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Internal Carotid Artery Hypoplasia Misidentified as Internal Carotid Artery Dissection

CASE REPORT

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ABSTRACT

Agenesis or hypoplasia of the internal carotid artery (ICA) may easily be confused with dissection or occlusion. We report a case of a 24-year-old female with complaint of acute left-hand hypoesthesia and a history of occasional intermittent numbness of her right hand with myoclonic jerking. Because previous imaging studies over 2 years were interpreted as occlusion of the left ICA secondary to carotid dissection, the treating physician had prescribed anticoagulant therapy. During transcranial Doppler (TCD) examination, the spectral waveform was unexpectedly normal, prompting a repeat review of all imaging due to the TCD results. Magnetic resonance angiography (MRA) revealed the same "flame-like" appearance of the ICA origin. Late-phase digital subtraction angiography showed a small caliber cervical ICA (occluded at the skull base). Computed tomography demonstrated absence of the carotid canal, confirming an absent intracranial portion of the ICA and establishing a correct diagnosis of left internal carotid hypoplasia. Vascular ultrasound and TCD examinations are noninvasive and inexpensive tools that can improve the interpretation and understanding of the clinical significance of other "static" radiographic tests (MRA, digital subtraction angiography). An accurate diagnosis is essential to avoid risky, aggressive treatment, such as anticoagulation for an "absent" dissection.

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KEYWORDS:
transcranial Doppler; internal carotid artery (ICA); agenesis; hypoplasia; vascular ultrasound

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INTRODUCTION

Dysgenesis, including agenesis, aplasia, and hypoplasia, of the internal carotid artery (ICA) is the congenital absence or insufficiency of the artery with unilateral or bilateral involvement. The left ICA is more frequently involved.[1,2] This artery rarely shows abnormal development, with an incidence estimated to be around 0.01%. The most common and nonspecific symptom in ICA dysgenesis is headache; however, most patients are asymptomatic due to the ample collateral circulation provided by the Circle of Willis.

ICA dysgenesis also may be discovered at birth if the ICA anomaly is accompanied by other, more obvious anomalies.[3] In this case, for example, a missing ICA was identified as the underlying cause of unexpected symptoms such as recurrent, unexplained epistaxis and secondary hypopituitarism.[4] Acquired conditions can have a similar appearance to these congenital anomalies. Consequently, ICA agenesis can be confused with atherosclerosis, arteritis, tubular fibromuscular hyperplasia, intimal dissection and Moya Moya disease.[2]

It is important to distinguish between agenesis, which can be compatible with normal life without intervention, and acquired conditions that may require proper treatment with endarterectomy or anticoagulation. ICA dysgenesis is most often found as an incidental finding on ultrasound, angiography, computed tomography (CT), magnetic resonance angiography (MRA), or digital subtraction angiography (DSA) of the head and neck.[5] The definitive diagnosis is made by confirming a missing or small carotid canal with well-developed collateral circulation at the base of the skull on MRI and CT.

Secondary cerebral aneurysms are common in people with ICA dysgenesis due to the dysplastic changes in the collateral arteries of the Circle of Willis from increased hemodynamic pressure.[1,2,6] The incidence of cerebral aneurysm in these patients is 24% to 34%, substantially higher than in the normal population (2-4%).[1] In cases of an absent/occluded ICA, the main vascular supply for the brain is predominantly the contralateral ICA in unilateral disease and the vertebrobasilar system in bilateral disease. Less commonly, the affected hemisphere is supplied by an intercavernous anastomotic vessel connecting the ICAs.[7,8]

To avoid prescribing risky, aggressive treatment (for example, anticoagulation therapies for an "absent" dissection), an accurate diagnosis of congenital vessel malformation is clearly needed. Agenesis and hypoplasia may easily be confused with dissection or occlusion. Vascular ultrasound modalities, including transcranial Doppler (TCD) examination, could improve the interpretation and understanding of other radiographic tests.[9] TCD is an ultrasound technique used to find osseous or soft tissue windows on the skull for Doppler ultrasound imaging of the major blood vessels supplying the brain. Here, we report a case where use of TCD examination uncovered the true diagnosis of ICA hypoplasia and allowed the patient to avoid risky, unnecessary anticoagulant therapy.

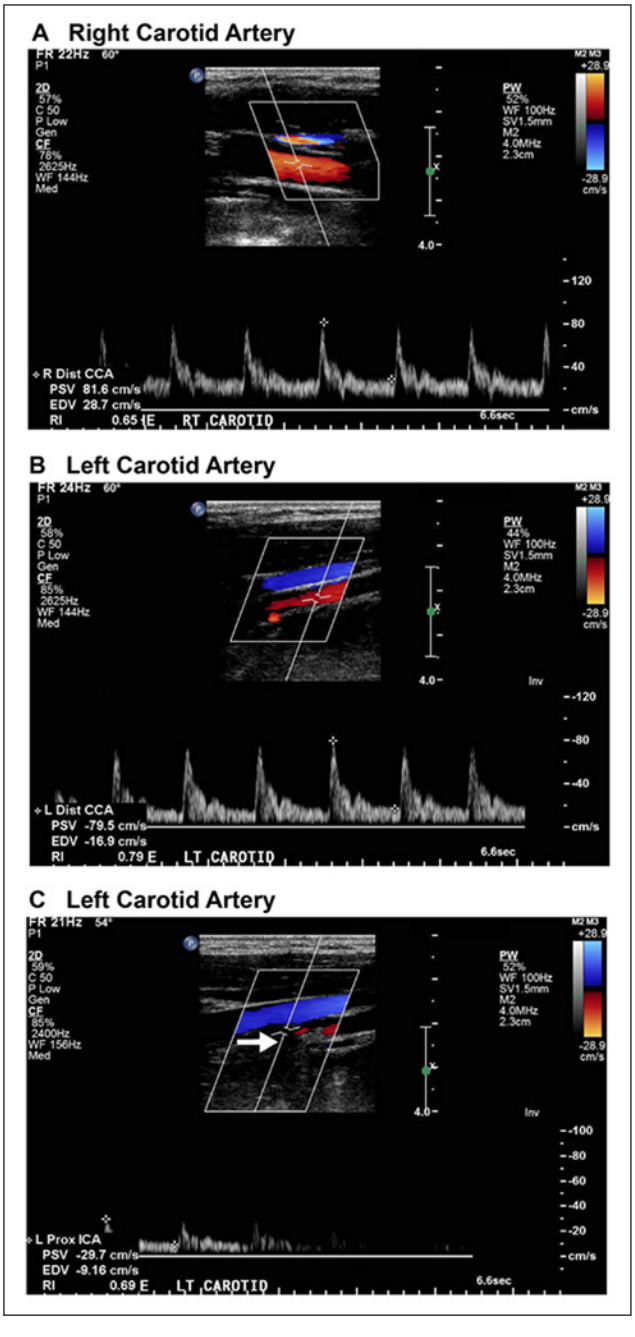

Figure 1 (A) Normal waveforms and velocity in the right common carotid artery (CCA); **(B)** small left CCA suggesting abnormal development; **(C)** proximal left internal carotid artery displaying "flame-like" appearance.

CASE

A 24-year-old female inpatient presented to the Houston Methodist Vascular Ultrasound Laboratory with a complaint of acute left-hand hypoesthesia and a history of occasional, intermittent numbness of her right hand with myoclonic jerking. She relayed a remote history of headache, dizziness, and memory loss with occasional blurring of vision. Her former medical history was significant for a left ICA occlusion, which was discovered 2 years prior. At that time, she was treated with anticoagulant therapy.

The original workup included an MRI of the brain and an MRA. Magnetic resonance venogram was normal, and MR angiography of the neck showed no visualization of the left ICA except an approximate 7-mm stump at the origin, with a small caliber left common carotid artery (CCA). Cerebral angiograms were interpreted as left ICA dissection with resultant severe diffuse narrowing of the ICA. The occlusion was reported to be at the distal cavernous/ophthalmic segment. The prior treating physician attributed the carotid dissection to a minor car accident. The patient was discharged on coumadin, which she took for approximately 1 year.

When she presented to Houston Methodist with her symptoms, we ordered a carotid duplex examination, which suggested a left ICA occlusion, although certain aspects were unusual. We observed a small left CCA (Figure 1A, B) and a large external carotid artery with multiple, prominent branches. These are not routinely seen on carotid duplex, except for the superior thyroid branch. Only the most proximal portion of the small-caliber left ICA was visualized, and it exhibited a flame-like appearance (Figure 1C). Doppler velocities were significantly reduced in the left ICA at 29/9 cm/sec (peak and end diastolic velocity) and a distal ICA occlusion was suspected. The Doppler waveform, however, did not reveal a spike below the baseline in the left CCA, as is routinely seen with ipsilateral ICA occlusion. The left vertebral artery was noted to be larger than normal, with high diastolic flow velocity. The right carotid examination was unremarkable. Due to the inconclusive duplex findings, we ordered further diagnostics.

An MRA of the head without contrast suggested "occlusion" of the left ICA with good cross-filling of the left middle cerebral artery (MCA) territory by way of the anterior communicating artery; a large left posterior communicating artery was noted (Figure 2A, B). MRA of the neck was performed, which also suggested occlusion of the left ICA with codominant vertebral arteries. MRA of the brain without contrast demonstrated an "occluded" left ICA with no definite acute infarction. Results from an echocardiogram were normal, and extensive bloodwork revealed only a vitamin B12 deficiency.

The patient was initially discharged on folic acid and vitamin B12 injections for her B12 deficiency, clopidogrel to reduce risk of platelet aggregation, and ibuprofen as needed for pain/headache. We instructed the patient to

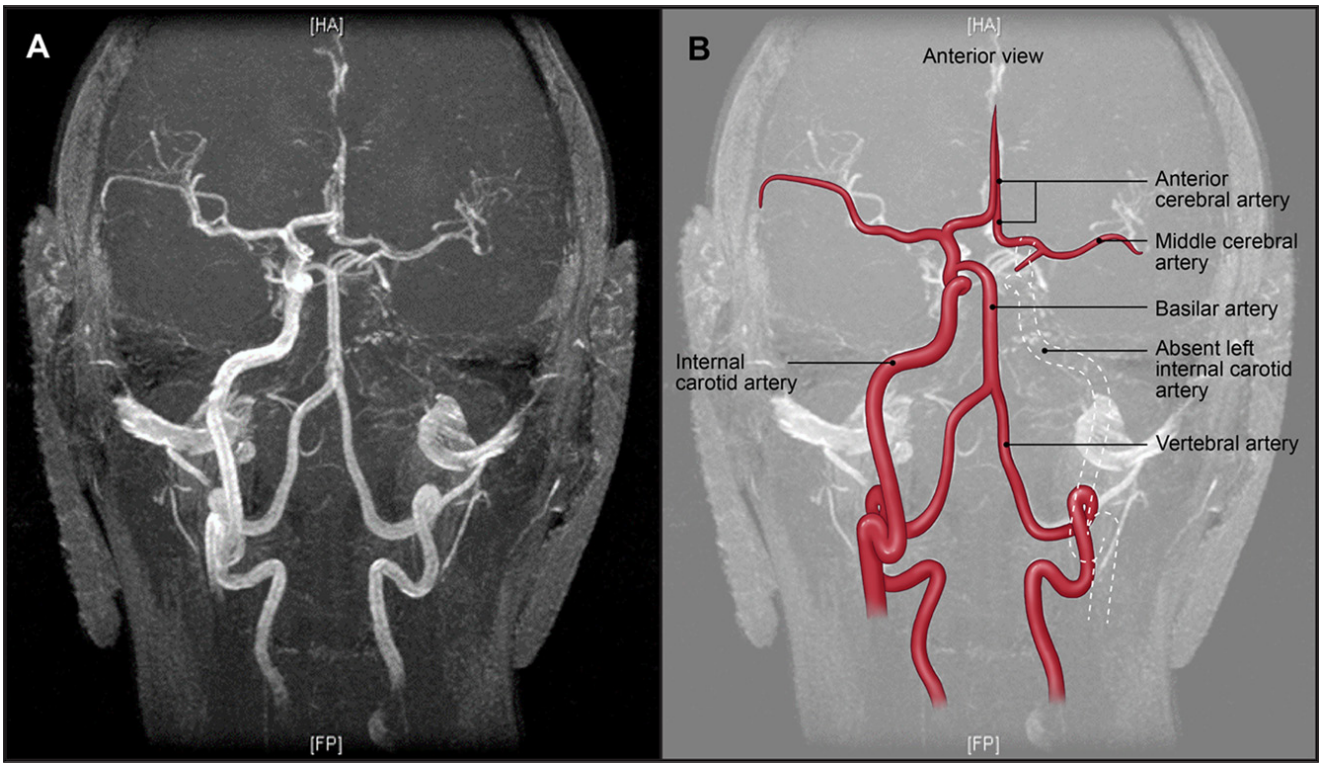

Figure 2 Absent left internal carotid artery with good cross-filling via contralateral anterior cerebral artery.

return as an outpatient for a hematology consult to rule out possible hypercoagulable state, an electroencephalograph to rule out possible seizure activity, a transcranial Doppler to rule out patent foramen ovale (PFO), and a 24-hour Holter monitor to rule out cardiac arrhythmias.

Of these outpatient tests, the only unusual finding was the TCD examination (Figure 3). A PFO was not present; however, the TCD findings revealed an "unexpectedly normal" left MCA spectral waveform, which showed no significant difference between the upstrokes and waveforms observed in both MCAs. In the event of an acute ICA occlusion, we expect to see retrograde flow and curve blunting. Retrograde flow happens because of the collateral blood flow provided through the opposite ICA via the anterior communicating artery. Collateral flow from the other side results in sufficient circulation but misses the sharp upstroke, leading to curve blunting. Because we did not see these expected changes, we inferred a strong ipsilateral collateral development, which often occurs in congenital anomalies. Furthermore, on the side of the occluded ICA, we saw the low resistance external carotid artery (ECA) dynamics, but the left ophthalmic artery displayed anterograde rather than retrograde flow on TCD, which is unusual in situations where the ECA takes over for the acutely closed ICA.

Our TCD finding prompted a complete review of all the previous imaging studies. The previous MRAs and DSA demonstrated the same "flame-like" appearance of the ICA origin as on duplex ultrasound. Late-phase DSA showed a small caliber cervical ICA, which occluded at the skull base (Figure 4A, C). These findings could have been consistent with the previous diagnosis of left ICA dissection; however, the revealing finding was in the CT result, which demonstrated absence of the carotid canal (Figure 5). This finding, along with the DSA demonstrating a small caliber cervical ICA, confirmed the diagnosis of ICA hypoplasia. Finally, we compared all 2007 films to 2005 films and noted no changes in the patient's disease process over this period.

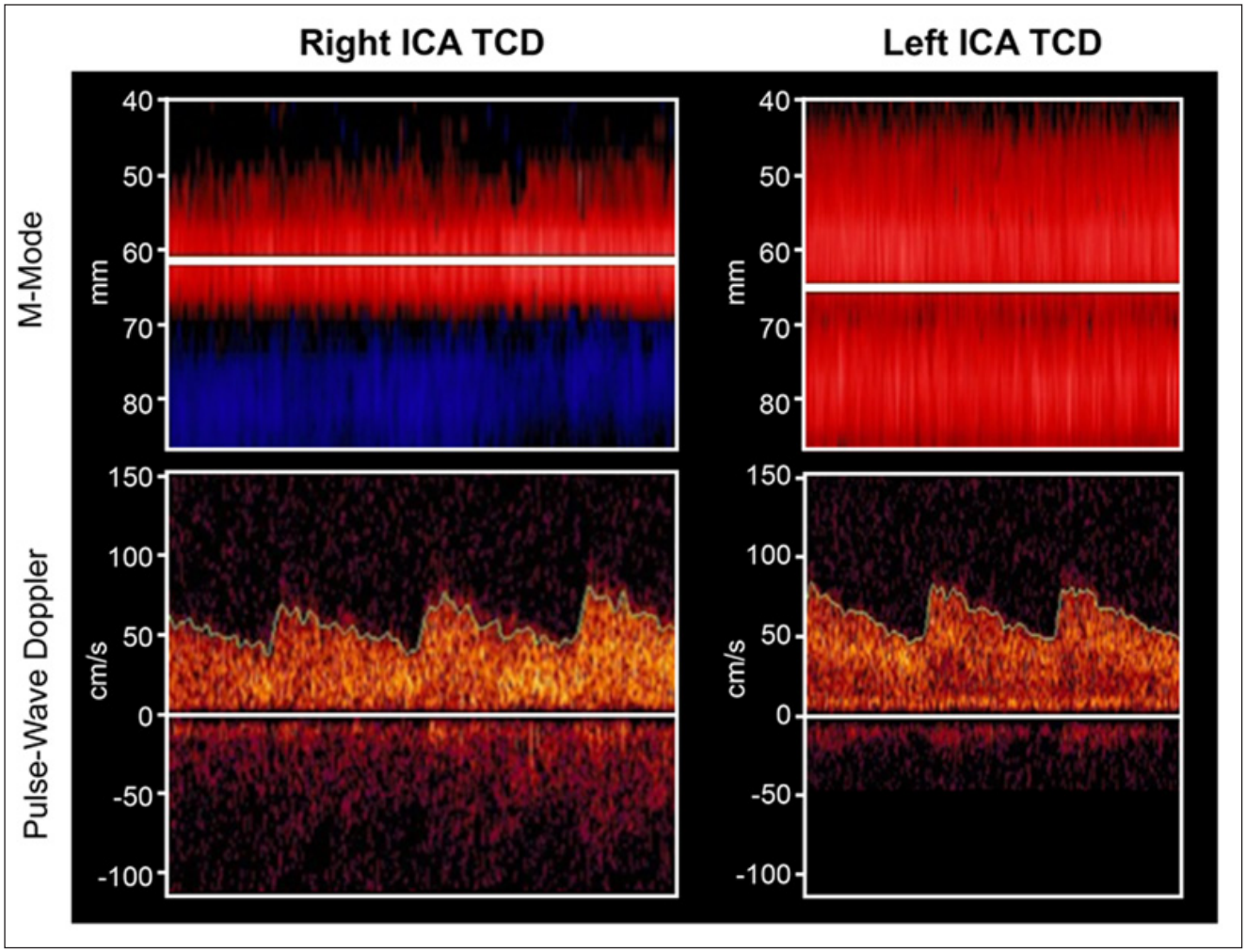

Figure 3 Transcranial Doppler image of the left and right middle cerebral arteries (MCA). Unexpectedly normal left MCA waveform.
ICA: internal carotid artery; TCD: transcranial Doppler

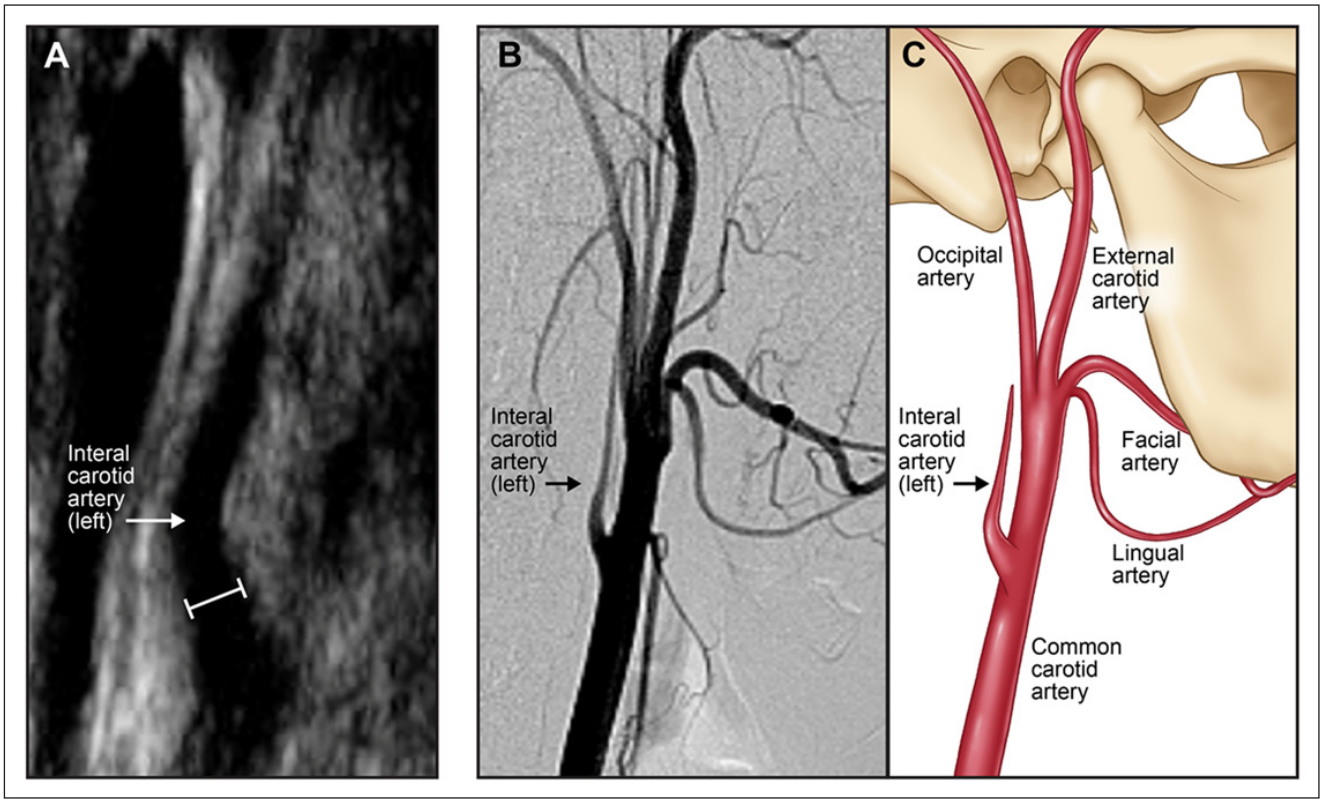

Figure 4 (A, B) Ultrasound and digital subtraction angiography demonstrating a tapered, "flame-like" left internal carotid artery. **(C)** Illustration of the anatomical configuration.

METHODS

Duplex ultrasound imaging was performed using a Phillips iU22 (Phillips Medical Systems) with a 7-4 MHz linear array transducer. Routine carotid duplex imaging was performed along with color flow during the scan. TCD was performed using a PMD 100 (Spencer Technologies) power m-mode Doppler TCD system.

DISCUSSION

This case illustrates the importance of meticulous attention to anatomic variations and Doppler signals of the vascular system to help diagnose and appropriately treat cases of rare conditions such as ICA dysgenesis. In patients who have anatomy outside of the normal realm, using extracranial duplex imaging can be challenging because the vessels proximal to the clavicle and distal to the mandible are not seen due to large bone masses. We must be astute in identifying subtle changes in carotid artery anatomy and Doppler waveforms. At times, these subtle variations are not readily identified, and these oversights can result in consequential differences in treatment plans and potentially severe side effects.

One should also exercise caution when performing Doppler waveform analysis to distinguish between the ICA and ECA. Normally, the ECA waveforms have little flow in diastole whereas the ICA waveforms have a large flow throughout diastole. In cases of occlusion (or absence) of the ICA, the Doppler waveforms of the ECA and its branches may resemble the ICA and may lead to erroneous vessel identification.[10] A smaller-than-normal ipsilateral CCA may raise suspicion for the missing or hypoplastic ICA and reveal the condition's congenital nature due to their common embryologic origin.

In this case, TCD provided significant insight to the correct interpretation of the patient's condition. TCD did not support a simple ICA occlusion in this young adult and therefore prompted a closer look. Although the origin of the ICA was slightly more proximal than the prominent ECA branches on ultrasound, it could have been easily mistaken for an additional ECA branch. It was a small ICA with reduced Doppler velocities. The multiple ECA branches demonstrated low resistant signals in what should have been a higher resistant bed. The "flare" of the proximal portion of the cervical ICA could easily mislead physicians to conclude a carotid artery dissection in all imaging modalities, a diagnosis more consistent with the patient's age and history.

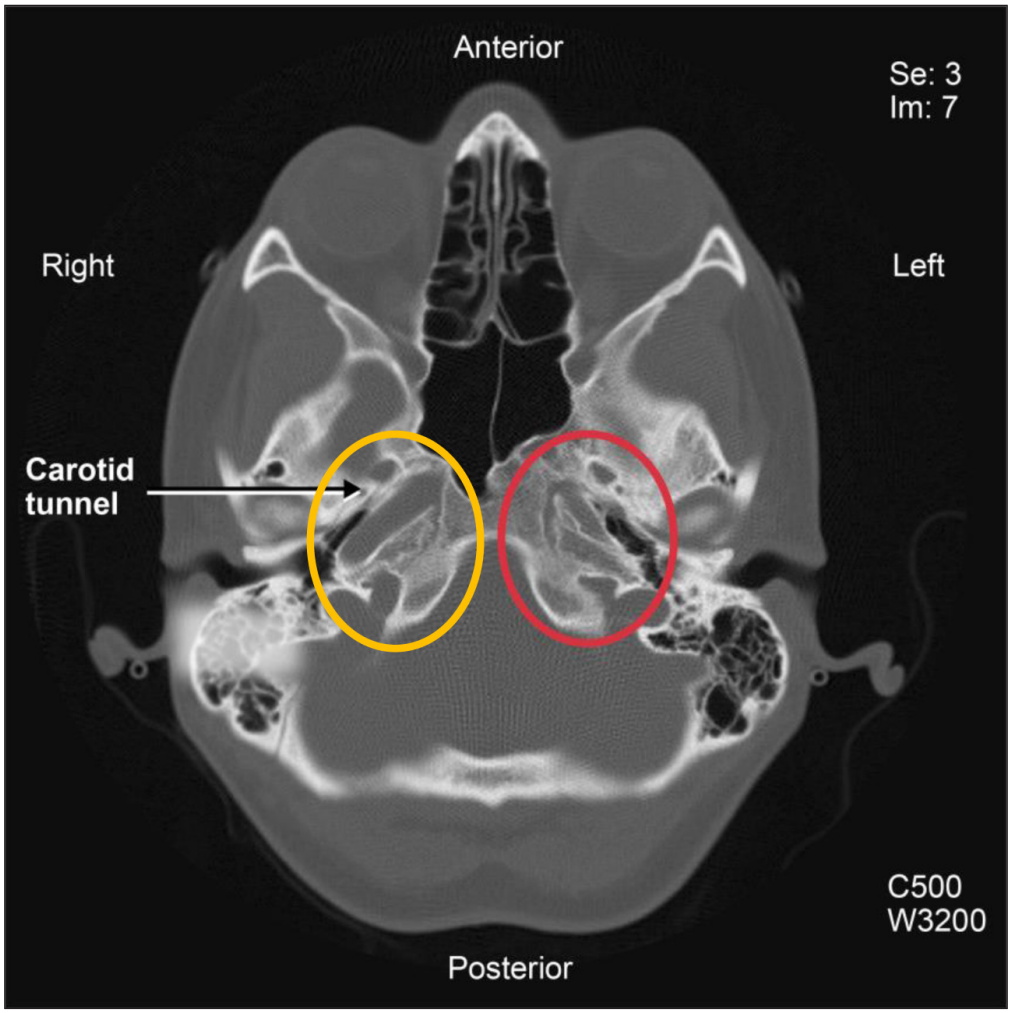

Figure 5 Computed tomography demonstrating absent carotid canal at the base of the skull on the left.

CONCLUSION

TCD and duplex ultrasonography are critical in determining the anatomy of extra- and intracranial carotid circulation. Carotid duplex without TCD could only predict distal disease; it could not provide certainty about the etiology of the direct finding. TCD stands alone as a noninvasive and inexpensive tool to assess intracranial and real-time flow dynamics. It is of immense value in adding insight to the findings of carotid Doppler and static imaging like CT and MRI. We suggest that TCD's real-time vascular assessment could also be complementary to other static radiographic images in young stroke patients. Here, we were able to avoid a high-risk therapeutic intervention—anticoagulation—by correctly interpreting the test results and establishing an accurate diagnosis using carotid duplex ultrasound and TCD.

Physicians and sonographers can be biased towards establishing a diagnosis that they see more frequently in clinical practice. Routines in clinical practice help to save time in evaluating patients, but they also can be a two-edged sword that causes hasty conclusions based on conditions most frequently seen. We must recognize the need to step back and look at each patient as an individual life and story to avoid "automated diagnostics" and minimize the risk of misinterpreting results.

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COMPETING INTERESTS

The authors have no competing interests to declare.

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