

CASE REPORT

Unilateral Hypoplasia of Venous System of Brain; A Contraindication to the Ligation of Internal Jugular Vein in the Treatment of Venous Pulsatile Tinnitus

Devrim Bektas, Refik Caylan

Associate Professor of Otolaryngology, KTU Medical School, Trabzon, TURKEY, (DB)

Professor of Otolaryngology, Department of Otolaryngology, Ankara Etlik İhtisas Hospital, Ankara, TURKEY, (RC)

Pulsatile tinnitus of venous origin is a relatively rare diagnosis. When it originates from turbulent flow within the transverse-sigmoid-jugular system, the surgeons are often tend to block tinnitus creating venous flow. In the presence of a contralateral underdeveloped venous system surgery, internal jugular vein ligation may lead to fatal consequences. We report a 30-year-old female patient with venous pulsatile tinnitus and contralateral hypoplastic venous system of the brain. In the planning of any surgical method having a major interference with the venous blood flow of the brain, the surgeon should make necessary measures to insure an adequate contralateral venous system.

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Introduction

Pulsatile tinnitus (PT) is a relatively infrequent diagnosis in otolaryngology practice. In most cases it originates from vascular structures within the head and neck area. According to its origin it is classified either as arterial or venous. Venous PT usually can be diagnosed by reduction or cessation of symptoms by internal jugular vein (IJV) compression.^[1] Most common encountered venous etiologies of PT are; pseudotumor cerebri syndrome^[2], jugular bulb abnormalities^[3], hydrocephalus associated with the stenosis of the sylvian duct^[4], Arnold-Chiari malformation^[4], abnormal condylar and mastoid emissary veins^[5,6], narrowing of the transverse dural venous sinus^[3], and idiopathic tinnitus (Venous hum tinnitus)^[2,7]. Most of the defined treatments for venous PT in the literature include impeding the turbulent blood flow causing noise detectable from the ear.^[8-10]

In cases which the noise becomes intolerable, ligation of the IJV is the most common used treatment.^[8] Development of contralateral tinnitus^[11] or recurrence of tinnitus^[12] are not the worst scenarios when the

treatment choice is blockage of venous flow of the tinnitus creating side. If the affected side is the only major route for brain's venous drainage such as in a case with contralateral hypoplastic or atresic transverse-sigmoid-jugular venous system surgery could have fatal consequences.

We report a venous PT in a 30-year-old female patient with an emphasis on the necessity of meticulous investigation of the origin of venous PT before committing an IJV ligation for cure.

Case Report

A 30-year-old female patient presented with a complaint of left-side tinnitus deteriorated in the previous months. Tinnitus had a roaring and crescendo-decrescendo character. The noise especially increased when the patient's head was turned to the right and decreased when the head was turned to the left. On physical examination the patient stated the disappearance of tinnitus on gentle palpation of the left IJV area. On otoscopic examination TM was noted as normal. Auscultation of left mastoid area revealed a humming sound, therefore indicating a jugular bulb

Corresponding address:

Devrim Bektas

KTU Tip Fak. KBB Servisi Trabzon, TURKEY

Phone: 90-533-477-53-73; Fax: 90-462-3258324 E-mail: devrimbektas@yahoo.com

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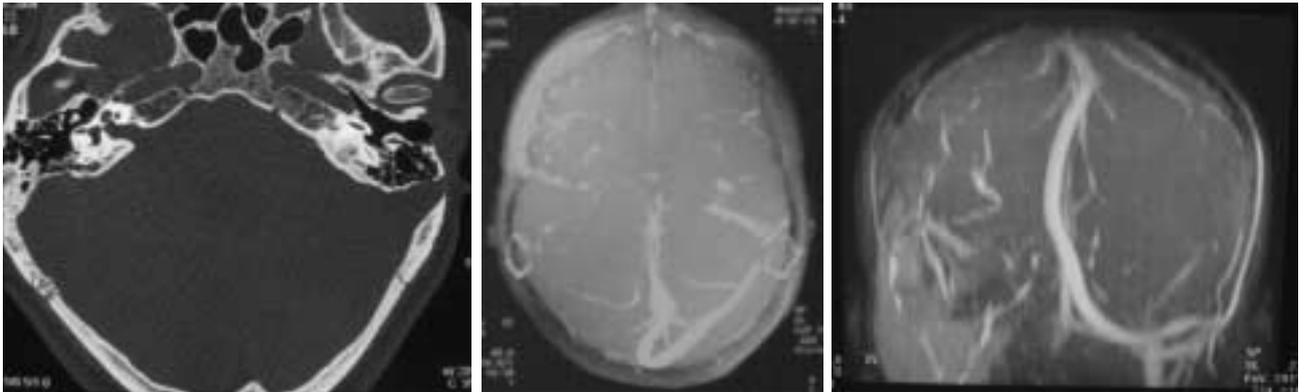


Figure 1. Axial CT image demonstrates laterally placed left sigmoid sinus (tinnitus side) with dehiscent outer mastoid cortex and an underdeveloped sigmoid sinus groove on right mastoid inner cortex (hypoplastic side)

Figures 2-3. Venographic MRI demonstrates hypoplasia of the anterior half of superior sagittal sinus, right transverse-sigmoid sinus junction and right internal jugular vein

originated venous PT. Pure tone audiogram demonstrated a 33 dB mixt hearing loss in the left ear. Computerized tomographic (CT) and Magnetic resonance imaging (MRI) studies were performed. On CT the patient had a dominant and laterally placed left sigmoid sinus with a dehiscent outer mastoid cortex. On MRI cisterna magna was dilated with deep brain sulci and hydrocephalic appearance. MRA was warranted, and the arterial system was found normal with no stenosis/ occlusion findings. Due to these findings flow quantification MRI and venographic MRIs were additionally obtained. Flow quantification MRI demonstrated apparent dilatation in all ventricular systems with increased BOS dynamics. On venographic MRI hypoplasia of the anterior half of superior sagittal sinus, right transverse-sigmoid sinus junction and right internal jugular vein were detected. Due to this gross pathology of the venous system, no surgical intervention was considered.

Discussion

Venous originated PT is a rare finding. When suspected, a gentle compression of IJV leading to disappearance of symptoms generally confirms the diagnosis.^[1] CT is generally helpful in the differential diagnosis of venous lesions such as dominant jugular bulb, laterally placed sigmoid sinus and otoscopically visible intratympanic mass. If a bony destruction in jugular foramen is detected in MRI, contrast-enhanced

CT is warranted to exclude a glomus jugulare tumour. MRV is useful in demonstration of the anatomy of dural sinuses.^[13]

Vascular PT generally originates from disturbance in the laminar blood flow either by increased flow or lumen stenosis.^[8,14,15] Since venous PT is usually due to an increased flow within the transverse-sigmoid-jugular venous system, treatment choices usually include unilateral interference of brain's venous drainage.^[16] Treatment options of the venous PT include mastoidectomy (when filled mastoid cells were the reason for increased transmission of sound from venous blood flow to the ear)^[8], ligation of the IJV^[7,10] endovascular coil obliteration of diverticulum of sigmoid sinus^[9], transmastoid reconstruction of sigmoid sinus (to provide a smooth reconstructed wall to eliminate venous turbulence in the presence of sigmoid sinus diverticulum)^[17] and diverting high jugular bulb via a retrosigmoid infralabyrinthine or subfacial approach^[18,19].

When a treatment option which is going to have a major interference with the venous blood flow of the brain is selected, the surgeon should rule out an ineffective contralateral venous system due to hypoplastic or even atresic dural sinuses. Disturbance in the hemodynamics of brain circulation could lead to acute pseudotumor cerebri and other disturbing neurologic conditions such as progressive visual

loss.^[16] The authors of this case believe that this is an understated notion in the literature which surgeons should be beware of.

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