

## Case Studies

## A Case of Intrathoracic Carotid Bifurcation Without Klippel–Feil Syndrome

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**Background:** Intrathoracic carotid bifurcation is a rare vascular anomaly, with only 8 cases reported. This vascular anomaly was recently correlated with Klippel–Feil syndrome, a rare congenital disorder involving fusion of the cervical vertebrae. **Methods:** A 70-year-old deaf mute man was admitted to our department because of right hemiparesis and right sensory disturbance. He displayed no abnormalities associated with Klippel–Feil syndrome. Diffusion-weighted imaging revealed acute multiple infarcts in bilateral hemispheres. Computed tomography angiography showed that the left common carotid artery bifurcated at the T2 vertebral level. No significant stenosis was seen in the carotid arteries. Transesophageal echocardiography confirmed a complicated atheromatous lesion at the aortic arch. **Results:** This case was diagnosed with aortogenic brain embolism, incidentally accompanied by left intrathoracic carotid bifurcation without Klippel–Feil syndrome. **Conclusions:** Intrathoracic carotid bifurcation can occur in cases without Klippel–Feil syndrome. The knowledge of this anatomic variation is important in evaluating vascular lesions of the carotid arteries in patients with stroke. **Key Words:** Intrathoracic carotid bifurcation—Klippel–Feil syndrome—ischemic stroke— aortogenic brain embolism.

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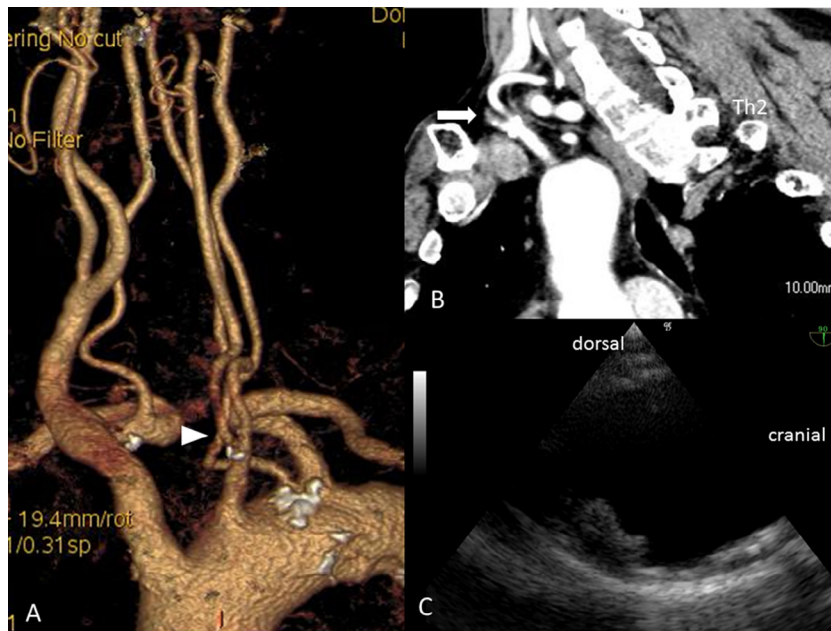
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### Case Description

A 70-year-old deaf mute man with dyslipidemia and hypertension was admitted to our department because of right hemiparesis and right sensory disturbance. On physical examination, he displayed no abnormalities associated with Klippel–Feil syndrome (e.g., short, webbed neck with low hairline). Diffusion-weighted imaging revealed acute multiple infarcts in bilateral hemispheres. Neither time-of-flight magnetic resonance angiography nor carotid ultrasonography revealed left carotid bifurcation within the cervical segment. The right common carotid artery (CCA) was normal. Computed tomography angiography showed that the left CCA bifurcated at the T2 vertebral level, and the left vertebral artery originated directly from the aortic arch (Figs 1, A,B). No significant stenosis was evident in the left CCA or internal carotid



**Figure 1.** (A) Volume-rendered computed tomography angiography in left anterior oblique view shows that the left common carotid artery (CCA) bifurcates at an extremely low level (arrowhead). No significant stenosis is evident in either the left CCA or the internal carotid artery. (B) Sagittal-view computed tomography angiography shows that the intrathoracic CCA bifurcation is located in the median position at the T2 vertebral level (arrow). (C) B-mode transesophageal echocardiography reveals a complicated atheromatous lesion with a thickness of 7 mm on the ventral wall at the aortic arch.

artery. Cervical and thoracic X-rays showed no vertebral abnormalities. Transesophageal echocardiography confirmed a complicated atheromatous lesion with a thickness of 7 mm at the aortic arch (Fig 1C). This case was diagnosed with aortogenic brain embolism, incidentally accompanied by left intrathoracic carotid bifurcation without Klippel–Feil syndrome.

## Discussion

Intrathoracic carotid bifurcation is an extremely rare vascular anomaly, with a relative incidence of .2%.<sup>1</sup> In the literature, only 8 case reports of intrathoracic carotid bifurcation have been described.<sup>1–5</sup> Among these, 3 were complicated with Klippel–Feil syndrome, an uncommon congenital disorder involving fusion of the cervical vertebrae. This segmental disorder has been implicated in the development of a low carotid bifurcation via third aortic arch anomaly.<sup>2–4</sup> Gailloud and colleagues assumed that this development of a lower bifurcation is attributable to persistence of the ductus caroticus, which exists between the third and the fourth aortic arches, accompanied with involution of the third aortic arch, and migration of the external carotid artery precursor toward the ductus caroticus.<sup>4</sup> Congenital comorbidities other than those causing deaf muteness were not observed in the

present case. Bilateral lower carotid bifurcation can be attributed to a segmental developmental disorder associated with Klippel–Feil syndrome. However, unilateral intrathoracic carotid bifurcation, as observed in our case, could be caused by a local disturbance during embryogenesis. Moreover, we emphasize the importance of knowledge about this rare anatomic variation when evaluating vascular lesions of the carotid arteries in patients with stroke.

## References

1. Vitek JJ, Reaves P. Thoracic bifurcation of the common carotid artery. *Neuroradiology* 1973;5:133-199.
2. Brill CB, Peyster RG, Keller MS, et al. Isolation of the right subclavian artery with subclavian steal in a child with Klippel-Feil anomaly: an example of the subclavian artery supply disruption sequence. *Am J Med Genet* 1987;26:933-940.
3. Rielly JP. Intrathoracic carotid bifurcation in Klippel-Feil syndrome. *J Vasc Surg* 2008;47:1071-1073.
4. Gailloud P, Murphy KJ, Rigamonti D. Bilateral thoracic bifurcation of the common carotid artery associated with Klippel-Feil anomaly. *AJNR Am J Neuroradiol* 2000;21:941-944.
5. Gomez CK, Arnuk OJ. Intrathoracic bifurcation of the right common carotid artery. *BMJ Case Rep* 2013;doi:10.1136/bcr-2012-007554.