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Case Report
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Incidental Internal Carotid Dolichoectasia: Case Report

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Abstract

Cerebral arterial dolichoectasia is an uncommon vascular disorder, which is mostly diagnosed incidentally. However, some patients present with deleterious vascular events or various clinical pictures of mass effect resulting from pressure by the large and tortuous artery. There is still no consensus guideline for the treatment or follow up of these patients, especially those who are asymptomatic. This angiopathy mostly affects the vertebrobasilar system and the carotid involvement is therefore, under represented and less addressed in the literature. Here, we present an incidental finding of dolichoectasia of the supraclinoid portion of the right internal carotid in a 60 year old woman with chronic hypertension, diagnosed by MR angiography.

Keywords: Dolichoectasia; Internal carotid; Vasculopathy; MR angiography

Introduction

Dolichoectasia is an uncommon but well recognized form of cerebrovasculopathy, characterized by irreversible dilation, elongation, and tortuosity of the cranial arteries [1]. Vertebrobasilar system seems to be particularly more vulnerable to this angiopathy, although rare cases with the dolichoectatic vessels in the anterior circulation have also been described [2]. Patients may present with mass effect or vascular accidents. However, most cases are detected incidentally [2,3]. It is believed that arterial hypertension with or without atherosclerosis is a major contributing factor to the formation and enlargement of the dolichoectatic vessels [4]. Herewith, we present an incidental finding of dolichoectasia in the supraclinoid portion of the Internal Carotid Artery (ICA) in a 60 year old woman with hypertension and hyperlipidemia.

Case Presentation

A 60-year-old woman with a history of arterial hypertension and hypercholesterolemia presented to the emergency department of our hospital with headache lasting for one day. Her blood pressure at admission was 178/92 mmHg. The headache was generalized with moderate severity and was relieved by controlling the blood pressure. She had no previous history of cerebrovascular or coronary accidents. Her neural examination was normal. An urgent brain non-contrast Computed Tomography (CT) scan revealed mural calcification of the supraclinoid portion of the right ICA with increased diameter and high density. There was a hypo density suspicious of thrombosis (Figure 1). Therefore, the patient was further evaluated by Magnetic Resonance Arteriography (MRA). Thrombosis was ruled out and the incidental evidence of dolichoectasia at the same segment was proven in MRA. There were no other vascular or any neural pathologic features detected on brain images (Figure 2). The patient was discharged home and a systematic follow up was advised.

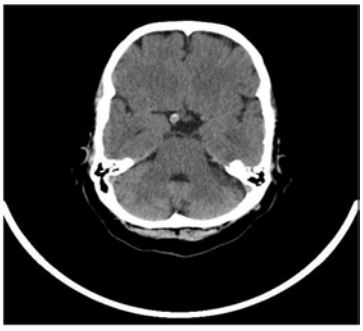

Figure 1: Brain computed tomography without contrast.

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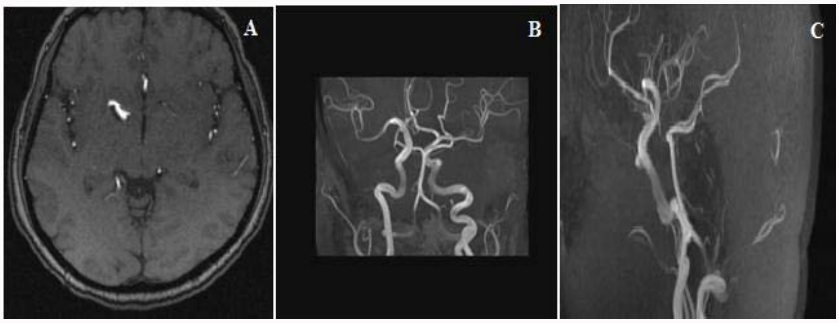

Figure 2: A) Brain magnetic resonance angiography. B) Evidence of dolichoectasia of supra clinoid portion of right side ICA is noted as incidental finding. C) A1 segment of the right anterior cerebral artery is hypoplastic (normal variation).

Discussion

Dolichoectasia, a Greek term meaning long "dolichos" and dilate "ectasis", is a rare vascular disorder of a not well-known pathophysiology. The prevalence is estimated about 0.08% to 6.5% in the general population, while the reported prevalence in series of patients with stroke ranges from 3% to 17% [1,5]. As in our patient, this arteriopathy is most commonly observed as an incidental finding in elderly patients with hypertension [4,6]. It is believed that continued shear stress on the arterial walls in the setting of prolonged systemic hypertension is associated with pathological remodeling of the vessel wall, leading to the dilative arteriopathy [7,8]. In support of this, histologic studies show degeneration of the internal elastic lamina as well as atrophic media secondary to the migration of smooth muscle cells as the key pathologic features of the dolichoectatic arteries [8-10]. Hypercholesterolemia can also traumatize and weaken the vessel wall, resulting in this dilative vasculopathy. Several other predisposing factors to the dilative arteriopathy are also described in association with dolichoectasia. Connective tissue disorders, congenital vascular malformations, inherited metabolic disorders, and inflammatory processes are shown to have higher risk of intracranial dolichoectasia [11-18]. Overall, multiple pathological processes might lead to the development of this angiopathy. However, the diagnosis of dolichoectasia in this patient most likely could be attributed to the systemic hypertensive vasculopathy [4]. As in our patient, most cases of dolichoectasia have been diagnosed incidentally [2,19]. However, this condition could manifest by compressive symptoms or acute vascular events, either hemorrhage or ischemia [20]. Because of lower available space in the territory of anterior circulation, dolichoectasia in the carotid tree has a higher chance than the vertebrobasilar system to be symptomatic [2,10]. Isolated II, III or VI nerve palsies have been reported as a result of direct compression by the tortuous, elongated, and ecstatic internal carotid artery [10,21,22]. Dolichoectasia more commonly causes cerebral ischemic infarction by thrombosis formation, embolism, or simply by occluding deep penetrating arteries [23,24]. In fact, lacunar strokes are shown to be more prevalent in those with dolichoectasia [23,25]. The rupture of affected vessel is less likely, but can result in deleterious intracranial or subarachnoid hemorrhage [10,26,27]. Angiography is the gold standard modality to diagnose cerebral vascular pathologies such as dolichoectasia [28]. However, the advent of new technologies such as MRA has reduced the need for invasive procedures in many clinical settings [2,8,29]. In this patient, brain CT scan was equivocal regarding the presence of thrombosis in the supraclinoid portion of the right ICA. However, MRA excluded the thrombotic formation and the headache was attributed to the transient rise of the hypertension, as this was resolved by controlling the blood pressure. This pathology preferentially affects the posterior cranial circulation and thus, vertebrobasilar dolichoectasia is more addressed in the existing literature [2]. Smoker's criteria are used to diagnose the vertebrobasilar dolichoectasia [8]. However, no validated diagnostic criteria is yet available for the anterior circulation and rather the diagnosis is made visually by larger than normal diameter of the arteries and the presence of tortuosity compared with expected values or the contralateral artery [2,30]. There is no consensus guideline for the treatment or even follow up of dolichoectasia in asymptomatic patients [2,6]. Therefore, more research is needed to better understand and manage this rare angiopathy, which may result in unfavorable outcomes with high morbidity and mortality. This is challenged by the rarity of this condition and thus, case presentations are of great value to help with clinical management of asymptomatic patients.

Conclusion

The course of dolichoectasia in the anterior circulation has been presented in a few case reports or small case series and almost always in symptomatic patients. More research is needed, especially in asymptomatic patients to confederate a guideline for the clinical management of this rare but important angiopathy.

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