Case Report

Dolichoectasia of the right internal carotid artery diagnosed incidentally by MR angiography in a 17-year-old girl

Akira Uchino MD, PhD\textsuperscript{a,*}, Masayuki Ohira MD, PhD\textsuperscript{b}

\textsuperscript{a}Department of Diagnostic Radiology, Saitama Medical University International Medical Center, 1397-1 Yamane, Hidaka, Saitama 350-1298, Japan

\textsuperscript{b}Department of Neurology and Cerebrovascular Medicine, Saitama Medical University International Medical Center, Hidaka, Saitama, Japan

\textbf{A R T I C L E I N F O}

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\textbf{A B S T R A C T}

Dolichoectasia of the cerebral artery, a rare disorder of arterial elongation, dilatation, and tortuosity, usually involves the vertebrobasilar system in elderly patients with hypertension and is associated with the development of atherosclerosis in the aging process. We present a very rare case in a 17-year-old girl with multiple sclerosis of dolichoectasia of the supraclinoid segment of the internal carotid artery that was found incidentally on magnetic resonance angiography and seems to represent a congenital arterial malformation.

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\textbf{Introduction}

Ectasia of the cerebral arteries with elongation is rare, most commonly observed in patients who are elderly and those with hypertension, frequently involves the basilar artery followed by the vertebral artery, a condition described as “dolichoectasia of the basilar artery” or “megadolicho basilar artery,” and can involve the anterior circulation, such as the internal carotid artery (ICA) and anterior cerebral artery [1-8].

In this paper, we use the term “dolichoectasia.”

We report an extremely rare case of dolichoectasia of the ICA found incidentally on magnetic resonance (MR) angiography in a 17-year-old girl with multiple sclerosis. Few such pediatric cases are reported [3,8-10].

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* Corresponding author.

E-mail address: auchino@saitama-med.ac.jp (A. Uchino MD, PhD).

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Case report

A 17-year-old girl with multiple sclerosis underwent cerebral MR imaging and MR angiography using a 3-tesla machine for evaluation of recently experienced right hemiparesis. Her history included left hemiparesis 3 years previously. Neurologic examinations on admission revealed only mild right hemiparesis and muscle weakness. Laboratory data including cerebrospinal fluid were unremarkable. MR imaging showed demyelinating lesions from the posterior limb of the internal capsule to the corona radiata, bilaterally (Fig. 1), involvement of the white matter of the right occipital lobe, and a dilated and tortuous vessel at the right side of the suprasellar cistern (Fig. 2). Subsequent MR angiography demonstrated a mildly dilated and markedly elongated and tortuous supraclinoid segment of the right ICA without aneurysm (Figs. 3, 4). We therefore diagnosed dolichoectasia of the right ICA and performed no further angiography because she demonstrated no symptoms related to this abnormality.

Intravenous treatment with high-dose corticosteroids improved her symptoms of mild right hemiparesis and resolved her muscle weakness, and no further MR angiography was performed.

Fig. 1 – Magnetic resonance (MR) imaging at the level of body of the lateral ventricle. (a) Diffusion-weighted image (DWI) shows hyperintense lesion at the left corona radiata (long arrow). (b) Fluid-attenuated inversion recovery (FLAIR) image shows hyperintense lesions, bilaterally (long and short arrows). Thus, the lesion on the left is regarded as acute demyelinating focus and that on the right, chronic stage demyelination, compatible with multiple sclerosis.

Fig. 2 – T2-weighted magnetic resonance images at the level of the suprasellar cistern show an abnormal flow void in the right side, which suggests aneurysm or ectasia of the right internal carotid artery. Both the right optic tract and right medial temporal lobe are mildly compressed.
Fig. 3 – Anteroposterior (a) and right lateral (b) projections of magnetic resonance angiography show a mildly dilated and extremely long and tortuous supraclinoid segment of the right internal carotid artery.

Fig. 4 – Anteroposterior (a) and right lateral (b) projections of volume-rendering images of magnetic resonance angiography of the right carotid system demonstrate the lesion more clearly. The right posterior cerebral artery is fetal type.

Discussion

Dolichoectasia is usually seen in the vertebrobasilar system, can affect the anterior circulation, especially the anterior cerebral artery and its branches [3,8], and can affect both the vertebrobasilar system and anterior circulation simultaneously [2,4-7]. Most patients are aged and hypertensive. Multiple pathophysiological processes, including prolonged hypertension of the arterial system, might contribute to its development. Histologic studies support the hypothesis of underlying degeneration of the internal elastic lamina and thinning of the media secondary to smooth muscle atrophy [6]. However, in our young patient, congenital weakness of the arterial wall might have contributed to the formation of dolichoectasia [11,12]. Baccin et al. [10] reported segmental dolichoectasia of the intracranial arteries in 2 children with PHACE syndrome (P, posterior fossa malformations; H, hemangioma; A, arterial anomalies; C, coarctation of the aorta and cardiac anomalies; E, eye (ocular) anomalies), who represent the few pediatric patients with dolichoectasia reported in the English-language literature [3,8,9]. In children and young adults without hypertension, dolichoectasia can be regarded as a type of congenital vascular malformation. Sako et al. [13] reported a case in a young adult of an extremely tortuous posterior inferior cerebellar artery that was diagnosed as a pure arterial malformation.

Most cases of dolichoectasia, like ours, are asymptomatic, but the various clinical symptoms that do arise are attributable to 3 mechanisms – compression, rupture, and ischemia [9]. Symptoms related to the mass effect of dolichoectasia depend on the affected artery and grade of
both dilatation and elongation. The most common cause of symptoms is compression of the surrounding neural structure. In our patient, any increase in lesion diameter in time may injure the right optic tract and right oculomotor nerve. Hudson and Gonsalves [1] reported a case of unilateral ventricular obstruction at the foramen of Monro by dolichoectasia of the contralateral ICA. Patients with multiple dolichoectasia tend to be symptomatic [5,7].

Conclusions

We report an extremely rare case of dolichoectatic ICA in a 17-year-old girl with multiple sclerosis, whose age suggests the condition’s origin as congenital arterial malformation.

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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