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Case Report

Bilateral congenital absence of the internal carotid arteries: a case report

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ABSTRACT

Congenital absence of the internal carotid artery is a rare occurrence. Even more infrequent are cases where the patient has a bilateral absence of the internal carotid arteries. Reported is a case of a 52-year-old woman who presented with optic nerve neuropathy, and was incidentally discovered to have a congenital bilateral absence of her internal carotid arteries. During computed tomography angiography imaging looking for cerebral venous thrombosis, related to her preexisting condition of bilateral elevated optic discs and residual left optic neuropathy, the findings were made. The absence of the arteries is not always recognizably symptomatic, with most findings being incidental through imaging studies only. This is because collateral flow allows for sufficient cerebral circulation. However, this condition puts such patients at higher risk for conditions such as aneurysms and subsequently strokes where the collateral flow exists.

Introduction

Bilateral congenital absence of the internal carotid arteries (ICA) is an uncommon condition, impacting less than 0.01% of the general population [1,2,3,4]. Absence of the ICA most often presents unilaterally in patients, thereby making instances of bilateral hypoplasia even more so intriguing [3]. This natural absence may be consequent of hypoplasia (incomplete development), agenesis (no development), or aplasia (no development despite the presence of developmental precursors). The result of this absence is collateral blood flow, typically from the circle of Willis; for this reason, this abnormality is oftentimes asymptomatic and detected incidentally via imaging such as computed tomography (CT), angiography, or magnetic resonance imaging (MRI) [2,3]. However, symptoms that may present in some patients include recurrent headache, blurred vision, and convulsions [4]. This collateral flow may also occur through persistent embryonic vessels or through anastomotic branches of the external carotid artery (ECA) [2]. If there is a lack of adequate collateral flow, cerebrovascular accidents (CVA) and/or intracranial hemorrhage could occur and thereby this issue may present as cerebrovascular disease [5,6]. The presented case involves a 52-year-old female

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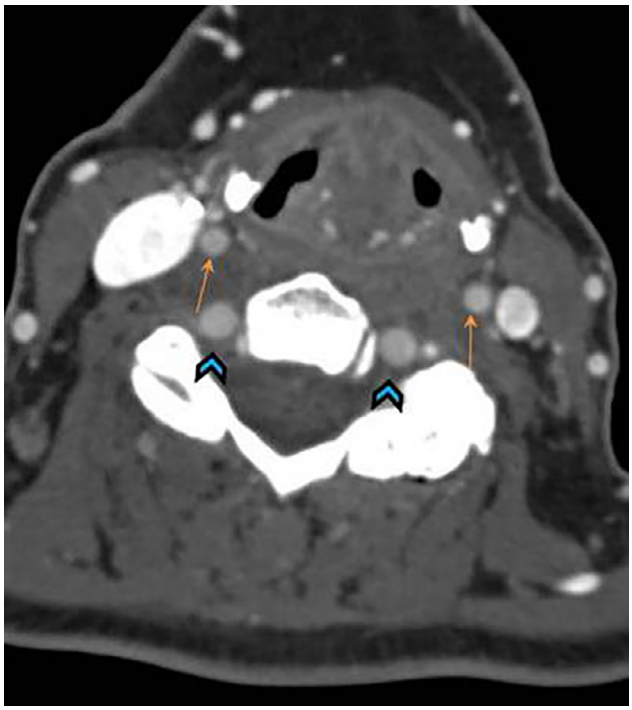

Fig. 1 – Agenesis of the bilateral internal carotid arteries. CT angiogram head/neck at the level of the C5 vertebral body, where the internal carotid artery (ICA) is absent. The common carotid arteries continue as the external carotid arteries (orange arrows). Neither intracervical nor intracranial portions of the ICAs were identified. Enlarged bilateral vertebral arteries were identified (blue arrowheads). (Color version of figure is available online.)

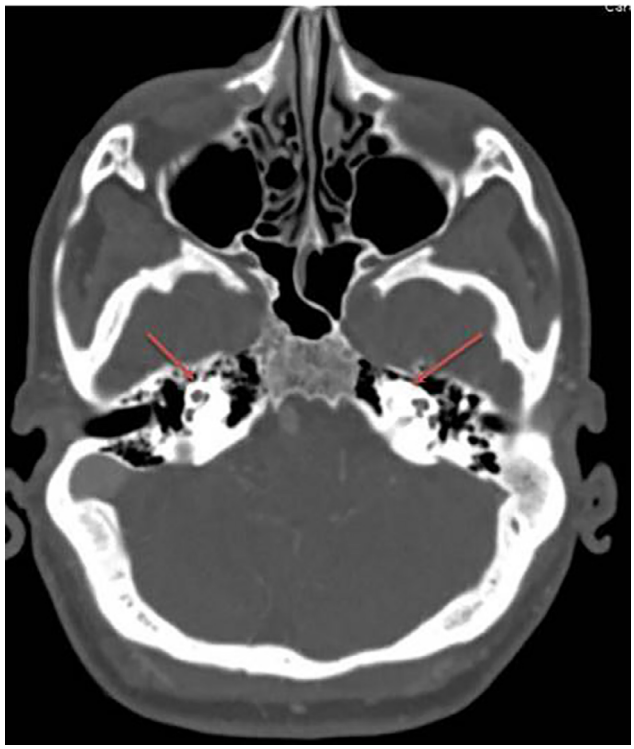

Fig. 2 – Empty petrous carotid canals. Axial CT angiogram at the level of the petrous portions of the carotid canals demonstrates absence of carotid canals within the expected portions of temporal bones on both sides (red arrows). The finding favors absence of internal carotid arteries (ICA). (Color version of figure is available online.)

patient with an incidental finding of bilateral absence of the ICAs.

Case report

A 52-year-old female presented for a routine follow-up appointment related to her bilateral elevated optic discs and residual left optic neuropathy. She had constant headache, especially when laying down flat. A computed tomography angiography (CTA) of her head and neck was done (see Figs. 1–5). While inspecting the major branches of the aorta in the neck, the right common carotid originated from the brachiocephalic trunk and the left common carotid artery arose from the arch of the aorta. However, there was no bifurcation of the common carotid arteries at the expected level superior to the thyroid cartilage (see Fig. 1). The vessels identified in this area were determined to be the external carotid arteries as they followed the appropriate anatomical course and had the expected branches in the neck with no intracranial extension. The expected course of the intracervical and intracranial portions of the ICA failed to demonstrate any visible vessels (see Fig. 2). Additionally, petrous carotid canals were not identified on either side. These findings were representative of bilateral absence of the ICAs. The CTA head revealed patent anterior cerebral circulation, which was found to be from compensatory enlarged feeding posterior communicating arteries (PCOM), prominent basilar artery, posterior cerebral arteries (PCA) and codominant enlarged vertebral arteries (see Figs. 3–6). This patient also had hypoplastic left internal jugular vein, sigmoid, and transverse sinuses.

Discussion

Absences of carotid arteries have been documented as early as 1787 per postmortem examination, with a case in a living patient as early as 1954 via angiography [1,2]. Interestingly, the most documented cases involving absence of the ICA are unilateral and most commonly reported on the left side [1,7]. Our case report is of particular interest because it demonstrates bilateral absence of the vessels. The cause of congenital unilateral carotid absence has been attributed to disruption of the embryo by physical and hemodynamic pressure upon the embryo [2]. Such stresses are thought to include amniotic band constriction and folding of the embryo's neck region to one