A Rare Complex Case Report: Bilateral Congenital Internal Carotid Artery Hypoplasia Associated with Ruptured Left True Posterior Communicating Artery Aneurysm

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Abstract

Congenital internal carotid artery hypoplasia is a rare condition characterized by underdevelopment or reduced caliber of the internal carotid artery during embryonic development. This anomaly presents significant challenges in management, particularly in neurosurgical interventions for cerebrovascular events. We present a case report of a 67-year-old male who presented with subarachnoid hemorrhage and intraparenchymal hemorrhage extending as intraventricular hemorrhage due to a ruptured left true posterior communicating artery aneurysm, associated with intraoperative findings of left internal carotid artery aneurysm, accompanied by incidental findings of bilateral congenital hypoplasia of the internal carotid artery on computed tomography angiography. Surgical intervention involved a left frontotemporal craniotomy, during which both aneurysms were successfully clipped. This case underscores the critical importance of meticulous preoperative evaluation, utilizing advanced neuroimaging modalities to identify such anomalies, particularly in patients with acute cerebrovascular events. Furthermore, it emphasizes the necessity for meticulous surgical planning and intraoperative vigilance to effectively manage associated vascular pathologies.

Keywords

- congenital internal carotid artery hypoplasia
- cerebrovascular anomalies
- neurosurgical interventions
- aneurysm
- bilateral

Introduction

The internal carotid artery (ICA), as one of the primary blood vessels supplying the brain, typically undergoes minimal variation during embryonic development. Congenital anomalies affecting the cerebral vasculature represent a spectrum of conditions posing challenges in diagnosis, management, and prognosis. Congenital internal carotid artery hypoplasia (CICAH) is a rare occurrence with significant implications, particularly in neurosurgical interventions for cerebrovascular events. ICA hypoplasia, characterized by underdevelopment or reduced caliber during embryonic development, can lead to various cerebrovascular complications, including aneurysm formation and rupture, contributing to morbidity and mortality. Among these anomalies, congenital hypoplasia of the bilateral ICA with associated aneurysms presents a

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complex clinical scenario requiring careful evaluation and tailored treatment strategies. Diagnosing congenital hypoplasia of the bilateral ICA and associated aneurysms requires a multidisciplinary approach incorporating neuroimaging modalities such as magnetic resonance angiography, computed tomography angiography (CTA), and digital subtraction angiography. These imaging studies not only aid in confirming the diagnosis but also provide crucial information regarding the anatomical configuration of the vasculature and the extent of the aneurysm. The management of congenital hypoplasia and associated aneurysms necessitates careful consideration of various factors, including the patient's clinical presentation, the size and location of the aneurysm, and the underlying cerebrovascular anatomy. Treatment options may include conservative management with surveillance, endovascular interventions such as coil embolization or stent placement, or surgical approaches such as clipping or bypass procedures. This article aims to report a complex case scenario presenting with subarachnoid hemorrhage, intra-parenchymal hemorrhage extending as intraventricular hemorrhage found to be from ruptured left true posterior communicating artery (PCoA) aneurysm associated with intraoperative findings of left ICA aneurysm, and incidental finding of bilateral CICAH on CTA with whole circulation depends on the vertebrobasilar system.

Case Report

A 67-year-old male presented to the neurosurgery department with a history of two episodes of projectile vomiting followed by a sudden onset of loss of consciousness in the morning. Patient's relative took the patient to a local hospital where a computed tomography (CT) of brain was done and subsequently the patient was referred to our hospital for further management. The patient's medical history includes hypertension for 2 years, self-stopped medications for 2 months, and a history of left-side gangliocapsular region infarct 3 years back for which the patient was managed conservatively. Upon presentation, the patient was hemodynamically stable, with a pulse rate of 100 beats per minute, blood pressure measuring 160/90 mmHg, and an oxygen saturation level of 94% on room air. His neurological status, as assessed by the Glasgow Coma Scale (GCS), was 8/15 (E2V1M5). Pupillary examination revealed bilateral 3 mm pupils reacting to light. Patient had right-sided hemiplegia with power in the upper and lower limbs graded as 1 on the Medical Research Council (MRC) scale, while the power in the left upper and lower limbs was graded as 4 MRC. Bilateral plantar reflexes were elicited as downgoing. Prior imaging conducted at the referring institute consisted of a plain CT of the brain, indicating acute diffuse thick subarachnoid hemorrhage with 19*24*10 mm intraparenchymal hemorrhage in the left gangliocapsular region extending as intraventricular hemorrhage in all four ventricles (►**Fig. 1**). Subsequent CT cerebral angiography further delineated the vascular pathology, identifying a 2.8*3 mm left true PCoA saccular aneurysm (►**Fig. 2**) with a neck measuring 2 mm and having superolateral orientation surrounded by intraparenchymal

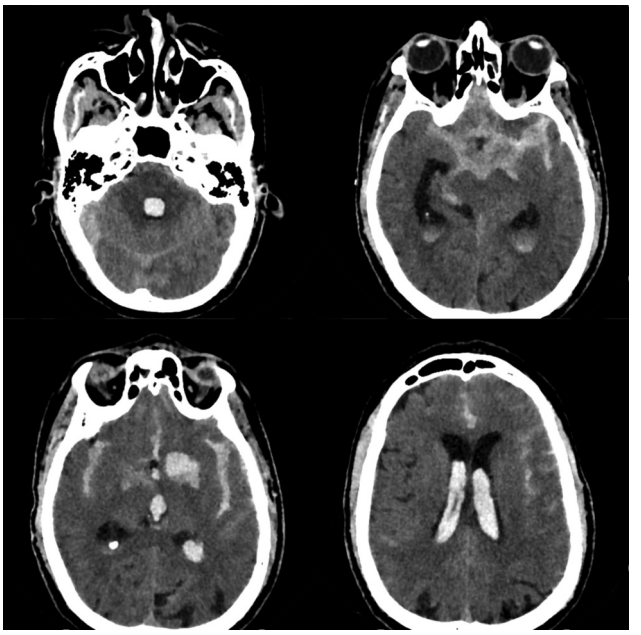

Fig. 1 Preoperative plain computed tomography of brain.

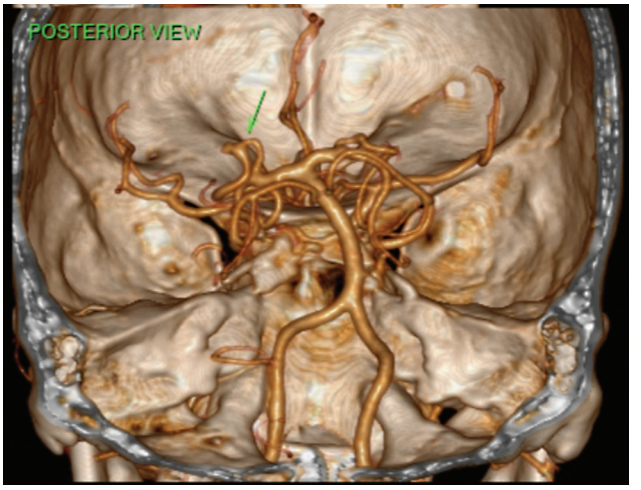

Fig. 2 Three-dimensional volume rendered preoperative image of intracranial angiogram showing true posterior communicating artery aneurysm.

hemorrhage as described above. The subarachnoid hemorrhage was classified as Hunt and Hess Scale 4, Modified Fischer CT Grade 4, and World Federation of Neurological Surgeons Grade 4 at the time of presentation to our institute. Moreover, the patient was found to have incidental findings of bilateral CICAH on CTA (►**Fig. 3**). The skull base CT bone window was also suggestive of bilateral hypoplastic carotid canal with a diameter of 3.1 mm on the right side and 3 mm on the left side (►**Fig. 4**). The intricate clinical situation underscores the paramount importance of timely and thorough management to effectively address the underlying vascular pathology and its associated sequelae. Left frontotemporal (pterional) craniotomy done with classical pterional incision. The sylvian fissure was opened. The optic nerve, ICA, and oculomotor nerve were identified. The incidental finding of bilateral CICAH was confirmed intraoperatively

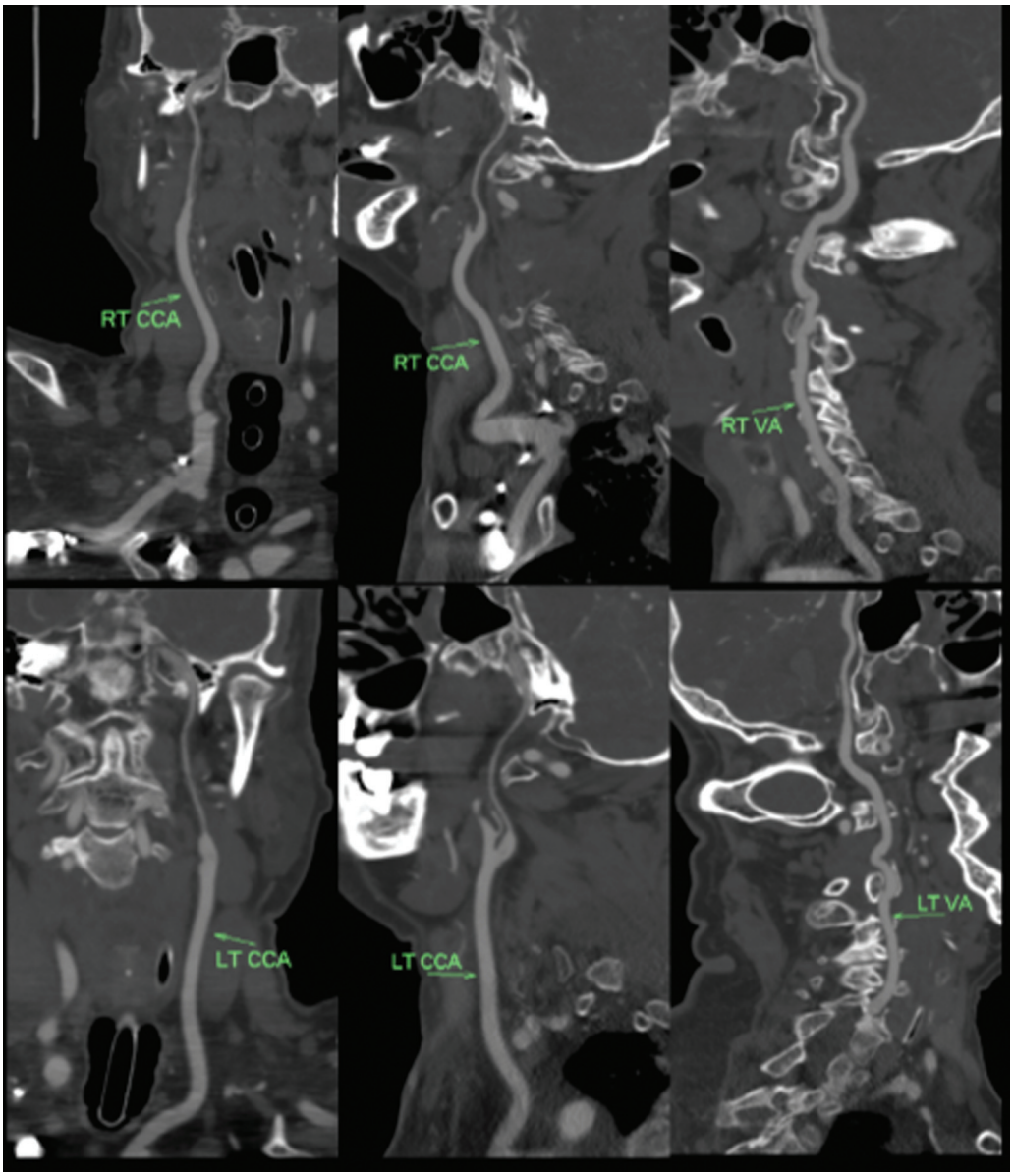

Fig. 3 Computed tomographic angiography of neck showing bilateral congenital hypoplastic internal carotid artery as compared to diameter of bilateral vertebral artery.

with the whole anterior cerebral circulation found to be dependent on vertebrobasilar circulation with resultant dilatation of vertebrobasilar circulation. The PCoA and basilar arteries were seen through the optico-carotid triangle. By using optico-carotid and carotico-oculomotor corridor, left true PCoA aneurysm was identified surrounded by intraparenchymal hemorrhage. Intraparenchymal hemorrhage was drained carefully. The aneurysm sac was dissected and a permanent clip was applied at the neck of the aneurysm (►**Fig. 5**). During dissection, the patient was also found to have a left ICA supraclinoid segment aneurysm adherent to the sphenoid wing which was not identified on prior preoperative imaging (►**Fig. 6**). With meticulous dissection, the aneurysm was dissected out and a permanent clip was applied at the neck of the aneurysm. Hemostasis achieved. Dura closed in a water-tight manner. The skin incision was closed in layers. The postoperative period was uneventful and the patient was discharged in a tracheostomized state on room air with a GCS of 9T/15 (E4VTM5).

Discussion

The ICA, as one of the primary blood vessels supplying the brain, typically undergoes minimal variation during embryonic development. Normal embryogenesis entails the origin of the ICA from the third arch artery and the dorsal aorta.[1] The first documented case of absent ICA was noted by Tode during an autopsy conducted on 10 October 1787.[2] Subsequently, Verbiest [3] reported the first confirmed case of CICAH through angiography. Despite early observations, the precise mechanism underlying CICAH remained elusive. Various hypotheses have been proposed to elucidate the etiology of CICAH. Keen[4] suggested that factors such as over-bending of the head, excessive pressure, and amniotic adhesion could

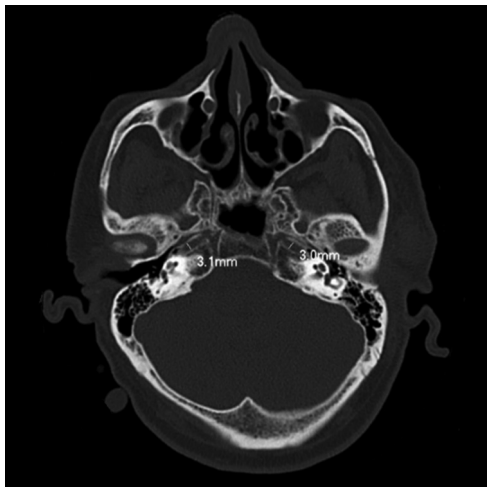

Fig. 4 Skull base computed tomography bone window showing bilateral hypoplastic carotid canal.

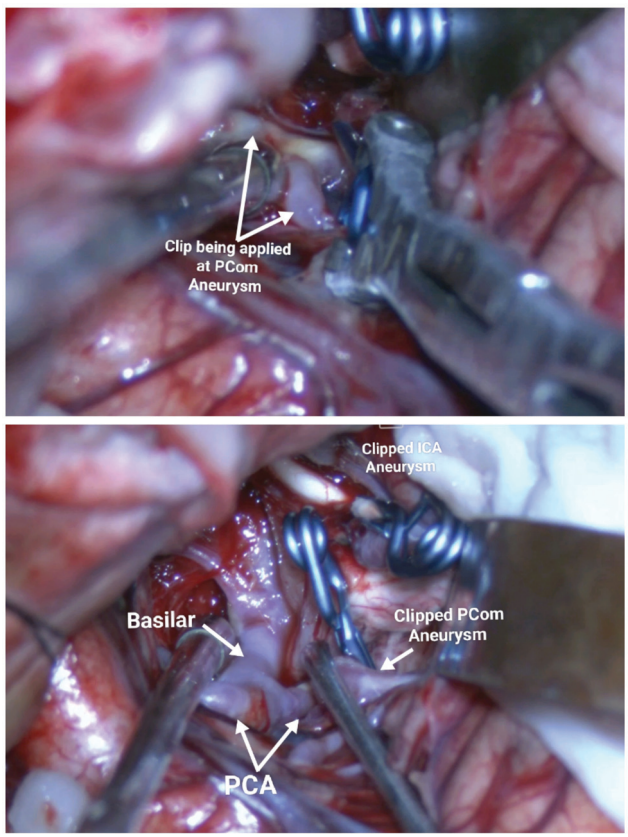

Fig. 5 Preoperative computed tomography angiography coronal view (**A**) and intraoperative image of incidental internal carotid artery aneurysm before and after application of permanent aneurysm clip (**B**).

contribute to the absence of ICA. Additionally, genetic studies have implicated certain genes, including FOXI3 and 22q11.2, in the pathogenesis of CICAH.[5,6] These findings underscore the multifactorial nature of CICAH and highlight the need for further research to elucidate its underlying mechanisms. The annual incidence of CICAH is about 0.01%.[2,6] CICAH usually occurs unilaterally. Bilateral CICAH has been reported but is even less common.[7] According to a study by Tran-Dinh et al,[1] hypoplasia of the left ICA is observed more often, with an incidence of approximately 1.5 times that of hypoplasia of the right ICA. Traditionally, CICAH is classified according to the classification of Lie[8] type 1: ICA aplasia/hypoplasia associated with the anterior communicating artery (ACoA) and PCoA hypertrophy. The ipsilateral anterior cerebral artery (ACA) is supplied by the patent ACoA, and the middle cerebral artery (MCA) is supplied by the hypertrophied PCoA. Type 2: ICA aplasia/hypoplasia associated with ACoA hypertrophy, with MCA and ipsilateral ACA supplied by ACoA. Type 3: bilateral ICA aplasia/hypoplasia. Vertebrobasilar circulation is responsible for the maintenance of anterior circulation of the brain. PCoA hypertrophy often occurs. Type 4: there is unilateral underdevelopment of a cervical segment of the ICA associated with transcavernous communication. It can be located behind the clivus, or run superior, through, or on the floor of the sella turcica.[9] Type 5: bilateral hypoplasia of the ICA. The ACA is supplied through a hypoplastic ICA and the MCA is through an enlarged PCoA. Type 6: aplasia/hypoplasia of the ICA associated with anastomosis of the ipsilateral external carotid artery (rete mirabilis). Our case showed a type 3 collateral circulation. The increased blood flow through collateral vessels and altered flow dynamics may cause ACoA and PCoA aneurysms.[10,11] In these patients, the prevalence of intracranial aneurysm is estimated to be about 24 to 34%, while in the general population, it is 2 to 4%.[10,11] Agenesis and hypoplasia of the carotid artery can be diagnosed by CT examination of the skull base. As per the study done by Tewari et al,[12] the mean antero-posterior diameter and width of the carotid canal are 0.535 and 0.683 cm, respectively. A small osseous carotid canal can be seen on the skull base CT in the case of CICAH. Recognition of CICAH is very important in the planning of surgical procedures such as transsphenoidal hypophyseal surgery and carotid endarterectomy. A preoperative diagnosis of this congenital anomaly before carotid endarterectomy is essential because both hemispheres may depend on a unilateral carotid artery. Before transsphenoidal hypophyseal surgery, the surgeon should foresee the possibility of finding an intracavernous vessel in any patient with no identifiable ICA.[13]

Conclusion

CICAH poses significant diagnostic and therapeutic challenges due to its association with cerebrovascular anomalies such as aneurysms. Our case report emphasizes the crucial role of meticulous preoperative evaluation and advanced neuroimaging techniques in identifying such anomalies, particularly in patients with acute cerebrovascular events. Additionally, meticulous surgical planning and intraoperative vigilance are paramount to address associated vascular pathologies effectively. The aim of publishing this article is to contribute to the global literature on CICAH with valuable insights for neurosurgeons to ultimately improve patient care and outcomes in similar clinical scenarios. Further research is warranted to

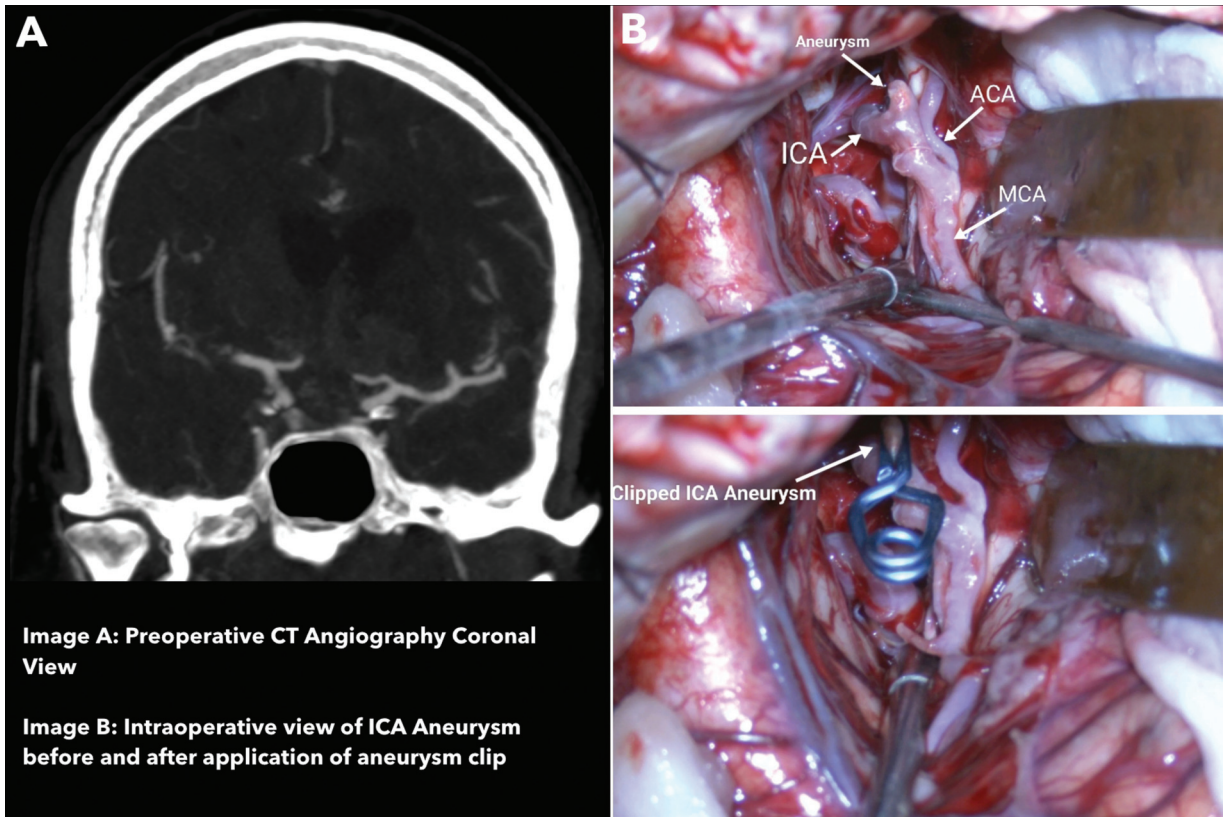

Fig. 6 Intraoperative image of true posterior communicating artery aneurysm during and after application of permanent aneurysm clip.

elucidate the underlying mechanisms of CICAH and optimize management strategies for improved patient outcomes.

Conflict of Interest
None declared.

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