International Journal of Otorhinolaryngology and Head and Neck Surgery
Narayankar NM et al. Int J Otorhinolaryngol Head Neck Surg. 2022 Mar;8(3):280-285
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20220485

Case Report

Nutrimentum vascularum as pulsatile tinnitus

Nivedita M. Narayankar*, Chandre Gowda B. V., S. Juthika Rai

Department ENT, MVJ Medical College and Research Hospital, Hosakote, Bangalore, Karnataka, India

Received: 22 January 2022
Accepted: 09 February 2022

***Correspondence:**
Dr. Nivedita M. Narayankar,
E-mail: dr.niveditamn.92@gmail.com

ABSTRACT

Tinnitus is one of the most common otological symptoms encountered in otorhinolaryngology. In recent times our understanding of tinnitus has significantly progressed but nonetheless, it still constitutes a very incapacitating symptom which greatly affects the quality of life. Objective tinnitus accounts for only 5% and has various aetiologies both non-otological and otological which include both non vascular and vascular. Vascular malformations like dural arteriovenous fistulas (DAVFs) constitute about 10% of objective tinnitus and hence are of academic interest to an otolaryngologist due to their rarity. We present one such case. A 26-year-old female presented to the department of ENT with chief complaints of a left pulsatile tinnitus of 2 months duration. A detailed clinical examination revealed no abnormality. A pure tone audiometry (PTA) was then done which once again showed hearing within normal limits. Given the clinical persistence of the symptom which was affecting her daily activities, further investigation of magnetic resonance imaging (MRI) brain was done. A suspicion of a dural arterio-venous fistula in the transverse sinus further warranted an arteriography which showed a low grade DAVF at the level of the left sigmoid sinus. DAVFs, although infrequent, constitute a treatable cause of tinnitus and hence should be considered as a possible diagnosis in all patients with pulsatile tinnitus, especially if it is unilateral. We also carried out a review of the current medical and surgical management in an attempt to better understand this rare yet intriguing condition.

Keywords: Pulsatile tinnitus, Objective tinnitus, Dural arterio-venous fistula

INTRODUCTION

Tinnitus is the conscious, usually unwanted perception of sound that arises or seems to arise without an external source.[1]

Tinnitus may be categorised as subjective, objective, primary or secondary.

Objective tinnitus accounts for only 5% and has various aetiologies both otological like Meniere's disease, Acoustic neuroma etc.; and non otological causes such as traumatic, infectious, oncological, pharmacological, systemic and vascular causes like glomus tumours and vascular malformations like DAVFs. In the presence of such varied aetiologies, it is imperative to use standardized protocols in the otolaryngology consult, without underestimating any sign, since it may very well be the first manifestation of a serious and potentially fatal pathology.[1-3]

The most frequent causes of objective tinnitus described in literature are arteriosclerosis of the carotid or subclavian artery and benign intracranial hypertension; however, DAVFs are a far less common cause.[1-3]

Less than 10% of patients who present with tinnitus suffer from pulsatile tinnitus.[4] Pulsatile tinnitus is described as a tinnitus producing sound of regular pulsations. This may be subjective or objective and there is usually a genuine physical source of sound.[5]

There are two plausible causes of pulsatile tinnitus: 1) Blood flow acceleration or changes in blood flow disrupts the laminar flow and the resulting local turbulence is audible. 2) Normal flow sounds within the body are perceived more intensely, either as a result of alterations

in the inner ear with increased bone conduction or as a result of disturbance of sound conduction leading to loss of the masking effect of external sounds.[1]

Pulsatile tinnitus is usually unilateral, unless the underlying vascular pathology is bilateral.

Somatosensory pulsatile tinnitus presents as bilateral tinnitus with no vascular cause but more often than not, it is possible to identify the cause of pulsatile tinnitus.[6] In addition to the patient's medical history and targeted clinical examination, imaging procedures also play an important role in diagnosis. However, despite careful examination, no cause is found in up to 30% of patients.[7]

The DAVFs consist of small anomalous connections or shunts between the collateral of a dural artery with the dural vein or sinus, constituting just 10%-15% of all intracranial arteriovenous malformations.[8] Most of them are idiopathic and a small percentage have an associated history of craniotomy, trauma or thrombosis of the dural sinus. They are more frequent between the 6th and 7th decade of life and in women, although men are the ones who frequently present with more severe clinical manifestations.

TSDAVF is a diagnostically elusive entity that is critical for the otologist to account for when confronted by pulsatile tinnitus in the face of normal otoscopy. If left untreated, TSDAVF may result in a catastrophic outcome.

CASE REPORT

A 26-year female presented to the ENT outpatient department with chief complaints of ringing sensation in left ear for a duration of 2 months. Patient was apparently normal 2 months ago, when she developed, ringing sensation (gushing sound) in left ear, which was synchronous with the heartbeat. It was insidious in onset, intermittent, progressive and more noticeable during night hours and was associated with sleep. There were no other otological symptoms like ear discharge, giddiness and decreased hearing. No aggravating or relieving factors. She also had a left earache since 2 months, sharp shooting pain, intermittent type, radiating from behind the ear to the left side of neck. She didn't have any history of trauma or surgery in the past, and no known comorbidities.

A detailed otological examination was conducted which revealed bilateral intact and normal tympanic membranes. Tuning fork tests were normal on both the sides. On auscultation of the mastoid, a vascular bruit was heard over left mastoid.

PTA showed (Figure 1) normal hearing in both ears. Impedance audiogram showed "A" type curve. Blood investigations were done, which showed CBC within normal limits, vitamin B12 levels were 128 pg/ml and Homocystiene levels were 120 mcmol/L (severe raise).

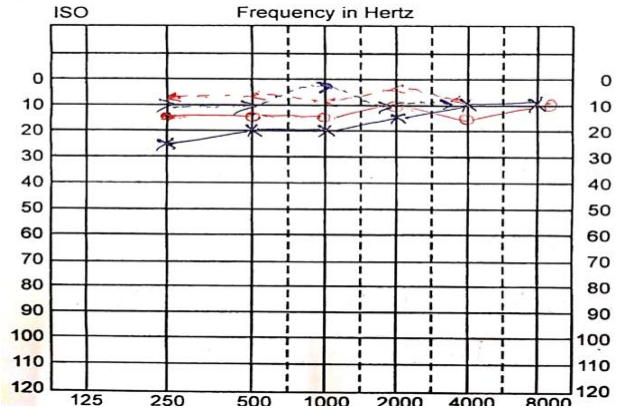

Figure 1: PTA of normal hearing in both ears.

Patient was further investigated with HRCT temporal bone which showed thinning and erosions of left mastoid bone with bony dehiscence, along the lateral aspect of sigmoid sinus with a prominent emissary vein foramen adjacent and posterior to left mastoid bone (Figures 2-4). MRI brain of the patient showed luminal irregularities in distal most part of left transverse sinus, left transverse sigmoid junction.

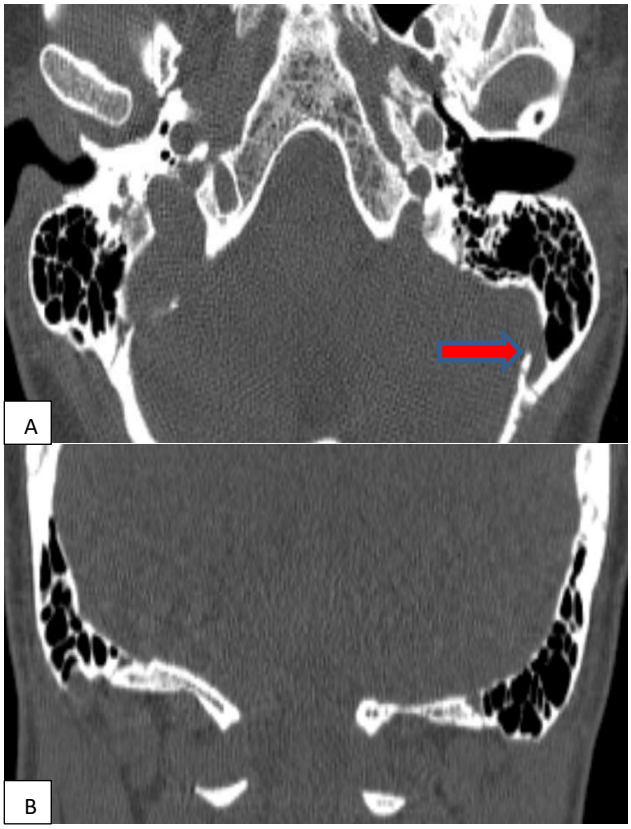

Figure 2 (A and B): HRCT temporal bone-axial view-showing thinning and erosion of medial wall of left mastoid sinus (Red arrrow) and coronal view-thinning of medial wall of left mastoid sinus.

Figure 3: HRCT temporal bone-A prominent emissary vein foramen posterior to the left mastoid sinus.

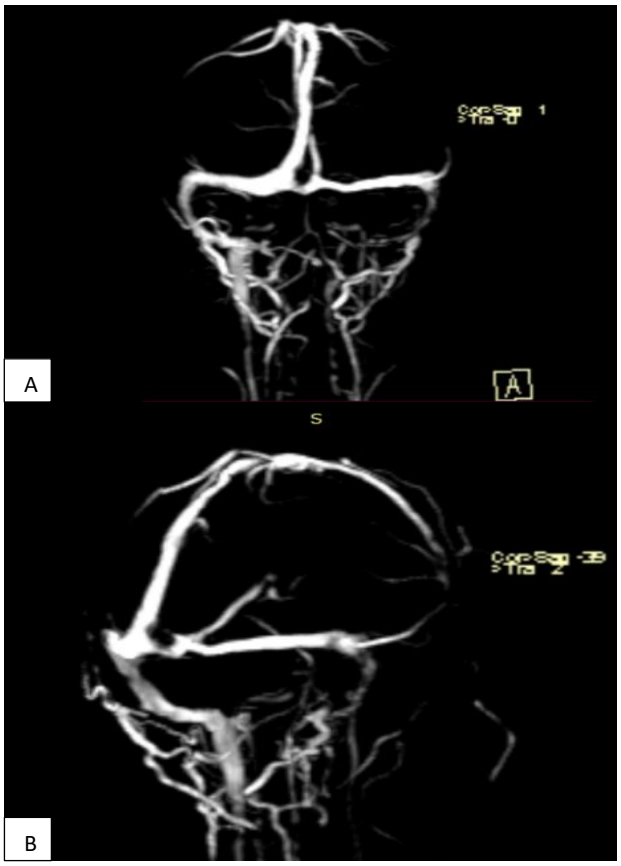

Figure 4 (A and B): MRV-thinning of left sigmoid sinus.

Patient was further evaluated with MRA and MRV which showed the following findings: 1) Multiple abnormal flow voids along the left sigmoid and transverse-sigmoid junction. 2) Dilated left external carotid artery branches with hypertrophied meningeal branches of left cavernous internal carotid artery. 3) Irregular filling defects and narrowing in the left sigmoid sinus which were suggestive of chronic venous thrombosis. 4) Prominent left sided vein of Labbe. 5) Prominent left venous sinus and torcula, suggestive of venous reflux.

Imaging features of MRV were suggestive of left transverse sigmoid dural arterio-venous fistula with multiple feeders from left ECA and ICA and venous reflux.

Patient was further subjected to 3 dimensional DSA which showed: 1) Type IIa (Cognard) dural arterio-venous fistula involving left transverse sigmoid sinus junction with venous reflux and left ICA, MMA, occipital artery feeders were noted (Figure 7). 2) Patient thereafter underwent-endovascular embolization of the left transverse sigmoid dural arteriovenous fistula done through femoral artery and vitamin B12 oral supplementation was given. 3) Patient was followed up after 2 weeks and subsequently after 12 weeks at which time she was completely symptom free.

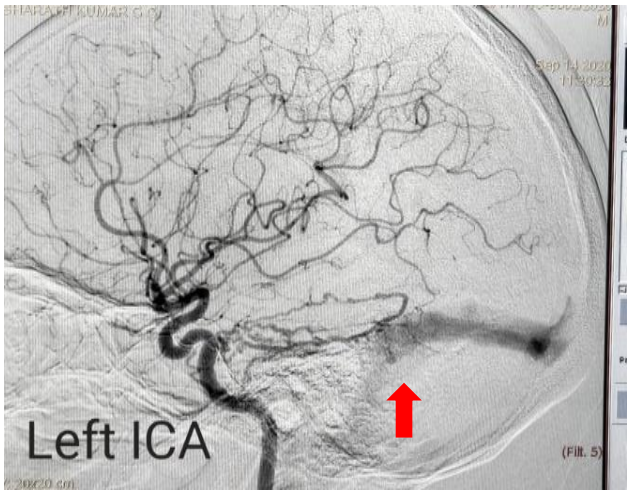

Figure 5: MR angiography-multiple abnormal flow voids along the left sigmoid and transverse-sigmoid junction.

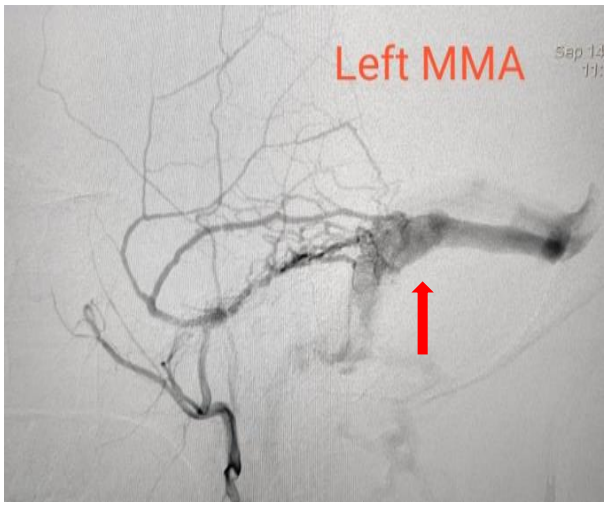

Figure 6: MRA-Dural arterio-venous reflux.

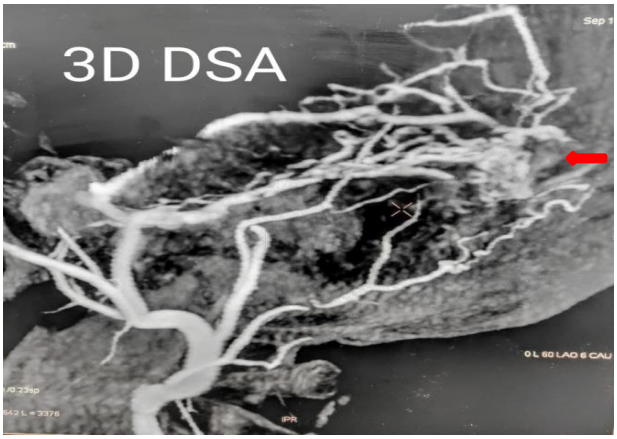

Figure 7: 3D DSA.

DISCUSSION

Pulsatile tinnitus has a myriad and varied spectrum of causes which can be accurately assessed only with a detailed clinical examination and an extensive diagnostic workup. The cause can be non-vascular like myoclonus and vascular such as, DAVF, arterio-venous malformation or aneurysm.[9] In transverse sigmoid DAVF (TSDAVF), the common causes of fistula formation are congenital vascular malformations, traumatic causes and pregnancy. Although DAVFs constitute only 10% of vascular causes; of all intracranial DAVFs, those that drain into the transverse and sigmoid sinus are the most common.[10-13] Fortunately, these types also have the most benign course.

Our patient was a young female, vegetarian by diet with no history of trauma. Her blood work up showed low Vitamin B12 levels, which is a cause for elevated levels of homocystiene. These elevated levels might have further caused the stenosis thereby resulting in fistula formation. Hence the probable cause of TSDAVF in our patient might be raised homocystiene levels.

Accurate diagnosis in pulsatile tinnitus is essential because in the majority of cases a treatable underlying aetiology can be identified. Failure or delay in the diagnosis of TSDAVF, or ascribing a diagnosis of exclusion, such as benign intracranial hypertension, in the face of true intracranial vascular malformation can have catastrophic consequences. TSDAVF is a dynamic disease that can be associated with significant morbidity and death. Studies have shown the crude risk of haemorrhage for DAVF to be 2% per year.[14] Moreover, delay in treatment often results in additional morbidity and higher-risk TSDAVF.[10,15]

There are other clinical manifestations such as vertigo, headache or ophthalmological symptoms that could be related to inner ear or intracranial pathology. It is also essential to include a complete personal clinical history and also the use of any frequent medications.[16,17] Complete physical examination besides the ORL explorations; which includes auscultation of the retro-auricular and orbital region looking for audible murmurs or tinnitus. Otoscopy findings in DAVFs are usually normal; however, the presence of a reddish trans-tympanic mass would lead to the suspicion of a jugulotympanic glomus, an aberrant or ectopic carotid or a proximal jugular bulb. Completing the evaluation with a PTA and tympanometry helps us to recognize the possibility of audiological involvement.[16,17]

Supportive investigations like 70% of the brain CT and 80% of the cerebral MRI done in patients with DAVF show suggestive alterations, which must be confirmed with more specific techniques such as a Doppler ultrasound, magnetic resonance angiography or a computed tomography angiography.[8] However, the definitive diagnosis is given by DSA, a procedure also performed towards a therapeutic management: Endovascular embolization. The appearance of knots in arteriography is pathognomonic of an arteriovenous malformation.[8,11,18-21]

Thus, accurately defining the extent of TSDAVF is important not only in guiding therapy, but also in counselling the patients on potential risks of therapeutic endeavours. The evaluation of pulsatile tinnitus, particularly with normal otoscopy, demands early angiography, should MRA be negative. In cases of normal otoscopy where such anomalies as a high-riding jugular bulb, glomus tumours, or even serous effusions are less likely, we advocate moving directly to MRI/MRA over CT scanning. As seen in our case, MRA, in conjunction with DSA, enhances diagnosis with vascular aetiology of pulsatile tinnitus including TSDAVF.

Sequele of TSDAVF is depression and fatal complications such as intracranial haemorrhage. In any patient with pulsatile tinnitus, a thorough clinical and radiological evaluation can prevent further fatal complications.

According to Borden and Cognard classification for grading DAVF has 3 types: Type I is a venous drainage into dural sinus with antegrade flow and the probability of associated complications is 2%. Type II and type III of Borden classification are associated with 39-79% of probable complications.[22]

Treatment must be individualized and patients with Borden type I and type I/IIa Cognard DAVFs, with controlled symptoms, should be followed periodically with imaging tests, with the purpose of recognizing any worsening/ change in their clinical condition. For the most severe and/or most symptomatic cases, endovascular embolization (arterial or venous) is the treatment of choice. The embolization of the drainage pathway is now recognized as important to successful treatment of DAVF. Halbach et al reported complete angiographic cure in 5 of 11 patients who underwent only

trans-venous embolization of the transverse-sigmoid sinus with either coils or glue.[13]

In our case, a 3 dimensional DSA showed: Type IIa (Cognard) Dural arterio-venous fistula involving left transverse sigmoid sinus junction with venous reflux and left ICA, MMA, occipital artery feeders were noted (Figure 7). Thereafter she underwent-endovascular embolization of the left transverse sigmoid DAVF through femoral artery (Figure 8 A and B).

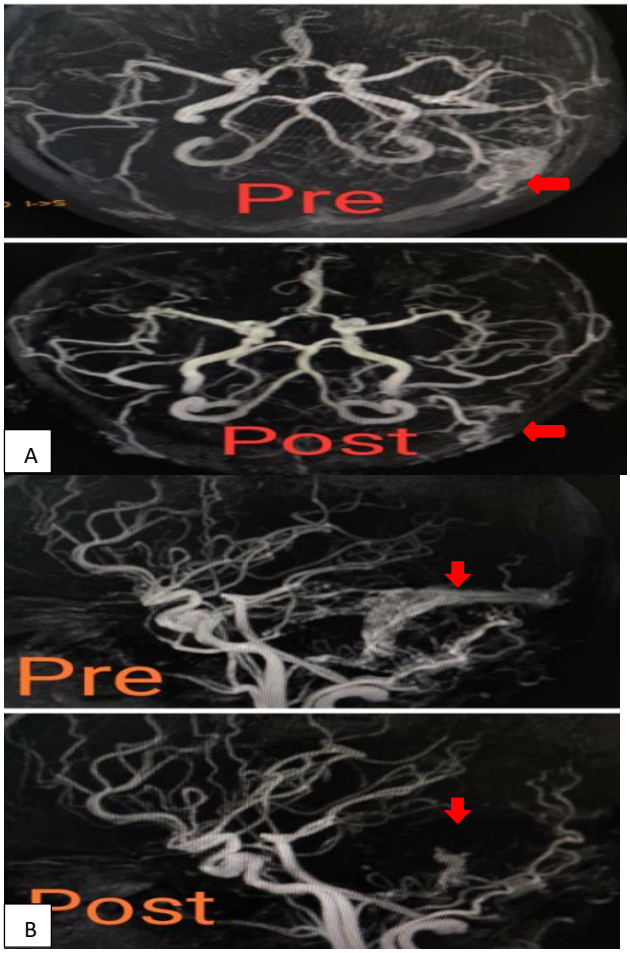

Figure 8 (A and B): Comparison preoperative and postoperative angiography images (circle of Willis).

Recent studies provide overriding evidence to suggest that elevated blood homocysteine levels can cause thrombosis and some 10-20% of coronary heart disease cases have been linked to elevated homocysteine levels.[23] Homocysteinemia may result from vitamin B12, folate and pyridoxine deficiencies which we postulate as a probable cause in our patient.[24]

CONCLUSION

Pulsatile tinnitus has a wide differential diagnosis. Traditionally the evaluation has included-examination, CT and MRI. While the advent of MRA enhances the ability to capture vascular lesions, there remain short-comings which can lead to undiagnosed TSDAVF. Super-selective arteriography with possible adjunctive embolization should be the mainstay in routine evaluation and treatment of pulsatile tinnitus. Early diagnosis and prompt treatment may save the patients from devastating consequences of TSDAVF-associated haemorrhage or the increased morbidity associated with therapeutic endeavours against advanced grade TSDAVF. In our case, a raised Homocystiene level might have been the cause for thrombosis and fistula formation, as there was no history of trauma, or previous medication.

A detailed and comprehensive work up and multimodality approach is mandatory in all patients presenting with pulsatile tinnitus with normal otological parameters, to enable the clinician to arrive at an accurate diagnosis thereby preventing catastrophic outcomes and restoring patient's quality of life.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Hofmann E, Behr R, Neumann-Haefelin T, Schwager K. Pulsatile tinnitus: imaging and differential diagnosis. Dtsch Arztebl Int. 2013;110(26):451-8.
2. Herraiza C, Aparicio JM. Claves diagnósticas en los somatosonidoso acúfenos pulsátiles. Acta Otorrinolaringológica Española. 2007;58(9):426-33.
3. In 't Veld M, Fronczek R, de Laat JA, Kunst HPM, Meijer FJA, Willems PWA. The incidence of cranial arteriovenous shunts in patients with pulsatile tinnitus: A prospective observational study. Otol Neurotol. 2018;39(5):648-53.
4. Kircher ML, Standring RT, Leonetti JP. Neuroradiologic assessment of pulsatile tinnitus. Otolaryngol Head Neck Surg. 2008;139:144.
5. Sismanis A. Pulsatile tinnitus. Otolaryngol Clin N Am. 2003;36:389-402.
6. Levine RA, Nam EC, Melcher J. Somatosensory pulsatile tinnitus syndrome: Somatic testing identifies a pulsatile tinnitus subtype that implicates the somatosensory system. Trends Amplif. 2008;12:242-53.
7. Mattox DE, Hudgins P. Algorithm for evaluation of pulsatile tinnitus. Acta Oto-Laryngol. 2008;128:427-31.
8. Gandhi D, Chen J, Pearl M, Huang J, Gemmete JJ, Kathuria S. Intracranial dural arteriovenous fistulas: classification, imaging findings, and treatment. AJNR Am J Neuroradiol. 2012;33(6):1007-13.
9. Shownkeen H, Yoo K, Leonetti J, Origitano TC. Endovascular Treatment of Transverse-Sigmoid Sinus Dural Arteriovenous Malformations Presenting as Pulsatile Tinnitus. Skull Base. 2001;11(1):5310-1.
10. Lalwani AK, Dowd CF, Halbach VV. Grading venous restrictive disease in patients with dural

arteriovenous fistulas of the transverse/sigmoid sinus. J Neurosurg. 1993;79:11-5.
11. Borden JA, WuJK, Shucart WA. A proposed classification for spinal and cranial dural arteriovenous fistulous malformations and implications for treatment. J Neurosurg. 1995;82:166-79.
12. Davies MA, Saleh J, Ter Brugge K. The natural history and management of intracranial dural arteriovenous fistulae. Int Neuroradiol. 1997;3:295-302.
13. Halbach VY, Higashida RT, Hieshima GB. Dural fistulas involving the transverse and sigmoid sinuses: results of treatment in 28 patients. Radiology. 1987;163:443-7.
14. Brown RD, Wiebers DO, Nichols DA. Intracranial dural arteriovenous fistulae: angiographic predictors of intracranial hemorrhage and clinical outcome in nonsurgical patients. J Neurosurg. 1994;81:531-8.
15. Ishii K, Goto K, Ihara K. High-risk dural arteriovenous fistulae of the transverse and sigmoid sinuses. AJNR Am J Neuroradiol. 1987;8:1113-20.
16. Dietz RR, Davis WL, Harnsberger R. MR Imaging and MR angiography in the evaluation of pulsatile tinnitus. AJNR Am J Neuroradiol. 1994;15:879-89.
17. Newton TH, Cronqvist S. Involvement of dural arteries in intracranial arteriovenous malformations. Radiology. 1993;93:1071-8.
18. An Y-H, Han S, Lee M, Rhee J, Kwon OK, Hwang G. Dural arteriovenous fistula masquerading as pulsatile tinnitus: radiologic assessment and clinical implications. Scientific Rep. 2016;6:36601.
19. Paredes I, Martinez-Perez R, Munarriz PM, Castaño-Leon AM, Campollo J, Alén JF et al. Fístulas durales arteriovenosas intracraneales. Experiencia con 81 casos y revisión de la literatura. Neurocirugía. 2013;24(4):141-51.
20. Cognard C, Gobin YP, Pierot L, Bailly AL, Houdart E, Casasco A et al. Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. Radiology. 1995;194(3):671-80.
21. Chung SJ, Kim JS, Kim JC, Lee SK, Kwon SU, Lee MC et al. Intracranial dural arteriovenous fistulas: analysis of 60 patients. Cerebrovasc Dis. 2002;13(2):79-88.
22. Invêncio Da Costa LF, Amaya FA, Riera MM, Álvarez-Buylla M. A Rare Case of Unilateral Pulsatil Tinnitus. Int Tinnitus. 2018;22(2):156-9.
23. Halbach VV, Higashida RT, Hieshima GB, Goto K, Norman D, Newton TH. Dural fistulas involving the transverse and sigmoid sinuses: results of treatment in 28 patients. Radiology. 1987;163(2):443-7.
24. Edirisinghe SP. Homocysteine-induced thrombosis. Br J Biomed Sci. 2004;61(1):40-7.
25. Engman M. Homocysteinemia: new information about an old risk factor for vascular disease. J Insur Med. 1998;30(4):231-6.

Cite this article as: Narayankar NM, Gowda CBV, Rai SJ. Nutrimentum vascularum as pulsatile tinnitus. Int J Otorhinolaryngol Head Neck Surg 2022;8:280-5.