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Case Report

Long segment dolichoectasia of the right internal carotid artery diagnosed by CT angiography

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ABSTRACT

Dolichoectasia of the cerebral artery, a rare disorder of arterial dilatation, elongation, and tortuosity, most frequently involves the vertebrobasilar system in elderly patients with hypertension and is associated with the development of atherosclerosis in the aging process. Dolichoectasia also can be seen in the carotid system, but it is usually seen at the intracranial short segment of the internal carotid artery (ICA) or anterior cerebral artery. We present a case of dolichoectasia of the long segment of the right internal carotid artery that extended from the upper cervical to terminal segment of the vessel that was diagnosed by computed tomography angiography in a normotensive 66-year-old woman with transient ischemic attack.

Introduction

Ectasia with elongation of the cerebral arteries is rare and most frequently involves the basilar artery, followed by the vertebral artery. Generally described as either "dolichoectasia of the basilar artery" or "megadolicho basilar artery," we use the term "dolichoectasia" in this paper. The condition can involve the anterior circulation, such as the intracranial segment of the internal carotid artery (ICA) and the anterior cerebral artery [1–6], and is most commonly observed in patients who are elderly and those with hypertension.

We report an extremely rare case of dolichoectasia of the long segment of the ICA extending from the upper cervical to terminal segment of the vessel that was diagnosed by computed tomography (CT) angiography in a

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Ethical standards: We declare that all human and animal studies have been approved by the Melbourne Health Research Ethics Committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

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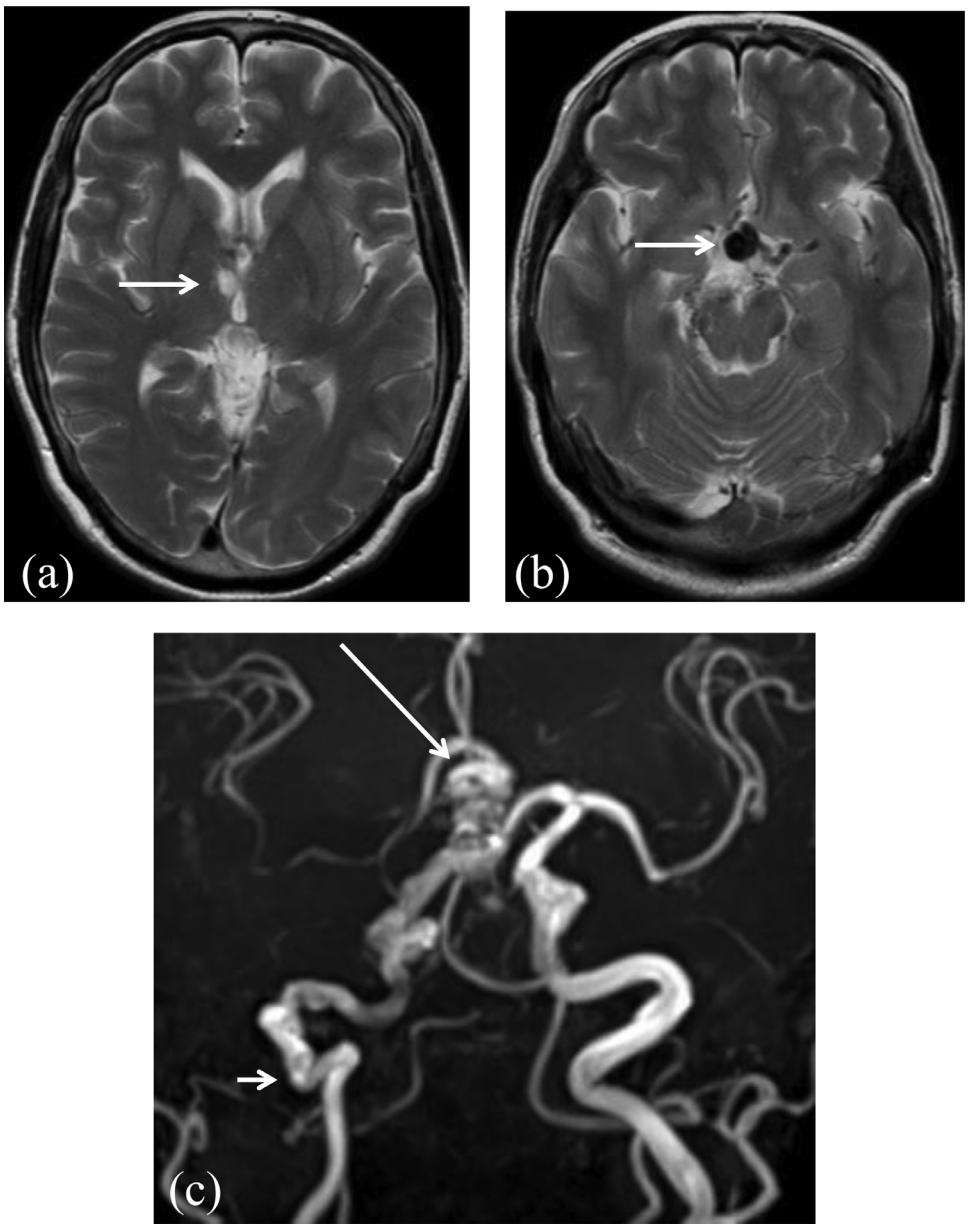

Fig. 1 – T2-weighted magnetic resonance (MR) imaging shows a small infarction (arrow) at the right thalamus (a) and an abnormal flow void (arrow) in the suprasellar cistern, suggestive of an aneurysm (b). Anteroposterior projection of MR angiography (c) shows abnormal configuration of the right internal carotid artery (ICA) from its upper cervical segment (short arrow) to the terminal segment (long arrow). The left ICA is dilated mildly.

66-year-old normotensive woman with transient ischemic attack (TIA).

Case report

A 66-year-old normotensive woman with transient hyperesthesia of the left hand and left face, indicative of TIA, visited a general hospital, where she underwent cerebral magnetic resonance (MR) imaging and MR angiography using a 1.5-tesla machine. MR imaging showed a small infarction in the anterior thalamus and a signal void in the suprasellar cistern (Fig. 1a and b). Simultaneously obtained MR angiography showed abnormal configuration of the long segment of the right ICA (Fig. 1c) and mild dilatation of the left ICA.

The patient was transferred to our institution for further evaluation and possible treatment. CT angiography showed extreme tortuosity and mild dilatation of the right ICA from its upper cervical to terminal segment (Fig. 2a and b) as well as mild dilatation of the left ICA. CT angiographic source images of the suprasellar region showed dilatation of the right ICA with calcification of the vessel wall (Fig. 2c and d). Single photon emission CT (SPECT) using N-isopropyl-p-[^{123}I]-

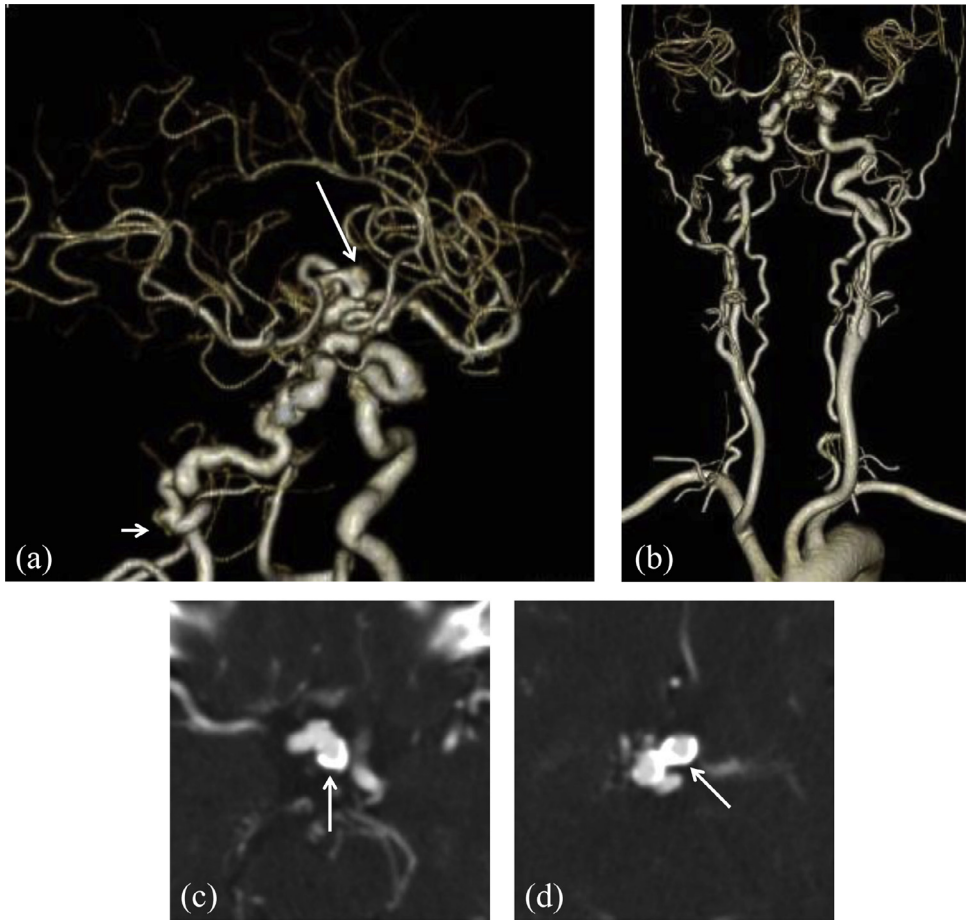

Fig. 2 – Right anterior oblique projection of computed tomography (CT) angiography (a) shows extremely tortuous and mildly dilated right internal carotid artery (ICA) from its upper cervical segment (short arrow) to the terminal segment (long arrow). The left ICA is also dilated mildly. Anteroposterior projection of CT angiography from the aortic arch to the intracranial region (b) shows no abnormality except for that described above. CT angiographic source images at the level of the suprasellar cistern (c, d) show a tortuous and dilated right ICA with calcified arterial wall (arrows).

iodoamphetamine showed mildly decreased blood perfusion in the right cerebral hemisphere (not shown).

The patient was treated conservatively for her mild symptoms and did not require surgery. She was sent back to the previous general hospital, experienced an uneventful clinical course for 5 years, and underwent no further angiography during this period in our institution.

Discussion

Dolichoectasia is characterized by dilatation, elongation, and tortuosity of the cranial arteries. Because dilatation seems its most important feature, the condition is commonly described as "dilatative arteriopathy" [3]. However, there are no generally accepted quantitative criteria for dilatative arteriopathy. Extremely dilated and elongated arteries are referred as "(giant) fusiform aneurysms." Our patient demonstrated mild dilatation as well as tortuosity from the upper cervical segment of the right ICA to its terminal segment on both MR angiography and CT angiography. CT angiographic source images also showed calcification of the dilated arterial wall. Thus, we diagnosed dolichoectasia of an extremely long segment of the right ICA.

Dolichoectasia is most frequently seen in the vertebrobasilar system, followed by the anterior circulation, especially the anterior cerebral artery and its branches [2], and the condition can affect the vertebrobasilar system and anterior circulation simultaneously [1,3–6]. Most patients are aged and hypertensive. Multiple pathophysiological processes, including prolonged hypertension of the arterial system, might contribute to its development. Histological studies support the hypothesis of underlying degeneration of the internal elastic lamina and thinning of the media secondary to smooth muscle atrophy [5]. However, though our patient was older, she was normotensive and her other major cerebral arteries appeared normal apart from the extreme length of the affected segment, which extended from the extracranial to terminal segment of the right ICA. Thus, we considered congenital weakness of the arterial wall a possible mechanism underlying the formation of dolichoectasia in our patient [7]. In children and

young adults without hypertension, dolichoectasia can be regarded as a type of congenital vascular malformation [8].

Most cases of dolichoectasia are asymptomatic, but the various clinical symptoms that do arise are attributable to 3 mechanisms–compression, rupture, and ischemia [5]. Symptoms related to the mass effect of dolichoectasia depend on the affected artery and grade of both dilatation and elongation and are most commonly attributable to compression of the surrounding neural structure. Our patient experienced a TIA and small infarction in the region associated with the abnormal right ICA. Sadahiro and associates [9] reported a case of repeated cerebral ischemia caused by dolichoectasia of the common carotid artery.

In a recent study of patients with dolichoectasia of the intracranial distal ICA, Jia's group [10] observed PHACE, a syndrome comprising posterior fossa malformation, hemangioma, arterial anomalies, cardiac defects, and eye abnormalities, in 11 of 20 patients. Our patient demonstrated only extreme elongation of the affected segment and can therefore be regarded as a typical case of dolichoectasia of the ICA without PHACE syndrome.

Conclusions

We report an extremely rare case of dolichoectasia of the long segment of the ICA that extended from the upper cervical to terminal segment of the vessel in a normotensive woman, whose long lesion suggests the condition's origin as congenital weakness of the arterial wall.

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