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SIGMOID SINUS DIVERTICULUM WITH PERISINUS PNEUMATIZATION CAUSING PULSATILE TINNITUS: A RARE ENTITY

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Tinnitus is a bothersome symptom with various etiologies. Pulsatile tinnitus constitutes only a small percentage of tinnitus types and is due to a vascular pathology close to the inner ear which causes a sensation of noise.

Many vascular lesions are associated with pulsatile tinnitus: paraganglionoma, persistent stapedial artery, a vascular loop in the internal auditory meatus or cerebello-pontine angle, carotid-vertebral stenosis, and arterio-venous malformation.

We report one case of pulsatile tinnitus that originated in a venous diverticulum of the sigmoid sinus and which was transmitted to the inner ear through perisinus pneumatisation. Treatment options for this condition are discussed, with insights from the literature.

Key words: pulsatile tinnitus (PT), venous hum, sigmoid sinus diverticulum (SSD), endovascular coiling, sigmoid sinus wall reconstruction, transmastoid surgery, sigmoid sinus wall dehiscence (SSWD), sinus decompression, idiopathic intracranial hypertension (IIH).

DIVERTÍCULO DEL SENO SIGMOIDEO CON NEUMATIZACIÓN PERISINUSAL CAUSANTE DE TINNITUS PULSÁTIL

Resumen

Los tinnitus son fenómenos curiosos con etiología diversa. El tinnitus pulsátil constituye un porcentaje muy pequeño del total de los casos y está producido por una patología vascular dentro del oído interno, percibida como ruido.

Muchas de las alteraciones vasculares se asocian con el tinnitus pulsátil, entre otras: paragangliomas, arteria estapedial persistente, lazos vasculares dentro del conducto auditivo interno o ángulo pontocerebeloso, estenosis de la arteria carótida y vertebral, así como malformaciones arteriovenosas.

Se presenta un caso de tinnitus pulsátil originado en el divertículo venoso del seno sigmoideo, trasmitido al oído interno por neumatización perisinusal. Los posibles tratamientos se comentan tomando en cuenta los apuntes encontrados en la bibliografía relevante.

Palabras clave: tinnitus pulsátil (PT), zumbido venoso, divertículo del seno sigmoideo (SSD), reconstrucción de la pared del seno sigmoideo, mastoidectomía, dehiscencia de la pared del seno sigmoideo (SSWD), descompresión de los senos, hipertensión intracraneal idiopática (IIH).

ДИВЕРТИКУЛ СИГМОВИДНОГО СИНУСА С ОКОЛОСИНУСНОЙ ПНЕВМАТИЗАЦИЕЙ, ВЫЗЫВАЮЩИЙ ПУЛЬСИРУЮЩИЙ ШУМ В УШАХ

Абстракт

Шум в ушах является интересным обстоятельством, которое может возникнуть вследствие разного рода этиологических факторов. Пульсирующий шум в ушах составляет небольшой процент всех случаев шума в ушах и возникает по причине сосудистой патологии в области внутреннего уха, воспринимающего её как шум.

Большое количество сосудистых изменений связано с пульсирующим шумом в ушах, в частности параганглиома, персистирующая **стременная артерия**, сосудистые петли во внутреннем слуховом проходе или мостомозжечковом угле, стеноз сонной и позвоночной артерии, а также артериовенозная мальформация.

Мы представляем случай пульсирующего шума в ушах, возникшего в венозном дивертикуле сигмовидного синуса, передающегося во внутреннее ухо благодаря околосинусной пневматизации. Возможности лечения обсуждены с учётом замечаний, содержащихся в соответствующей литературе.

Ключевые слова: пульсирующий шум в ушах (PT), вихревые венозные шумы, дивертикул сигмовидного синуса (SSD), эндоваскулярная эмболизация, реконструкция стенки сигмовидного синуса, мастоидектомия, дегисценция стенки сигмовидного синуса (SSWD), декомпрессия синусов, идиопатическая внутричерепная гипертензия (IIH).

UCHYŁEK ZATOKI ESOWATEJ Z OKOŁO ZATOKOWĄ PNEUMATYZACJĄ POWODUJĄCA PULSUJĄCE SZUMY USZNE

Streszczenie

Szumy uszne są ciekawym przypadkiem, wynikającym z różnych czynników etiologicznych. Pulsacyjny szum w uszach stanowi niewielki procent wszystkich przypadków i występuje z powodu patologii naczyniowej w obrębie ucha wewnętrznego, odbierającego je jako szum.

Wiele zmian naczyniowych jest związanych z pulsacyjnymi szumami usznymi, m.in. przyzwojak, przetrwała tętnica strzemiączkowa, pętle naczyniowe w przewodzie słuchowym wewnętrznym lub kąt mostowo-móżdżkowy, zwężenie tętnic szyjnej i kręgowej oraz malformacje tętniczo-żylne.

Przedstawiamy przypadek pulsacyjnych szumów usznych powstałych w uchyłku żylnym zatoki esowatej, przekazywanych do ucha wewnętrznego dzięki pneumatyzacji wokół-zatokowej. Możliwości leczenia omówiono uwzględniając uwagi zawarte w odpowiedniej literaturze.

Słowa kluczowe: pulsacyjne szumy uszne (PT), buczenie żylne, uchyłek zatoki esowatej (SSD), rekonstrukcja ściany zatoki esowatej, mastoidektomia, ubytek sciany zatoki esowtej (SSWD), dekompresja zatok [sinus decompression], idiopatyczne *nadciśnienie śródczaszkowe* (IIH).

Introduction:

Tinnitus is a common otological symptom, although pulsatile tinnitus (PT) accounts for only a small subset (approximately 4%) of the entire population with tinnitus [1]. Venous PT is more common than arterial, and this can be caused by numerous anomalies and/or variants. The condition has a significant negative impact on the quality of life of many patients. Surgical and endovascular interventions have proven successful for some lesions [1].Eisenman et al. have described clinical evaluation of patients with tinnitus and, depending on its etiology, possible treatments, which include surgical options[2].

Sigmoid sinus diverticulum (SSD) is a new and increasingly recognized treatable cause of PT. It occurs when the sigmoid sinus wall locally intrudes into the mastoid cells of the cortex[10]. Mehall et al. reported an early case in 1995[10] when SSD was considered an uncommon cause of PT. Since then, an increasing number of PT patients with SSD have been reported, and it is now the most common identifiable cause of venous-derived PT[10]. Sigmoid sinus wall dehiscence (SSWD), also known as sigmoid plate dehiscence, refers to a local bone wall defect that leads to direct contact between the sigmoid sinus and the surrounding mastoid cells.[10] SSD and SSWD are closely related and can be present alongside each other in patients with PT[17]. Two approaches have been developed to treat PT patients with SSD: endovascular coiling[14] and transmastoid surgery. [2,15,17] The first description of a transmastoid surgical approach was by Otto and colleagues in 2007[11]; since then a number of others have followed suit using various techniques and materials with generally successful outcomes[3,8,13,15,17].

In this case report we describe a unilateral sigmoid sinus diverticulum associated with perisinus pneumatization in a fit and well woman who presented with a one-year history of intrusive pulsatile tinnitus and was treated by surgical decompression. We describe this rare entity along with a review of the relevant literature.

Case Report

A 57-year-old woman presented with subjective unilateral persistent and progressive intrusive PT in her left ear, which had come on suddenly one year earlier without any known predisposing factors. Following the onset, the PT had progressively worsened and she was clearly struggling

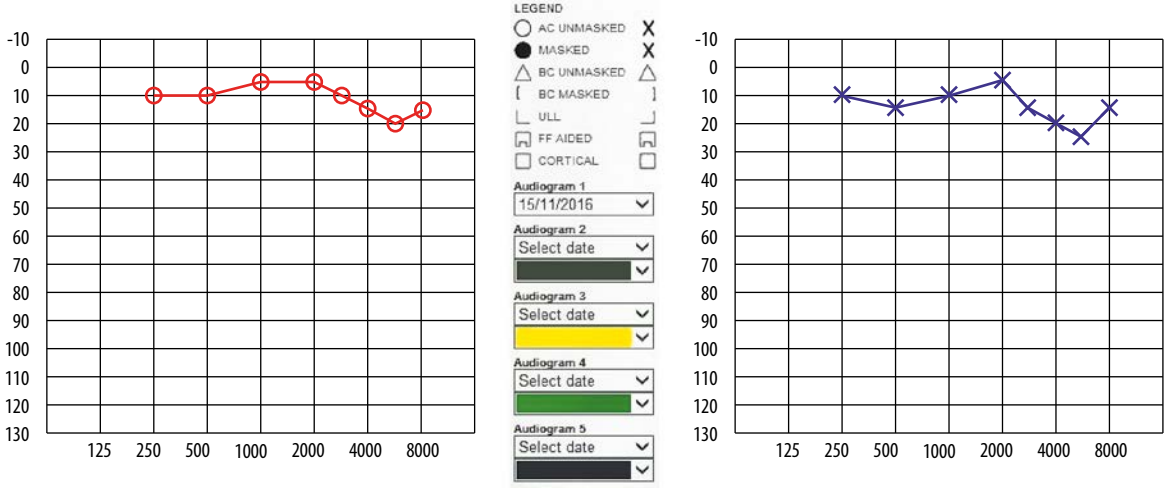

Figure 1. Pure tone audiometry of the patient showing bilateral symmetrical hearing within normal limits

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to cope. She said that the PT worsened when she lay on her left side, and it was mildly relieved with gentle compression of the ipsilateral neck, suggesting compression of the carotid sheath. There were no other neuro-otological symptoms and her speech and swallow were normal. She had no systemic vascular illnesses or familial history of paraganglionomas. Her past medical history was also unremarkable, except for well-controlled asthma and insomnia due to her intrusive tinnitus (for which she had been taking antidepressants for the past year). She scored her symptoms as 90% severity on the Tinnitus Handicap Index, meaning debilitating tinnitus aurium.

Examination revealed both tympanic membranes were healthy and all lower cranial nerves normal. There were no abnormal masses visible or palpable in her neck. There was no palpable thrill or audible bruit on the area over her left neck where she preferred to compress to get some relief. The only positive clinical finding was a faint hum which was audible on auscultation over her mastoid region; this prompted a suspicion that her intrusive PT may be due to a vascular malformation of the temporal bone. A pure tone audiogram and tympanometry were normal.

An MRI of the skull base and MRA of the head and neck was performed to rule out any evidence of vascular malformations or dural venous sinus stenosis. This demonstrated a subtle narrowing on the left side of the sigmoid sinus–transverse sinus junction. A CT venogram and an MRI of the internal acoustic meatus with contrast were then recommended by the neuro-radiologists to aid in further determining the origin of the tinnitus. These demonstrated a small sigmoid sinus diverticulum with perisinus pneumatisation around it, which was apparently transmitting the venous hum into the well-pneumatised mastoid air cells. This was then heard in the ear as the PT which the patient was experiencing. The patient's condition was discussed at the regional skull base multi-disciplinary team meeting (MDT); the consensus was to first rule out idiopathic intracranial hypertension (IIH) leading onto the diverticulum formation, since the patient was obese (BMI of 34.9). She was referred to a neuro-ophthalmologist and a neurologist who ruled out IIH with fundoscopy and spinal tap respectively.

The patient's condition was again discussed at the skull base MDT and two options for management were recommended. These were discussed with the patient: 1) surgical decompression of the sigmoid diverticulum with resurfacing and removal of the perisinus pneumatisation to cut off the transmission of signals; and 2) an interventional radiological procedure of placing an endovascular stent for the diverticulum to correct the turbulent venous flow (or its obliteration with endoluminal coiling). The patient preferred endovascular coiling; however, upon review of her MRA, the neuroradiologists decided not to pursue this avenue since the diverticulum was located at the sigmoid– transverse sinus junction, so that coiling would be very difficult and complete obliteration of this vital junction could lead to complications with a high risk of failure. Hence, they recommended that the first surgical approach was the most appropriate, and the MDT concurred.

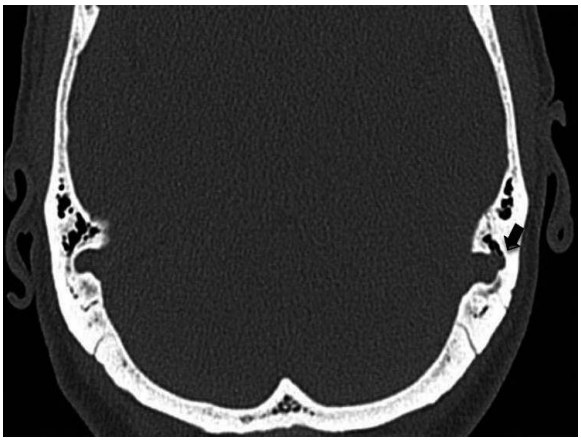

Figure 2. CT axial image showing the left sigmoid sinus diverticulum with perisinus pneumatisation (black arrow), which was thought to cause pulsatile tinnitus to be transmitted to the ear

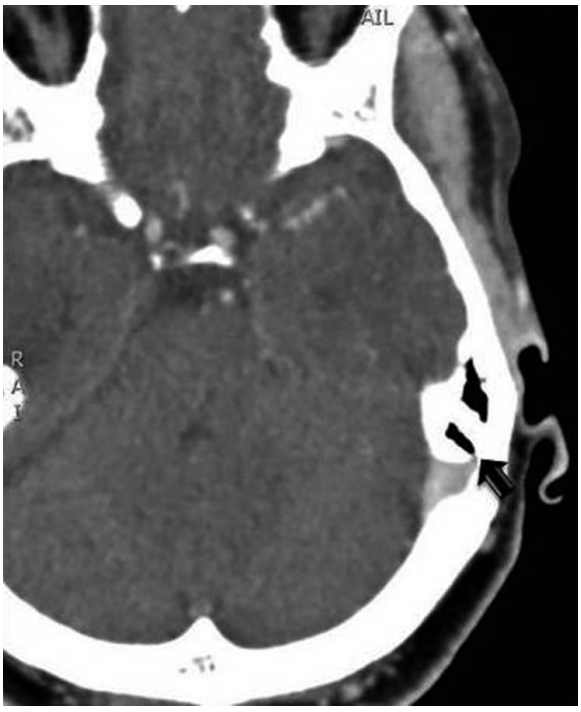

Figure 3. CT venogram axial image showing enhancement of left sigmoid diverticulum (black arrow)

The patient underwent successful transmastoid exploration and occlusion of the diverticulum. The diverticulum was fully decompressed by careful dissection of the adjoining perisinus pneumatisation, and bone paste with wax and fibrin glue was used to completely occlude its lumen, thereby blocking the tinnitus which had occurred due to the turbulent blood flow. The patient had a remarkable recovery in the immediate post-operative period with her tinnitus almost disappearing completely (her THI score was now 15%). She has been monitored over the past 4 months for any recurrence of PT at the neurotology service of our tertiary referral centre, but continues to be asymptomatic.

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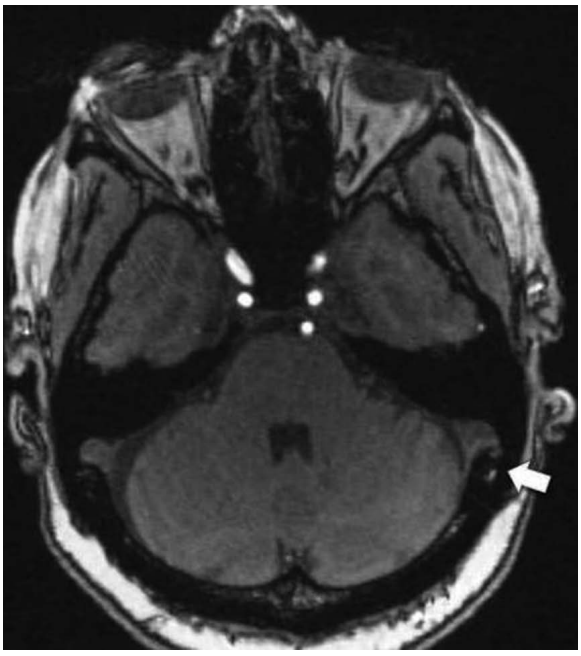

Figure **4**. MRI skull base T1W with contrast axial image showing the tortuous sigmoid diverticulum (white arrow)

Discussion

Tinnitus is a common otological symptom that affects 10–15% of the population worldwide[7]. It can be divided into pulsatile and non-pulsatile, with pulsatile patterns suggesting a vascular cause. Only 4% have vascular PT caused by rare venous abnormalities that transmit sound to the cochlea by bony or vascular structures[1]. PT is likely to be of arterial origin when symptoms worsen with gentle compression of the ipsilateral internal jugular vein, whereas it is likely to be venous if it is improved with the same manoeuvre[14]. The psychological impact of pulsatile tinnitus on many patients is so severe and incapacitating it can lead to depression or even suicide[16]. There are many causes of pulsatile tinnitus, although the sigmoid sinus diverticulum is one of the most common[1].

Sigmoid sinus wall anomalies (SSWA) include sigmoid sinus diverticulum and dehiscence, although a definitive classification scheme is lacking[2], and the precise etiology is not fully understood. Studies have shown that venous PT is more frequently found in female subjects (88.8%)[1]. This is thought to be likely associated with benign/ idiopathic intracranial hypertension (IIH) that is known to be more common in women with high BMI and can present with PT as the only symptom.

Harvey et al[6]. and Grewal et al[4]. have shown that many subjects with PT are women with elevated BMI, similar to the population affected by IIH. The median onset of PT is typically late middle age[6,13], which suggests that acute pressure erosion or a congenital defect in the sigmoid sinus is unlikely (since the sigmoid sinus is a low-pressure system)[6,13]. As in previous reports, all diverticulums were seen to develop in a well pneumatised mastoid cavity, as was seen with our patient[13]. However, Wenjuan et al. demonstrated that the magnitude of bone pneumatisation does not significantly differ between PT patients with SSD and healthy individuals[16]. A case report by Hou et al[8]. discussed PT as perhaps being an early manifestation of osteoporosis, or osteopenia due to the reduction of oestrogen and progesterone in the perimenopausal period.

When diagnosing PT and considering treatment modalities it is important to consider the cause of PT. Cheng et al[1]. retrospectively analysed contrast-enhanced CTs in 242 patients with unilateral venous PT to assess the various vascular anomalies and variants associated with venous pulsatile tinnitus. They found that 70% had more than one anomaly or variant on the symptomatic side, while 24% had only one anomaly or variant. This suggests that multiple lesions within the veins may trigger the sound of venous PT.[1] The most commonly found lesions were dehiscent sigmoid plate (86.4%), lateral sinus stenosis (55.8%), high jugular bulb (47.1%), and sigmoid sinus diverticulum (34.3%). It is thought that venous sinus stenosis and a high riding jugular bulb with diverticulum are associated with turbulent blood flow near the middle ear, which in turn is responsible for the objective PT[14]. It has been noted that SSD is mainly found on the side of the dominant brain venous systems, as was the case in our patient[1,9,14].

There are currently two main modalities described for treatment of PT: surgical or endovascular intervention. Endovascular treatment aims to embolize the diverticulum with coiling or stenting by correcting the turbulent flow of the SSD. Risks include coil migration, increase in intracranial pressure, and thrombosis (which typically necessitate anticoagulation in the perioperative period)[15]. Endovascular approaches also fail to repair SSWD, which is a significant cause of PT and has been shown to co-exist with SSD[15]. Transmastoid surgery aims to excise the SSD and repair the bony wall dehiscence using autologous or artificial materials. Risks are less than with the endovascular approach, as there is no surgery on blood vessels and no anticoagulation is needed in the perioperative period[15].

Sigmoid sinus wall reconstruction was first reported in 2007 by Otto et al[11]. for three patients with SSD and involved reconstructing the sigmoid sinus wall via the extraluminal placement of either the temporalis muscle and fascia or bone wax. Eisenman et al.[3] successfully used a similar surgical technique; however they used a soft tissue graft of temporal fascia between the dura and the posterior fossa bony plate to reconstruct the soft tissue sinus wall. Sing et al. presented a case series of PT from the sigmoid sinus diverticulum and the middle cranial fossa diverticulum that were successfully treated with transmastoid resurfacing of the venous wall with firm materials (including bone wax, harvested autologous cortical bone chips, autologous bone paste, and fibrin glue)[13]. Guo et al. showed that mechanical compression of the sigmoid sinus is an effective treatment, even if it may be accompanied by sigmoid sinus diverticulum[15].

PT needs to be evaluated thoroughly as it is likely to be caused by more than one lesion. Failing to address all the causes explains why some patients experience no relief or report recurrence after surgery or coiling, since the chosen intervention may address only one cause[1]. No randomized controlled trial data is available to decide which intervention

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is better. More literature is now available on the endovascular approach, mainly due to recent advancement in the interventional radiology field. Since the condition is rare, it is hard to find a large surgical case series from a single centre to support surgical intervention as the primary modality of management. Hence, it is prudent to discuss such cases in a skull base MDT to arrive at a consensus.

Conclusion

Sigmoid sinus diverticulum is a very rare entity which can lead to disturbing pulsatile tinnitus. It is vital to use appropriate high-resolution imaging to clearly diagnose the condition; it is also important to rule out co-existent lesions and idiopathic intracranial hypertension which can contribute to symptoms. All such cases need multidisciplinary input (as was done in our case) in order to weigh up the various treatment modalities and choose the safest option. As experienced in the case of our patient, the surgical approach was a safe option which did not require long-term anticoagulation. There has been a good outcome with immediate resolution of the pulsatile tinnitus. Long term surveillance for recurrence is continuing and remains paramount.

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