Case Report

Carotid Artery Hypoplasia Incidentally Found in Adult Patient: Case Report

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Congenital hypoplasia of the internal carotid artery (ICA) is a rare congenital anomaly. It is usually found incidentally during radiological examination for other reasons. Most patients are usually asymptomatic but a few of them may present with headache, Horner's syndrome, or transient ischemic attack (TIA). We presented herein an asymptomatic case of left carotid artery hypoplasia found on magnetic resonance imaging (MRI) during routine checkup. MRI with magnetic resonance angiography (MRA) showed severe hypoplasia of the left ICA from carotid bifurcation to petrous segment with collateral flow from the enlarged posterior communicating artery (PCOM) to supply the left middle cerebral artery (MCA). Recognition of this anomaly is very important when planning carotid endarterectomy or transsphenoidal hypophyseal surgery, even in case of acute brain attack from thromboembolic causes.

Keywords: *Carotid artery, Internal, Congenital anomalies, Magnetic resonance imaging, Agenesis, Hypoplasia*

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Congenital absence of the internal carotid artery (ICA) is a rare congenital anomaly occurring in less than 0.01% of the population[1,2]. The term "absence" is referred to a spectrum of developmental abnormalities including agenesis, aplasia, and hypoplasia of the ICA. Agenesis and aplasia represent total absence of the artery. Hypoplasia is characterized by ICA narrowing along its entire course because of incomplete development. In such cases, the anterior cerebral artery (ACA) and middle cerebral artery (MCA) on the side of the absence are usually supplied by the contralateral ICA through the circle of Willis or posterior circulation through the posterior communicating artery (PCOM)[2]. Patients are usually asymptomatic but a few of them may present with headache[3], Horner's syndrome[4,5], hormonal dysfunction[6], or transient ischemic attack (TIA)[2]. We reported a case of left carotid artery hypoplasia in asymptomatic case diagnosed on magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) during routine checkup.

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A 76-year-old woman with underlying well-controlled hypertension underwent first time MRI of the brain and MRA during routine checkup. She denied any abnormal neurological symptoms. The MRI and MRA of the brain showed diminished flow-related enhancement of the left petrous ICA (Fig. 1A) and collateral flow to the left cerebral hemisphere through the circle of Willis via a patent anterior communicating artery (ACOM) (Fig. 1B). No perceivable flow-related enhancement within the left supraclinoid ICA on the compressed image was noted. Hypoplasia of the left carotid canal was apparently shown on T1W image (Fig. 1C), which confirmed the congenital anomaly of the small-sized left ICA. The MRA of the neck also showed diffused small size of the left common carotid artery (CCA) and very small size of the entire left ICA from its origin up to petrous part (Fig. 1D).

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Discussion

Agenesis, aplasia, and hypoplasia of the ICA are rare congenital anomalies occurring in less than 0.01% of the population[1,2]. The term "absence" is referred to a spectrum of agenesis, aplasia, and hypoplasia of the ICA. Agenesis of ICA is caused by total nondevelopment of ICA, whereas aplasia and hypoplasia of ICA are respectively nondevelopment or incomplete development of the ICA despite the presence of embryonic precursor of the vessel[1,2,7]. The exact mechanisms of these developmental anomalies remain unknown. Therefore, agenesis and aplasia represent total absence of the artery, unlike hypoplasia, which is characterized by diffuse ICA narrowing along its entire course. Unilateral ICA abnormalities are distinctly more common than bilateral ICA agenesis or hypoplasia[2]. In these ICA abnormalities, there are

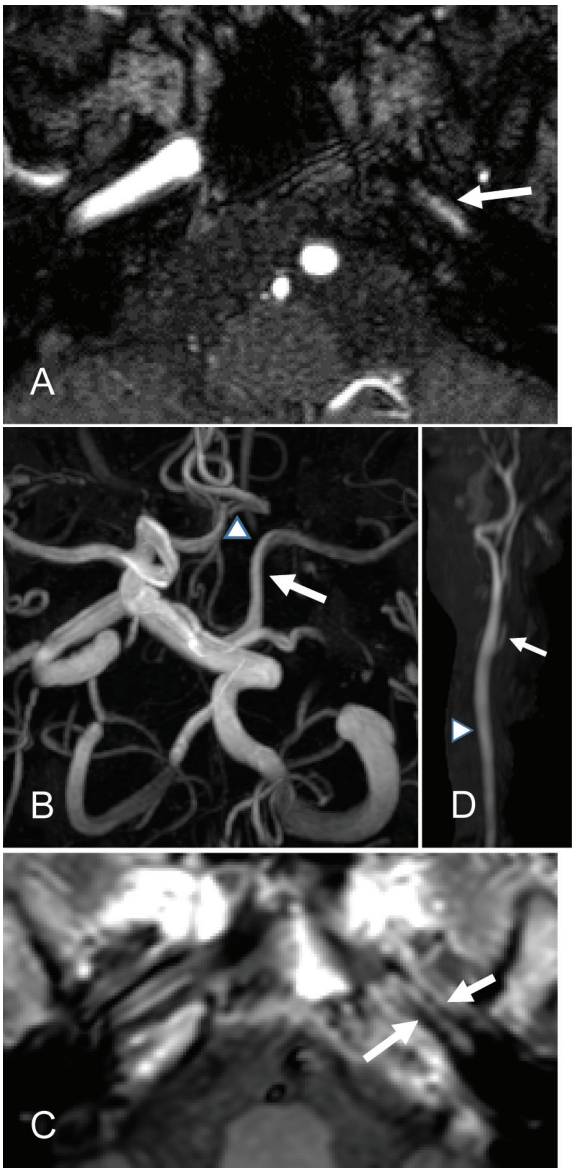

Fig. 1 Hypoplasia of the left ICA. (A) Source image from a 3D time-of-flight (TOF) MRA shows diminished flow-related enhancement within the petrous part of the left ICA (arrow). (B) Compressed image from the 3D TOF MRA shows an enlarged left PCOM (arrow) extending forward to supply the left MCA. The left ACA is supplied via a patent ACOM (arrowhead). There is no perceivable flow-related enhancement within the left supraclinoid ICA on this compressed image. (C) Axial T1W image at the level of the petrous ICA shows hypoplasia of the left carotid canal (arrows). (D) 3D TOF MRA of the neck shows diffusely diminished flow-related enhancement within the left ICA (arrow) and diffuse small size of the left CCA (arrowhead).

three major collateral pathways to supply the anterior cerebral circulation. The most common collateral pathway is through the enlarged PCOM to supply the MCA, as of this patient. The ACA on the side of ICA abnormalities are usually supplied by the contralateral ICA through the circle of Willis via a patent ACOM[2,5,8]. Other rare collateral pathways may be an anastomosis between the external carotid artery (ECA) and ICA at the skull base or an intercavernous anastomosis with the contralateral ICA(2,8,9). Agenesis of ICA may be misdiagnosed as ICA occlusion. To distinguish ICA agenesis from acquired ICA occlusion, computed tomography (CT) scan of the skull base should be performed focusing on the ipsilateral carotid canal[10]. This is the carotid canal normally develops in the setting of presence of embryonic ICA at five to six weeks of gestation. Therefore, demonstration of an absent or small carotid canal indicates a congenital absence, and helps differentiate from acquired causes of ICA occlusion or narrowing, such as chronic dissection, severe atherosclerosis, or fibromuscular dysplasia[2,10,11]. Moreover, the presence of ipsilateral CCA hypoplasia suggests the diagnosis of congenital absence of the ICA[11].

Although many cases of congenital absence of the ICA remain asymptomatic due to the presence of collateral circulation, these patients may present with neurological symptoms later in life due to cerebrovascular insufficiency. Congenital absence of the ICA has a high prevalence of intracranial aneurysms of 24% to 34%, compared to a 2% to 4% prevalence in the general population[2,7]. The aneurysms are possibly related to altered flow dynamics and increased flow through the collateral vessels along with congenital defects of the vessel wall, as the ACOM is the most common site of associated aneurysm. Owing to increased risk of aneurysm, clinical and radiologic surveillance in these patients has been indicated[9]. The patients with congenital absence of the ICA may present with migraine-like headache[3,8], rarely congenital Horner's syndrome[4,5], hormonal dysfunction[6], TIA[2], or symptoms related to pressure effects from enlarged collateral vessels. Recognition of congenital absence of the carotid artery is important when planning carotid endarterectomy, as both cerebral hemispheres may be dependent upon a single critical atheromatous carotid artery. In addition, the recognition of this condition is important in the setting of thromboembolic disease, as embolism from atherosclerotic disease in the contralateral carotid artery or vertebrobasilar system can occur. Consideration of this anomaly may help prevent the missed diagnosis of carotid dissection or

high-grade carotid stenosis, as the imaging findings of ICA hypoplasia can mimic radiographic string sign in carotid dissection or severe carotid stenosis[11,12]. Finally, failure to recognize the intercavernous collateral pathway can have serious complications during transsphenoidal hypophyseal surgery[8].

Conclusion

Agenesis, aplasia, and hypoplasia of the ICA are rare congenital anomalies. The major collateral pathways include the circle of Willis, persistence of embryonic vessels, anastomosis between ECA and ICA, and intercavernous anastomosis. Although many cases are asymptomatic and incidentally detected, recognition of this anomaly is important in the setting of thromboembolic disease, during planned carotid endarterectomy or transsphenoidal hypophyseal surgery, and detection of associated cerebral aneurysms.

What is already known on this topic?

Congenital absence of the ICA is a rare congenital anomaly. Collateral circulations from contralateral ICA or vertebrobasilar system play an important role to supply the cerebral hemisphere on the affected side. Patients are usually asymptomatic, but a few patients may present with symptoms related to cerebrovascular insufficiency or pressure effects from enlarged collateral vessels. Recognition of this anomaly is important during planned carotid endarterectomy, or transsphenoidal hypophyseal surgery to prevent serious complications. In addition, recognition of this anomaly helps prevent the missed diagnosis of chronic dissection or high-grade carotid stenosis.

What this study adds?

There are two points of interest. Firstly, this is the first report of congenital absence of the ICA in Thai population. Secondly, CT scan of the skull base should be done to identify the hypoplastic carotid canal that helps differentiate ICA agenesis or hypoplasia from acquired causes of carotid occlusion or stenosis. If CT scan of skull base is not done, identification of the hypoplastic carotid canal on axial T1W or T2W MRI can be used.

Potential conflicts of interest

None.

References

1. Makowicz G, Poniatowska R, Lusawa M. Variants of cerebral arteries - anterior circulation. Pol J Radiol 2013; 78: 42-7.
2. Given CA 2nd, Huang-Hellinger F, Baker MD, Chepuri NB, Morris PP. Congenital absence of the internal carotid artery: case reports and review of the collateral circulation. AJNR Am J Neuroradiol 2001; 22: 1953-9.
3. Zhang ZY, Sato S, Liu L, Xue S, Liu ZJ, Tian ZH, et al. Congenital hypoplasia of bilateral internal carotid artery with migraine-like headache with aura. Chin Med J (Engl) 2015; 128: 421-2.
4. Deprez FC, Coulier J, Rommel D, Boschi A. Congenital horner syndrome with heterochromia iridis associated with ipsilateral internal carotid artery hypoplasia. J Clin Neurol 2015; 11: 192-6.
5. Dinç H, Alioglu Z, Erdöl H, Ahmetoglu A. Agenesis of the internal carotid artery associated with aortic arch anomaly in a patient with congenital Horner's syndrome. AJNR Am J Neuroradiol 2002; 23: 929-31.
6. Stagi S, Traficante G, Lapi E, Pantaleo M, Becciani S, Mortilla M, et al. Agenesis of internal carotid artery associated with isolated growth hormone deficiency: a case report and literature review. BMC Endocr Disord 2015; 15: 58.
7. Luk YS, Man EM, Sy AN. Bilateral hypoplasia of the internal carotid arteries. Singapore Med J 2010; 51: e163-e165.
8. Kumaresh A, Vasanthraj PK, Chandrasekharan A. Unilateral agenesis of internal carotid artery with intercavernous anastomosis: a rare case report. J Clin Imaging Sci 2015; 5: 7.
9. Quint DJ, Boulos RS, Spera TD. Congenital absence of the cervical and petrous internal carotid artery with intercavernous anastomosis. AJNR Am J Neuroradiol 1989; 10: 435-9.
10. Quint DJ, Silbergleit R, Young WC. Absence of the carotid canals at skull base CT. Radiology 1992; 182: 477-81.
11. Nicoletti G, Sanguigni S, Bruno F, Tardi S, Malferrari G. Hypoplasia of the internal carotid artery: collateral circulation and ultrasonographic findings. A case report. J Ultrasound 2009; 12: 41-4.
12. Kaya O, Yilmaz C, Gulek B, Soker G, Cikman G, Inan I, et al. An important clue in the sonographic diagnosis of internal carotid artery agenesis: ipsilateral common carotid artery hypoplasia. Case Rep Radiol 2014; 2014: 516456.

ภาวะการเจริญพร่องของหลอดเลือดแดงคาโรติดซึ่งพบโดยบังเอิญในผู้ป่วยวัยผู้ใหญ่: รายงานผู้ป่วย 1 ราย

ธีรพล ปัญญาปิง

ภาวะการเจริญพร่องของหลอดเลือดแดงคาโรติด เป็นความผิดปกติแต่กำเนิดที่พบได้ยาก โดยมักจะวินิจฉัยได้โดยบังเอิญจากการตรวจด้วยภาพทางรังสีด้วยอาการนำต่าง ๆ กันไป ผู้ป่วยส่วนใหญ่มักจะไม่มีอาการผิดปกติ แต่มีส่วนน้อยที่มาด้วยอาการผิดปกติ ได้แก่ ปวดศีรษะ, Horner's syndrome หรือ อาการจากภาวะสมองขาดเลือดชั่วคราว เป็นต้น ผู้นิพนธ์ได้นำเสนอผู้ป่วยรายหนึ่งที่มีภาวะการเจริญพร่องของหลอดเลือดแดงคาโรติด โดยผู้ป่วยรายนี้ไม่มีอาการผิดปกติทางระบบประสาทใด ๆ ซึ่งตรวจพบจากการตรวจด้วยเครื่องเอ็มอาร์ไอร์ ในการตรวจสุขภาพทั่วไป ผู้ป่วยรายนี้จัดว่าเป็นผู้ป่วยคนไทยรายแรกที่มีการรายงานความผิดปกตินี้ ภาพเอ็มอาร์ไอร์ของหลอดเลือดสมองในผู้ป่วยรายนี้พบว่าหลอดเลือดแดงคาโรติดข้างซ้ายมีขนาดเล็กมากตั้งแต่บริเวณคอไปจนถึงบริเวณฐานกะโหลก โดยพบว่ามีการขยายใหญ่ของหลอดเลือด PCOM เพื่อที่จะไปเลี้ยงหลอดเลือด MCA ซึ่งการตระหนักถึงความผิดปกตินี้มีความสำคัญอย่างยิ่งในการวางแผนการผ่าตัดหลอดเลือดแดงที่คอหรือการผ่าตัดผ่านช่องโพรงอากาศด้านหลัง แม้กระทั่งในกรณีของภาวะสมองขาดเลือดชั่วคราวที่มีสาเหตุจากการอุดตันของลิ่มเลือดในหลอดเลือด