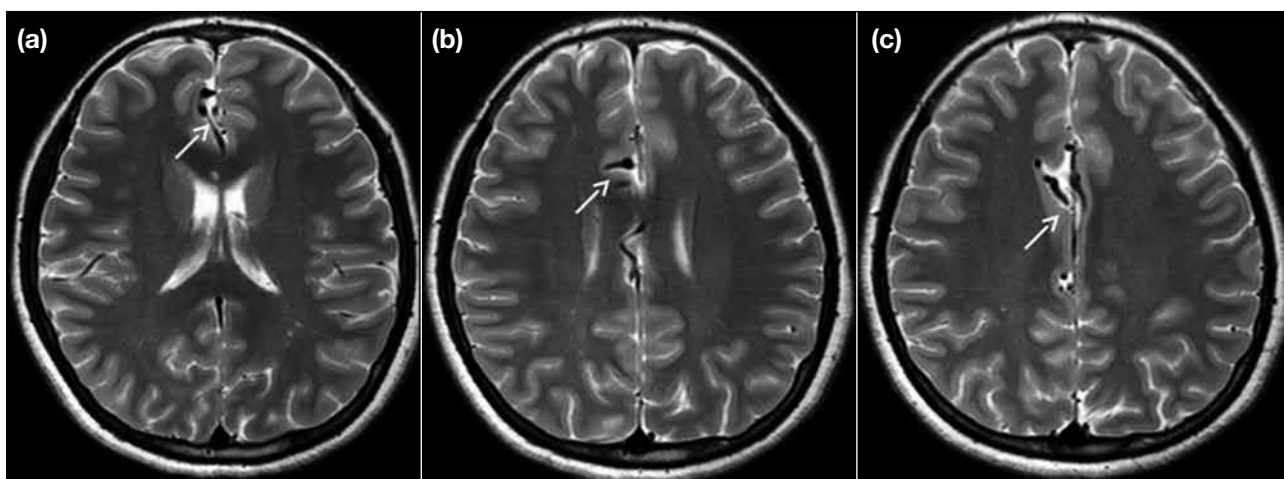


**Figure 4.** Consecutive axial T2-weighted magnetic resonance images (TR 8187 ms; TE 80 ms; flip angle 90°; in-plane resolution: 0.75 mm × 0.94 mm; slice thickness: 2.5 mm) showing the dilated and tortuous callosomarginal and pericallosal branches of the right anterior cerebral artery (white arrows) that measure 2.3-3.0 mm. These deviate laterally and compress the right medial frontal lobe. There is no underlying cerebral parenchymal ischemia or infarct, particularly along the anterior cerebral artery territory.

abnormality. In patients <40 years, the cause of dolichoectasia is largely undetermined but generally presumed to be non-atherosclerotic and congenital.[10] Loss of internal elastic membrane and tunica media in the vessel walls has been observed in histological examinations.[11] This gave rise to the hypothesis that juvenile dolichoectasia is related to inborn connective tissue diseases similar to that of Marfan or Ehlers-Danos syndromes. Intimal disruption from dissection has also been proposed based on other histological studies.[12] However, the exact pathophysiological mechanism remains unclear. For the more elderly population, the pathogenesis is associated with advanced atherosclerotic changes related to hypertension or hyperlipidaemia.[11]

The distinction of AVM or DAVF from dolichoectasia is important, because their clinical management differs. For AVM or DAVF, definitive treatment includes endovascular therapy, radiosurgery, or surgery. For dolichoectasia, asymptomatic patients can be managed conservatively or with clip construction, while symptomatic patients may require surgery. For dolichoectasia of the posterior circulation, the preferred surgery involves parent vessel occlusion with or without bypass.[12,13] Clip reconstruction or transposition of the involved vessel may also be considered. For dolichoectasia of the anterior circulation, surgery involves parent vessel occlusion with bypass, clip reconstruction or wrapping of the affected vessel with

synthetic material.[12,13] Limited evidence suggests that surgical interventions for dolichoectasia of the anterior circulation may have better outcomes than those for posterior circulation.[4] For ACA involvement, success may depend on the presence of abundant leptomeningeal collateral circulation.[13] Given the various methods of treatment, the available resources and expertise will influence the therapeutic approach if the patient opts for active management.

The prevalence of dolichoectasia affecting the posterior circulation is greater than that affecting the anterior circulation. Therefore, imaging diagnostic criteria for diagnosing dolichoectasia of the posterior circulation via imaging is better established. Smoker's criteria describe dolichoectasia of the basilar artery and are based on

CTA findings and use three quantitative measures of basilar artery morphology to define dolichoectasia of the posterior circulation.[5,6] The basilar artery vessel laterality, bifurcation height, and basilar artery diameter measure tortuosity, elongation and the degree of dilatation, respectively. The laterality of the basilar artery (tortuosity) is defined as abnormal if any part of the artery is lateral to the clivus or dorsum or is at the cerebellopontine angle (Table 1). This is based on CT studies which found that the normal position of basilar artery is midline and, when paramedian, it is medial to the lateral margin of the clivus or dorsum. Bifurcation height (elongation) is defined as abnormal when the basilar tip is above the suprasellar cistern (Table 2). This is based on CT findings which found the basilar tip to be below the third ventricle floor in 92% of cases. For basilar artery diameter, the diameter is considered normal if it measures 1.9 mm to 4.5 mm, ie any diameter >4.5 mm is abnormal. In contrast, Ubogu and Zaidat[2] provide criteria for defining tortuosity and elongation of the vertebral arteries. For laterality (tortuosity), any portion of the vertebral artery with deviation >10 mm perpendicular to a straight line joining its intracranial entry point to the basilar artery origin is considered abnormal while if the vertebral portion or origin of the basilar artery is above the level of the pontomedullary junction, then it is considered elongated. Neither Smoker's nor Ubogu's criteria enable differentiation of dolichoectasia from AVM or DAVF.

Currently there are no criteria for anterior circulation dolichoectasia, and identification on CTA or MRA relies on visual assessment of vessel shape abnormality. Gutierrez et al[14] used asymmetry, tortuosity, and compression of the parenchyma to define dolichoectasia of the anterior circulation, whereas Brinjikji et al[7,8] quantified the degree of aneurysmal dilatation as twice the normal diameter of the artery based on an angiographic atlas. Gutierrez et al[14] first looked for asymmetrical vessels identified using coronal views of CTA or MRA

**Table 1.** Criteria for assignment of basilar artery position for vessel laterality. Vessel laterality is abnormal if the most lateral position of the basilar artery is lateral to clivus or dorsum or at the cerebellar pontine angle.[5]

| Position* | |
|---|---|
| 0 | Midline throughout |
| 1 | Medial to lateral margin of clivus or dorsum sellae |
| 2 | Lateral to lateral margin of clivus or dorsum sellae |
| 3 | In cerebellopontine angle cistern |

* Most lateral position identified throughout course of basilar artery.

**Table 2.** Criteria for assignment of basilar bifurcation height. Basilar bifurcation height is abnormal if it is at above the suprasellar cistern.[5]

| Height | |
|---|---|
| 0 | At or below dorsum sellae |
| 1 | Within suprasellar cistern (one cut above dorsum) |
| 2 | At level of third ventricle floor (one cut above suprasellar cistern) |
| 3 | Indenting and elevating floor or third ventricle (two or more cuts above suprasellar cistern) |

**Table 3.** Definition of vascular segments and aneurysmal dilatation.[7]

| Vascular segment | Definition of vascular segment | Definition of aneurysmal dilatation |
|---|---|---|
| Cavernous ICA | Entry point of ICA into cavernous segment to the ophthalmic artery origin | ≥8.5 mm |
| Supraclinoid ICA | ICA from ophthalmic artery origin to the ICA bifurcation | >8.0 mm |
| M1 segment of MCA | Origin of MCA to the MCA bifurcation | ≥5.0 mm |
| Cavernous ICA | Entry point of ICA into cavernous segment to the ophthalmic artery origin | ≥8.5 mm |

Abbreviations: ICA = internal carotid artery; MCA = middle cerebral artery.

reconstruction, which was suggestive of elongation, then used axial images to assess if these vessels compressed on the brain parenchyma as well. If the vessels were also tortuous, the artery was labelled as dolichoectasia. The degree of aneurysmal dilatation described by Brinjikji et al[7] includes cavernous ICA, supraclinoid ICA, and M1 segment of middle cerebral artery but does not include ACA (Table 3). In our index subject, the distal ACA branches showed abrupt dilatation, obvious lateral tortuosity, and compression of the brain parenchyma, suggestive of dolichoectasia. However, an apparent communication with adjacent cortical veins did not allow a confident diagnosis of dolichoectasia, and DSA was still required to exclude AVM or DAVF.

As highlighted in this case, CT angiography has limitations for diagnosis of dolichoectasia of the anterior circulation, as it cannot show the real-time blood flow. Thus, DSA will still be required in such cases as the dynamic component of DSA allows confident identification of the presence or absence of an abnormal early venous drainage that would be seen in AVM or DAVF but not in dolichoectasia.

Radiologists should consider this differential diagnosis and recommend DSA for more definite diagnosis.

## CONCLUSION

Dolichoectasia of the anterior cerebral circulation is a rare disease that may be difficult to differentiate from AVM or DAVF when based on CT radiological findings. This case report highlights the value of DSA in differentiating dolichoectasia from AVM or DAVF owing to its ability to capture the real-time blood flow properties of the diseased artery. Differentiating dolichoectasia from AVM or DAVF is important, to guide further investigations or treatment.

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