

CrossRef DOI of original article:

Bilateral Internal Carotid Artery Agenesis: Very Rare Cause of Bilateral Pulsatile Tinnitus

Aynur Aliyeva¹, Ziya Karimov², Ozlem Yagiz³ and Fidan Rahimli Alekberli⁴

¹ University of Korea,

Received: 1 January 1970 Accepted: 1 January 1970 Published: 1 January 1970

Abstract

Bilateral internal carotid artery agenesis is a very rare congenital anomaly. Intracranial blood circulation in the affected internal carotid artery side is provided by collateral vasculature, contralateral internal carotid artery through the anterior communicating artery, and from the vertebrobasilar system through the posterior communicating artery. People with this abnormality may be asymptomatic for many years. However, patients may also be present with ischemic stroke or subarachnoid hemorrhage, headache, and blurred vision. Diagnosis is usually incidental by the performance of carotid artery doppler ultrasonography or cervical/cranial magnetic resonance imaging owing to other symptoms. We report a case of bilateral internal carotid artery agenesis with pulsatile tinnitus lasting five years in the light of clinical presentation and imaging findings.

Index terms— agenesis, bilateral carotid artery, congenital anomaly, pulsatile tinnitus.

1 Introduction

Tinnitus is the perception of sound in the absence of a corresponding external acoustic stimulus. This perception of the sound is associated with activity in the peripheral and central nervous systems that does not match the resonant or mechanical activity in the cochlea. The etiology of tinnitus development and maintenance is still unclear. One of the most common causes of tinnitus is pathological changes along the auditory pathway. Many abnormal conditions of the cochlea, such as sudden hearing loss, noise induced hearing loss, presbycusis or the use of ototoxic drugs, accompany this pathology. These lesions causing hearing loss can result in abnormal neuronal activity in the central auditory pathway. Other risk factors for tinnitus development or maintenance are rarely known (1-2). Tinnitus can be sometimes observed in the patients with normal hearing. The patient describes hearing as similar to ringing, roaring, buzzing, or other sounds. 10%-15% of the population had tinnitus. It has two types: subjective and objective (3)(4). Objective tinnitus is caused by a vascular or muscular origin. The first may be caused by either venous, arterial sources, or arteriovenous shunting. Vascular internal flow reflects rhythmically to the ear in accordance with the heartbeat and blood flow due to the disruption of turbulence. In patients with advanced pulsatile tinnitus, the clinician can sometimes hear these pulsatile sounds during auscultation of the neck and around the ear area. And also as, muscular pulsatile tinnitus results from myoclonus spasm of muscles, most commonly the palatal, tensor tympani, and stapedius. The most common form of tinnitus is non-pulsatile tinnitus, mostly associated with hearing loss and ear diseases (5,6). Pulsatile tinnitus is uncommon, not associated with ear disease but more often in the presence of abnormal extracranial or intracranial blood vessels or intracranial hypertension. Pulsatile tinnitus caused by intracranial hypertension is not synchronized with the heartbeat, and its differential diagnosis is more difficult. Intra-arterial or intravenous angiography, Computer tomography (CT) angiography, and magnetic resonance are used to diagnose classic vascular pulsatile tinnitus.

In this case, we report a patient with an infrequent vascular cause of ipsilateral tinnitus -bilateral internal carotid artery agenesis.

43 **2 II.**44 **3 Case Report**45 **4 T**

46 The patient was a 54-year-old white man who came to the ENT department with a 5-year history of bilateral
47 tinnitus. The tinnitus prevented the patient from sleeping. A physical examination, routine otorhinolaryngology
48 controls, audiometry, and other tests were normal. Also, tinnitus was not heard during auscultation. We decided
49 to evaluate the patient for another rare cause of tinnitus. A carotid duplex ultrasound study showed no bilaterally
50 internal carotid arteries and blood flow. We performed cranial-cervical Magnetic resonance imaging (MRI) and
51 CT angiography of the patient. CT angiography demonstrated agenesis of the bony carotid canal and smaller
52 cavernous sinus (Figure ??). Blood circulation was supplied via the posterior and anterior communicating arteries.
53 Bilateral ophthalmic arteries were supplied from meningeal arteries.

54 After consultation with the radiologist, the patient was definitively diagnosed with bilateral carotid agenesis.
55 It was concluded that pulsatile tinnitus occurred in the patient in accordance with the rhythm of the heartbeat
56 due to abnormal blood flow and turbulent flow. The patient was informed, all cardiological and cranial scans
57 were performed, and no additional pathology was found. It was reported that there was no clear and definitive
58 treatment for pulsatile tinnitus, and the patient was satisfied with this opinion and continued his routine controls.

59 **5 III.**60 **6 Discussion**

61 It should be considered the difference between agenesis, aplasia, and hypoplasia. Agenesis is a completely
62 unsuccessful development with absent primordial tissue by definition. Aplasia is an absent organ in spite of the
63 presence of the main primordial tissue. Hypoplasia is incomplete development, although primordial tissue is
64 presented. Demonstration of the carotid canal is a nuance for differentiation. Even though MRI angiography is
65 performed, CT of the head and neck should be administered to determine the presence/absence of a bony carotid
66 canal (14). In our case, ICA is absent without a carotid canal, so we described it as agenesis. Also, we found a
67 smaller cavernous sinus in the patient that is compatible with other cases in the literature (7).

68 The most common cause of objective vascular tinnitus is an arteriovenous malformation (AVM) in the posterior
69 fossa (15). ICA agenesis is a very rare cause of tinnitus. Cohen et al. Reported one case of tinnitus caused by
70 unilateral carotid artery agenesis (7).

71 Diagnosis is usually made incidentally with the performance of carotid artery doppler ultrasonography or
72 cervical/cranial MRI and CT due to other symptoms (7). Also, CT or MRI angiogram is required to confirm the
73 diagnosis and assess differential diagnoses (8). Differential diagnoses include congenital or acquired ICA stenosis,
74 common carotid artery hypoplasia, and other aortic arch anomalies (12,16,17).

75 We report a rare case of bilateral tinnitus caused by bilateral ICA agenesis. There are less than 30 "Bilateral
76 internal carotid artery agenesis" cases in the current literature. According to our knowledge, our report is the
77 first case in which bilateral ICA causes tinnitus.

78 Internal carotid artery (ICA) agenesis is a very rare congenital anomaly with <0.01% incidence (7,8). Tode
79 reported the first absence of ICA in 1787 (9). Though unilateral ICA defect is common, a bilateral variant
80 is also reported 6 . It is usually asymptomatic because of blood supply through anastomosis in the circle of
81 Willis. In addition, blood circulation is possible with continuous embryologic vessels like the trigeminal artery
82 and collaterales that originate extracranially 3,4 . But, it occasionally may be symptomatic and be associated
83 with other congenital anomalies and acquired defects, like partial brain hypoplasia, a functional deficit of cranial
84 nerves, etc. (10)(11)(12)(13). Because of decreased blood supply in the affected size, adaptation may be provided
85 unusual vasculature. So, it can cause abnormal vascular conditions like aneurysms and AVMs. Congenital ICA
86 defects were described less in light of clinical neurological conditions. The clinical presentation includes commonly
87 ischemic or hemorrhagic stroke, migraine, subarachnoid hemorrhage, contralateral motor weakness, Horner's
88 syndrome, external ear defect, decreased hearing, and rarely tinnitus (7,8,12,13). Although the affected side is
89 more susceptible to ischemia and can potentially cause clinical symptoms in unilateral cases, the contralateral
90 side is becoming sensitive because of decreased circulation due to the shift of circulation to the affected side.

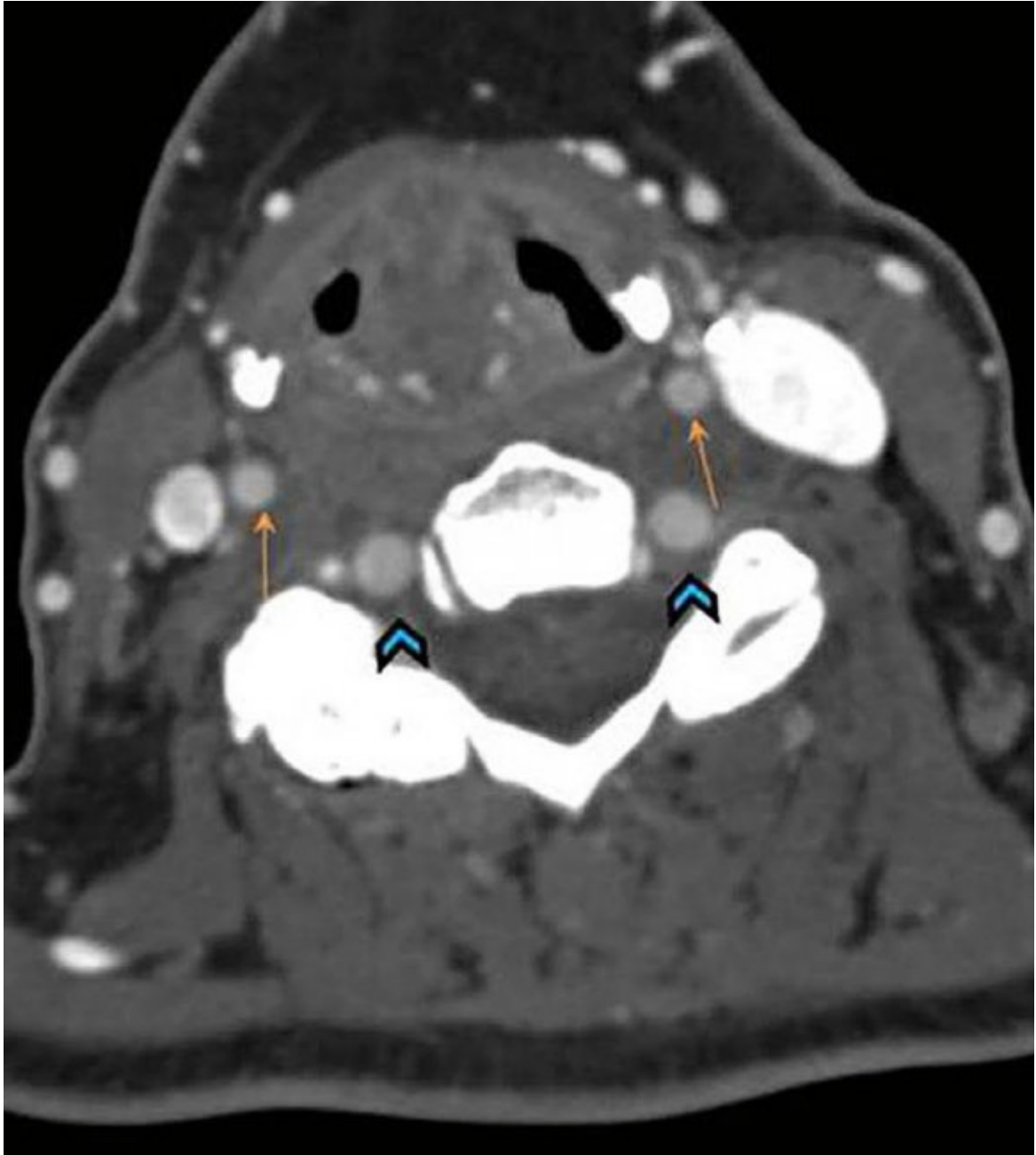


Figure 1:

.1 Conflict of interest:

The authors declare that they have no conflict of interest.

Ethics approval: For A Case Report N/A Consent to participate: Informed consent was obtained from the participant before including him in the case report.

[Tode. Medizinisch Chirurgische Bibliothek] , *Tode. Medizinisch Chirurgische Bibliothek* 10.

[Objective ()] , Iglauer S Objective . 10.1001/ARCHOTOL.1933.03580060159001. *WITH REPORT OF FOUR CASES. Arch Otolaryngol* 1933. 18 (2) p. .

[Paparella et al. ()] , M Paparella , O Shumric , W B Otolaryngology , Saunders . 1973.

[Baguley et al. ()] , D Baguley , D McFerran , D Hall , Tinnitus . 1016/ S0140- 6736. *Lancet* 2013. 382 (9904) p. .

[Kaya et al. ()] *An Important Clue in the Sonographic Diagnosis of Internal Carotid Artery Agenesis: Ipsilateral Common Carotid Artery Hypoplasia. Case Rep Radiol*, O Kaya , C Yilmaz , B Gulek . 10.1155/2014/516456. 2014. 2014 p. .

[Pavaci et al.] ‘Analysis of the audiological characteristics and comorbidity in patients with chronic tinnitus’. S Pavaci , F Tortorella , A B Fioretti , A M Angelone , L D R Businco , M Lauriello . 10.4081/audiores.2019.231. doi.org/10.4081/audiores.2019.231 *Audiol Res* 2019 (2) p. 231.

[Rumboldt et al. ()] ‘Bilateral congenital absence of the internal carotid artery’. Z Rumboldt , M Castillo , S Solander . 10.1007/s00330-002-1742-2. *Eur Radiol* 2003. 13.

[Given et al. (2001)] ‘Congenital Absence of the Internal Carotid Artery: Case Reports and Review of the Collateral Circulation’. C A Given , F Huang-Hellinger , M D Baker , N B Chepuri , Pearse Morris , P . 2022./pmc/articles/PMC7973856/ *AJNR Am J Neuroradiol* 2001. Accessed December 28. 22 (10) p. 1953.

[Dhiman et al. ()] ‘Congenital Third Nerve Palsy Associated With Midbrain Hypoplasia Due to Bilateral Segmental Internal Carotid Artery Agenesis’. R Dhiman , B Gorimanipalli , D R Swamy , S Sharma , A Garg , R Saxena . 10.1097/WNO.0000000000000666. *J Neuroophthalmol* 2018. 38 (4) p. .

[Cohen et al.] *Internal carotid artery agenesis: diagnosis, clinical spectrum, associated conditions and its importance in the era of stroke interventions*, J E Cohen , J M Gomori , R R Leker . 10.1179/016164110X12767786356273. <http://dx.doi.org/101179/016164110X12767786356273>. 2013 32 p. .

[Graham et al. ()] ‘Magnetic resonance imaging in internal carotid artery agenesis with computed tomography and angiographic correlation—case reports’. C B Graham , F J Wippold , G W Capps . 10.1177/000331979905001009. *Angiology* 1999. 50 (10) p. .

[Clarós et al. ()] ‘Major congenital anomalies of the internal carotid artery: Agenesis, aplasia and hypoplasia’. P Clarós , R Bandos , I Gilea . doi: 10.1016/ S0165-5876(99. *Int J Pediatr Otorhinolaryngol* 1999. 49 (1) p. .

[Berkman et al. ()] ‘Metin ORAKDÖ?EN Agenesis of the Left Internal Carotid Artery Associated with Anterior Communicating Artery Aneurysm: Case Report Anterior Kominikan Arter Anevrizmas? ile Sol ?nternal Karotid Arter Agenezisinin Birlikteliği: Olgu Sunumu’. Z Berkman , M Erfişah?n , N B?ber , H Somay . *Turk Neurosurg* 2007. 17 (4) p. .

[Ward et al. ()] ‘Operative treatment of surgical lesions with objective tinnitus’. P H Ward , R Babin , T C Calcaterra , Konrad Hr . 10.1177/000348947508400405. *Ann Otol Rhinol Laryngol* 1975. 84 (4) p. . (Pt 1)

[De Ridder et al. ()] ‘Phantom percepts: tinnitus and pain as persisting aversive memory networks’. D De Ridder , A B Elgoyhen , R Romo , B Langguth . *Proceedings of the National Academy of Sciences of the United States of America* 2011. 108 (20) p. .

[Park and Jeon ()] ‘Pulsatile Tinnitus Arising from Aberrant Internal Carotid Artery at Nasopharynx’. C W Park , E J Jeon . 10.7874/KJA.2012.16.1.35. *Korean J Audiol* 2012. 16 (1) p. 35.

[Axelsson ()] ‘Tinnitus—a study of its prevalence and characteristics’. A Axelsson , A . *British Journal of Audiology* 1989. 23 (1) p. .