

Cerebral aneurysms associated with segmental dilative arteriopathy of the circle of Willis

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Abstract

Background: Dilative arteriopathy is a form of dolichoectasia. It is sometimes observed in the posterior circulation, and it may be associated with various type of stroke. Herein, we report two unusual cases of saccular aneurysms associated with a segmental dilative arteriopathy located in the anterior circulation.

Case Descriptions: The first case is a 39-year-old woman with irregular tortuosity and coiling of the left internal cerebral artery along with saccular aneurysms in this artery. The second case is a 45-year-old woman presenting with a ruptured saccular aneurysm in the coiling of the anterior cerebral artery. In both cases, the aneurysm was clipped successfully, and the patients recovered uneventfully with no neurological deficits.

Conclusion: Dilative arteriopathy of the circle of Willis is an unusual anomaly and is characterized by tortuous and elongated arteries, which are sometimes observed in patients with a congenital anomaly. This report describes two cases of saccular aneurysm associated with dilative arteriopathy of the circle of Willis with no medical history, which to the best of our knowledge has not previously been described.

Key Words: Anterior cerebral artery, arteriopathy, dolichoectasia, subarachnoid hemorrhage

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INTRODUCTION

Dolichoectasia is a broad concept that may include many diseases. Intracranial dolichoectasia is characterized by enlargement, tortuosity, and elongation of the major arteries of the brain and is typically observed in the posterior circulation.^[6] Various anomalies have been diagnosed as dolichoectasia, including fusiform aneurysms, coiling, and multiple vascular enlargements. In any case, dilation is the most striking feature;

therefore this condition is now referred to as dilative arteriopathy. Although dilative arteriopathy is frequently asymptomatic in children, it may be associated with stroke, subarachnoid hemorrhage, cranial nerve palsy, compression of the midbrain or obstructive hydrocephalus in adults.^[14,15,18] In this report, we describe two cases of saccular aneurysm associated with segmental dilative arteriopathy of the internal cerebral artery (ICA) or anterior cerebral artery (ACA), one of which was a ruptured aneurysm.

CASE REPORTS

Case 1

A 39-year-old woman with no remarkable medical or familial history underwent magnetic resonance (MR) imaging for headache. MR angiography and three-dimensional (3D) computed tomographic (CT) angiography revealed irregular tortuosity of the ICA from the ophthalmic artery to the terminal portion (C1, C2) on the left side and saccular aneurysms in the left C1. Cerebral angiography revealed that the ICA coiled twice between the ophthalmic artery and the anterior choroidal artery and contained four saccular aneurysms distal to the posterior communicating artery [Figure 1a]. All other intracranial vessels appeared completely normal.

The patient was taken for microsurgical exploration. The left ICA was dilated and coiled. During temporary occlusion of the ICA, the artery collapsed completely; no arterial dissection or atherosclerotic changes were observed [Figure 1c and e]. Accordingly, the aneurysm was successfully clipped at the neck [Figure 1d and f]. Postoperative angiogram revealed

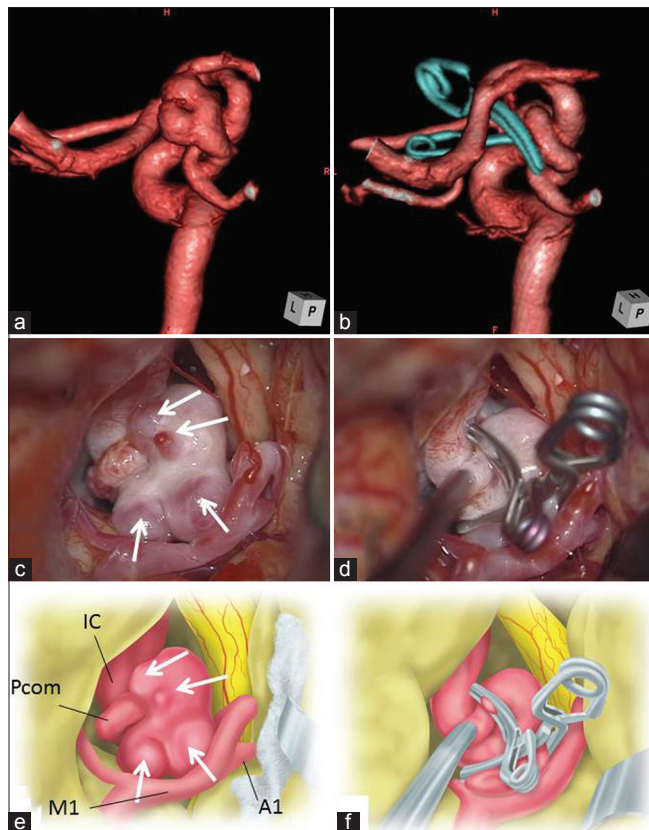


Figure 1: (a) Three-dimensional digital subtraction angiography (DSA) showing the elongated and tortuous left internal cerebral artery (ICA) with saccular aneurysms (white arrow). (b) Postoperative DSA showing successful clipping of these aneurysms. Intraoperative photographs (c and d) and illustrations. (e and f) Enlarged and tortuous left ICA with saccular aneurysms (arrow), and successful clipping of these aneurysms

obliteration of the aneurysmal neck and the absence of residual opacification [Figure 1b]. The patient recovered uneventfully and was discharged with no neurological deficits. Aneurysm has not recurred at the same site in the 5 years since the surgery.

Case 2

A 45-year-old woman with no remarkable medical or familial history suddenly developed a severe headache and nausea and was immediately transported to our hospital by ambulance. At the time of admission, she experienced mild consciousness disturbance but no other neurological deficits. She had no history of head trauma or connective tissue disorder. CT revealed diffuse subarachnoid hemorrhage, and 3D CT angiography and cerebral angiography revealed irregular tortuosity of the horizontal portion of the ACA (A1) on the left side and a saccular aneurysm in the left A1 segment [Figure 2a, c and d]. All other intracranial vessels appeared completely normal.

On day 1, the patient was taken to the operating room for microsurgical exploration, and a left frontotemporal craniotomy was performed. The dome of the aneurysm, including the rupture point, was buried in the frontal lobe. The left A1 was dilated, elongated, and tortuous [Figure 2e, f, and g]. The neck of the aneurysm was apparent, however, and the aneurysm was successfully clipped at the neck [Figure 2f and h]. Postoperative angiogram revealed complete clipping of the aneurysm [Figure 2b]. The patient recovered uneventfully and was discharged 22 days after surgery in an ambulatory condition with no neurological deficits. Aneurysm at the same site has not recurred in the 7 years since the surgery.

DISCUSSION

Several diseases have been implicated as potential causes of dilative arteriopathy, including both congenital and acquired conditions.^[2,6] From a pathological perspective, arterial dilation is caused by aberrant vascular remodeling,^[6] which is induced by the disruption of the internal elastic lamina. Breakdown and remodeling of the internal elastic lamina are controlled by early growth response protein 1 and matrix metalloprotease (MMP).^[16] Overexpression of MMPs damages the fibronectin mesh that smooth muscle cells used to travel within the vessel wall,^[10] and arterial dilation arises as a result. Morphologically, dilation of the arteries can develop according to two patterns. One is a nonsegmental pattern in which dilation occurs over a comparatively wide area.^[8] The other is a segmental pattern expressing geotropism, which might be related to focal dysfunction. Nonsegmental dilative arteriopathy represents the fragility of the entire arterial system when exposed to a specific trigger. This

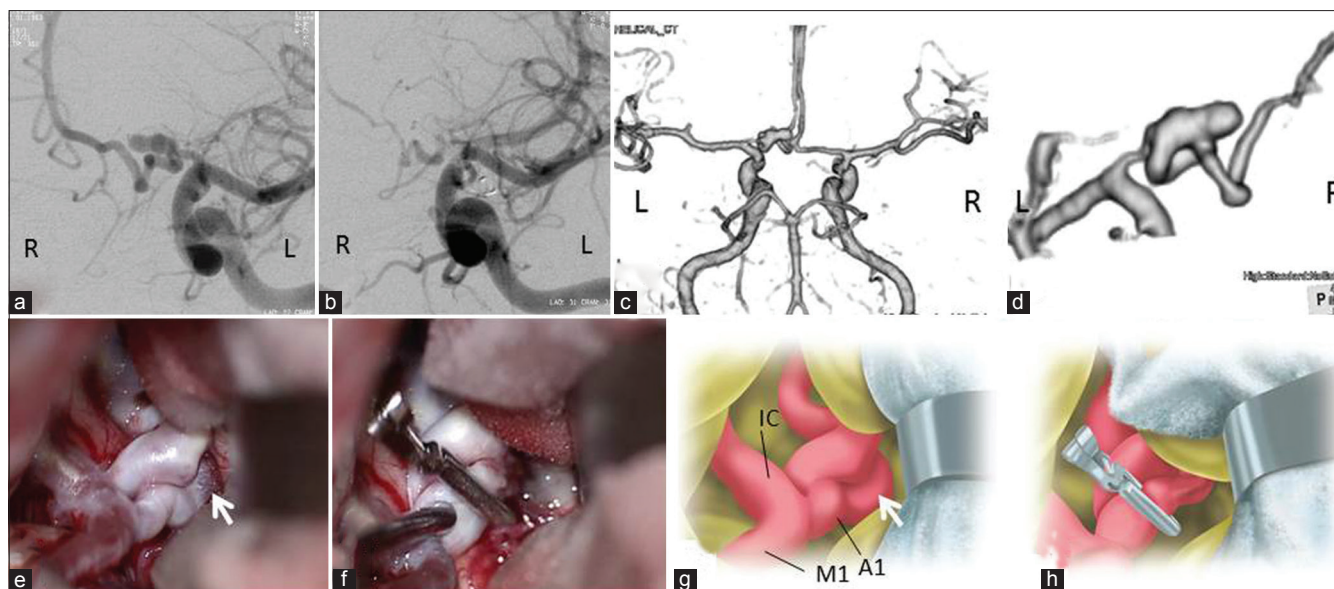


Figure 2: (a) Digital subtraction angiography (DSA) showing the elongated and tortuous left anterior cerebral artery (ACA) with saccular aneurysm (white arrow). (b) Postoperative DSA showing successful clipping of the aneurysm. Preoperative three-dimensional computed tomographic angiography (c and d), intraoperative photographs (e and f) and illustrations. (g and h) Enlarged and tortuous left ACA with saccular aneurysm (arrow), and successful clipping of the aneurysm

condition is found in patients with atherosclerosis, alpha-glucosidase deficiency, Marfan's syndrome, Ehlers–Danlos syndrome, Loyaes–Dietz syndrome, AIDS, Fabry's disease, and sickle cell disease.^[4,6,9,11] Although several genetic disorders are included in this category, our two cases were negative for these genetic diseases. Therefore, further genetic investigation was conducted. Segmental dilative arteriopathy, in contrast, is related to the segmental identity of the vessel and its associated segmental vulnerability. This condition is sometimes found in patients with PHACES syndrome or moyamoya disease.^[2,17] Cerebral vasculopathy in PHACES syndrome chiefly comprises arterial anomalies, with the arteries of the circle of Willis most commonly involved.^[7] Segmental dilative arteriopathy can also derive from embryological factors: Focal defects and destruction of the internal elastic lamina have been implicated in its pathogenesis.^[6,13] Lasjaunias advocated for the identification of seven segments within the ICA,^[8] namely, (1) cervical, (2) ascending petrous, (3) horizontal petrous, (4) ascending cavernous, (5) horizontal cavernous, (6) clinoid, and (7) terminal. Segmental arteriopathy is induced primarily through congenital dysgenesis of one ICA segment. Based on this theory, the carotid rete mirabile might be also subject to segmental arteriopathy. Anomaly of the terminal portion of the ICA or the ACA is extremely rare.^[3,5] In one of our two cases, vascular abnormality was limited to the region between the anterior choroidal artery and the posterior communicating artery. In the other case, vascular abnormality was limited to segment A1. These cases provide support for the segmental arteriopathy hypothesis.

Although associated saccular aneurysms are typically induced by hemodynamic stress due to dilative arteriopathy, most aneurysms associated with dolichoectasia are dissecting aneurysms or fusiform aneurysms.^[12] As for the causes of our cases, hemodynamic stress resulting from the unique flow associated with vulnerable vessels might be involved in addition to congenital factors, given the relatively broad-based lesions from irregular segments. Careful long-term observation would be required to confirm this. The aneurysms in our cases were saccular aneurysms that were confirmed perioperatively and successfully clipped. Aneurysm clipping will certainly not be possible in all cases, however, for such cases, flow alteration treatment using a bypass graft should also be available as an alternate strategy.^[1] With either treatment method, careful long-term observation is required, especially as a better understanding of these arteriopathies may improve our ability to predict stroke in affected patients.

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