

## CASE REPORT

# Pulsatile tinnitus caused by a dilated left petrosquamosal sinus

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## Abstract

The emissary veins, like the petrosquamosal sinus (PSS), are residual valveless veins, which connect the intracranial dural venous sinuses and the extracranial venous system. Rarely, they may cause pulsatile tinnitus (PT). A 22-year-old woman developed in the first week of puerperium worsening headaches, vomiting, and diplopia, and the accentuation of a PT in the left ear that she presented for eight years. The clinical examination findings nine days after delivery were unremarkable, with the exception of a left sixth nerve palsy, and a peculiar sensibility of the left temporo-mandibular joint. High-resolution computed tomography (HRCT) revealed an osseous canal in the air cells of the left temporal bone compatible with a PSS. CT and magnetic resonance (MR) imaging/MR-venogram detected signs of thrombosis of the superior sagittal sinus, and of the left lateral sinus. Laboratory tests revealed severe inherited thrombophilia. We used antithrombotic therapy (body weight-adjusted subcutaneous low-molecular weight heparin for three weeks, followed by indefinite therapy with warfarin), and the headaches, vomiting, and diplopia resolved within four days of treatment. A follow-up MR-venogram performed two weeks later indicated complete recanalization of the intracranial dural venous sinuses. The PT was improved after two weeks of medical therapy, so she could adapt to it without intervention on the PSS. The early initialization of an efficient medical therapy had a great impact on her favorable evolution. PSS could be identified in her case on HRCT.

**Keywords:** petrosquamosal sinus, pulsatile tinnitus, dural sinus thrombosis, inherited thrombophilia, puerperium.

## Introduction

Tinnitus is a perception of sound in the ear, generally without corresponding external sound (subjective tinnitus). It may be pulsatile and synchronous with the patient's heartbeat or non-pulsatile and continuous [1, 2]. Several vascular conditions can lead to pulsatile tinnitus (PT), such as arterial bruit, venous hum, arteriovenous malformations, and vascular tumors [1, 2]. Dilated emissary veins (EVs) may also cause tinnitus [3–6]. Posterior fossa EVs pass through cranial apertures and participate in extracranial venous drainage of the posterior fossa dural sinuses, in addition to the internal jugular vein (IJV), or instead of this vein if it is aplastic or thrombosed [1, 6]. EVs are usually small and asymptomatic in healthy people [1–6].

We report here an extremely rare case of a patient with unilateral PT induced by an abnormally large, prominent posterior fossa EV, called the petrosquamosal sinus (PSS). It courses along the petrosquamosal fissure of the temporal bone, connecting the sigmoid or transverse sinus with the external jugular vein (EJV) [3–6]. The main purpose of our paper is to elucidate the mechanism of the PT in our patient, and to show the appearance of the PSS on high-resolution computed tomography (HRCT) of the temporal bone.

## Case presentation

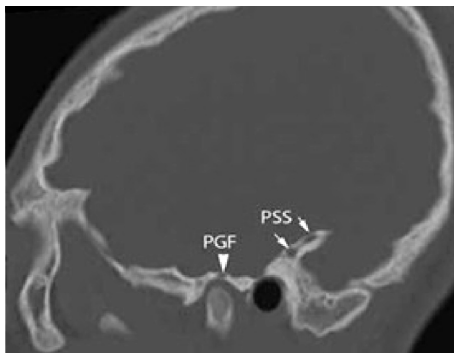
A 22-year-old woman developed worsening headaches

(refractory to ordinary medical treatment), associated with vomiting, and diplopia in the first week after an uncomplicated vaginal delivery. Her first pregnancy had been uneventful, and she had no medical history, with the exception of an associated subjective PT in the left ear for eight years, which had accentuated during pregnancy, and after accouchement. She had no past head or neck trauma, and no exposure to ototoxic drugs or extremely loud noise. We examined her nine days after delivery, following a complex protocol, including neurological, otorhinolaryngological, gynecological, and systemic examination, direct ophthalmoscopy and color fundus photography, high-resolution unenhanced computed tomography (HRCT) scans of the temporal bones, magnetic resonance (MR) imaging/MR-venogram, and laboratory data. CT scans were studied regarding the visibility of the fine sutures, fissures and small canals and the occurrence of vascular anomalies. All the veins and canals of the posterior fossa and both mastoid regions were evaluated by obtaining thin multiplanar reformatted (MPR) images (1 mm collimation, bone window, sharpest filter). This setting allowed good delineation of the emissary veins and venous canals [1–6].

Her blood pressure was 120/70 mmHg. She presented normal visual acuity, and the color fundus photography revealed a bilateral papilledema. Other examination (including gynecological) findings were unremarkable, with the exception of a left sixth nerve palsy, and a

peculiar sensibility of the left temporo-mandibular joint. She did not complain of hearing impairment, or dizziness, and the otoscopic examination and pure-tone audiogram were normal. The PT presented in the form of a high-pitched, “whooshing” sound in the left ear. It was similar with an ear-ringing synchronous with the heart beat, but was not audible even with a stethoscope around the left ear and mastoid area (subjective PT). The PT was heard about 10% of daytime before pregnancy, especially during physically stressful conditions. It became louder during pregnancy, and it was heard about 25% of the day during the first week after delivery. She described attenuation of PT by the Valsalva maneuver, or by manual compression of the left posterior auricular area, or of the left IJV. On the other hand, the PT was unaffected by compression of the right posterior auricular area, or of the right IJV.

HRCT of the temporal bone revealed an unusually distended osseous canal that passed through the superolateral portion of the left temporal bone compatible with a left PSS (Figure 1).



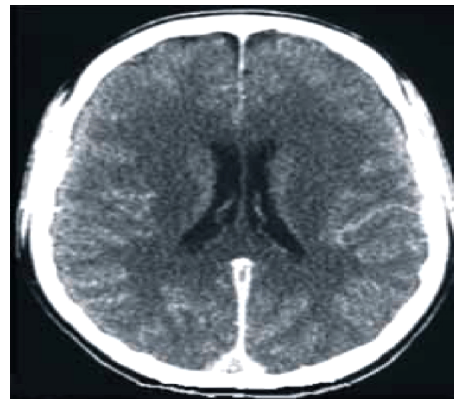
**Figure 1** – *Sagittal multiplanar reformatted (MPR) temporal computed tomography (CT) image shows a petrosquamosal sinus (PSS) sulcus on the left, located on the upper portion of the petrous bone (arrows) and ending in the postglenoid foramen (PGF).*

The diameter of the bony canal containing the left PSS was 2.6 mm. A connection with the left transverse sinus at its junction with the left sigmoid sinus was found in her case. Drainage of the left transverse sinus occurred through this relatively large PSS. The vessel originated on the upper part of the left sigmoid sinus, coursed antero-inferiorly over the superior portion of the left temporal bone and terminated near the posterior part of the left temporo-mandibular joint (TMJ). The anterior-lateral branch of the left PSS exited the skull through a postglenoid foramen (PGF) situated in the left temporal squama (Figure 1).

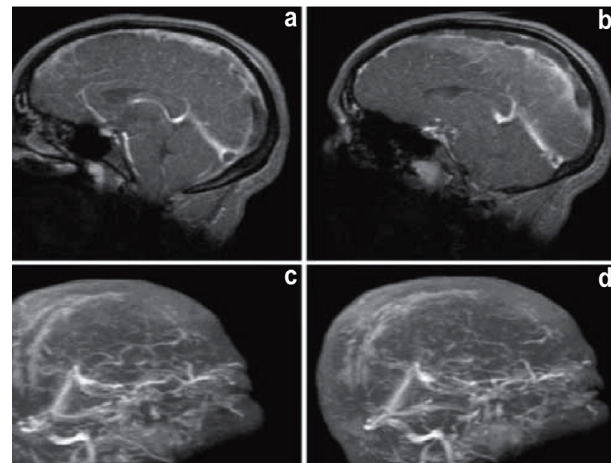
The left jugular foramen was normally developed. The patient had no associated inner ear or middle ear malformations (she presented normal aeration of the middle ear bilaterally).

Unenhanced CT scan, followed by contrast enhancement (Figure 2) and non-contrast-enhanced magnetic resonance (MR) imaging plus MR-venogram (Figure 3, a–d) detected direct and indirect signs of thrombosis of the superior sagittal sinus (SSS), and of the left lateral sinus (LS). MR imaging revealed hyperintense signal in the regions of the superior sagittal sinus (SSS), and the left lateral sinus (LS) [fluid attenuated inversion recovery (FLAIR) images], with concomitant hyperintensity on T1- and T2-weighted

images (the thrombus were bright on both T1- and T2-weighted images). MR-venography showed absence of flow-related signal within the SSS, and the left LS. These imaging results were consistent with dural sinus thrombosis.



**Figure 2** – *Axial enhanced CT image nine days after delivery. Empty delta sign. The thrombus is hypodense within the superior sagittal sinus (SSS), whose walls are clearly enhanced by the injection. (Direct sign of thrombosis of the SSS).*



**Figure 3** – *(a–d) Sagittal magnetic resonance (MR) imaging/MR-venogram. Images 10 days after delivery. SSS and left lateral sinus (LS) thrombosis.*

MR imaging and venography showed the presence and patency of the left PSS, because they identified a patent vein inside this canal, hypointense on T1-weighted images that enhanced homogeneously after contrast medium injection (straight-type PSS).

MR-venography showed a structure with a signal intensity equal to that of the transverse sinus, communicating with the left EJV. The left sigmoid sinus was equal relative to that on the right side. The left jugular foramen was normal. We saw no abnormalities of the skull base.

Laboratory tests revealed an elevated D-dimer, and a severe inherited thrombophilia, represented by homozygous factor V Leiden gene mutation, and heterozygous prothrombin G20210A mutation.

We used antithrombotic therapy [body weight-adjusted subcutaneous low-molecular-weight heparin (LMWH) for three weeks, followed by indefinite oral anticoagulation with warfarin], and the headaches, vomiting, and diplopia resolved within four days of treatment. A follow-up MR-venogram performed two weeks later indicated complete

recanalization of the venous sinuses. The PT was improved after two weeks of medical therapy, so she could adapt to it without intervention on the PSS. The outcome was favorable and the patient did not present any other neurological symptoms/signs during a follow-up of one year.

## Discussion

PT is generally subjective, but sometimes audible (objective) to the examiner by ear or by a stethoscope, which was not our case [1, 2]. It was attributed in our patient to a venous cause, because it had accomplished all three criteria for venous PT: (a) the PT was present in one ear; (b) the PT sound was persistent and synchronous with the heart beat; and (c) the PT was resolved after compression of the IJV ipsilateral to the tinnitus and when turning the head to the PT side [7, 8]. Venous PT is more common than arterial PT, and based on the otoscopic examination and all imaging studies used in our case, other possible vascular etiologies of her PT were excluded, such as fistula, arterio-venous malformations, vascular tumors, etc. [7–10]. On the other hand, venous PT can be caused by numerous multiple vascular anomalies and variants on the symptomatic side: large EV, sinus thrombosis, dehiscent sigmoid plate, lateral sinus stenosis, sigmoid sinus diverticulum, jugular bulb diverticulum, etc. [7, 9, 11, 12]. In our case, it was produced by a large EV, called PSS, and accentuated during pregnancy, and after accouchement by thrombosis of the SSS, and of the left LS. The PSS is an EV of the posterior fossa connecting the intra- and extracranial venous networks; its main drainage pathway is the EJV [3, 4, 6, 13]. On the other hand, the major cerebral venous outflow pathways are represented in adults by the IJV, for supine position, and the vertebral venous system (VVS), for the upright position [3, 6, 14, 15]. According to different authors [6, 14], the venous drainage of the posterior fossa follows highly asymmetric patterns with frequent anatomical variations, especially of the transverse and sigmoid sinuses. The latter are subject to lateral dominance, the right side being frequently more developed than the left side, but also to segmental variations of the transverse and sigmoid sinuses [6, 14] (which was not our case, because the two LS were equal). Regardless, most variations involving the LS allow conservation of the IJV and VVS as their major outflow pathways for encephalic drainage [6, 14].

PSS arises from the dorsolateral portion of the transverse sinus, before its junction with the sigmoid sinus, and before the confluence of the transverse sinus with the superior petrosal sinus [3, 4, 6, 13]. It collects the middle and superficial temporal veins, the masseterine veins, and the internal maxillary veins [3, 4, 6, 13]. San Millán Ruíz *et al.* [6] asserted that, in adults, only the connections of the PSS with the middle meningeal veins and the transverse sinus usually persist (like in our case), while the lateral connection with the deep temporal veins is lost [3, 4, 13]. PSS has two drainage pathways: one anteroinferiorly into the retromandibular vein through the postglenoid foramen (which was identified in our case by HRCT) and the other anteromedially into the pterygoid venous plexus *via* the foramen ovale [3, 4, 13]. While it usually regresses during fetal and early

postnatal life, a dilated PSS occasionally persists into adulthood, like in our female patient.

San Millán Ruíz *et al.* [6] evaluated 13 anatomic corrosion casts of the cerebral venous system. In three instances, they identified a PSS, with a typical appearance of a diploic channel (*i.e.*, rounded irregular contours and a tortuous course), which seems to correlate with the CT scans presented by Marsot-Dupuch *et al.* [4]. According to San Millán Ruíz *et al.* [6], the proximity of the PSS to temporal bone and its small size make its detection by CT angiography (CTA), MR angiography (MRA) or even digital subtraction angiography (DSA) difficult, explaining the discrepancy in the detection of a PSS observed between post-mortem and imaging examinations [6, 16]. Different authors [4, 6] suggested that the PSS is probably amenable to identification by imaging techniques only when it persists as a large channel, or when it is contained within a bony canal. Both conditions were applicable to our patient. In such cases, HRCT and MR imaging can assess both the anatomy and function of PSS accurately. According to different other studies [3, 7, 17], reformatted sagittal and coronal HRCT sections helped us show the origin and pathways of the PSS. MR venography was performed in her case to determine the presence and patency of the left PSS [4]. In our patient, the left identified PSS presented a diameter of 2.6 mm. Marsot-Dupuch *et al.* asserted that the diameter of the PSS is usually as small as that of a silk thread. It occasionally may be as large as 2 to 4 mm [4]. In another study [18], using high-resolution CT venography (HRCTV), Zhao *et al.* noted that the average diameter of the PSS was 1.4 mm. Twenty-nine (74%) temporal bones (TBs) had a PSS origin from the dorsolateral surface of the transverse sinus before its junction with the superior petrosal sinus (position A); three (8%) TBs had a PSS origin from the ventro-inferior surface of the transverse sinus after or before the junction (position B or C); seven (18%) TBs had a PSS without definite origin (position D) [18]. Eighteen (46%) TBs had a PSS course in a lateral bony canal/groove (lateral canal type); 15 (38%) TBs had a PSS course in petrosquamosal fissure (PSF) (PSF type); six (15%) TBs had a PSS course in both (lateral canal/PSF type) [18]. A postglenoid foramen (PGF) was detected in 25 (64%) TBs [18], like in our case. They concluded that HRCTV can mostly identify the characteristics of PSS similar to its anatomical findings and the optimal imaging technique has the potential to improve its clinical management [18]. Although the HRCTV is the superior tool to depict the venous structures, especially those with small diameter and the related bony canals [18], Pekçevik *et al.* [13] believe that this minimally invasive technique usually fails to represent certain collateral venous channels due to the reverse blood flow and the supine patient position. We did not use HRCTV in our case.

Song *et al.* [19] reviewed retrospective medical records and temporal bone computer tomography (TBCT) findings. They found a total of 20 PSS cases. Based on the shapes of PSS demonstrated on TBCT, PSS was classified into tortuous and straight types. The course and thickness of PSS were also investigated. The average thicknesses of PSS between tortuous and straight types were compared. They noted that the mean diameter of the bony canal that PSS courses on TBCT was  $2.57 \pm 0.88$  mm. Its maximal

and minimal diameters were 4.2 and 0.7 mm. The average diameter of tortuous type PSSs ( $3.04 \pm 0.75$  mm) was significantly larger compared with that of straight-type PSSs ( $2.09 \pm 0.76$  mm) ( $p < 0.05$ ) [19], like in our case: 2.6 mm.

Marsot-Dupuch *et al.* [4] suggested that the persistence of a PSS in adults was more frequent in patients with a skull base malformation associated with middle ear and venous anomalies (when the sigmoid sinus is absent or severely hypoplastic). They used HRCT in all cases, complemented by MR venography in three. We found no association with such anomalies in our patient. However, we observed only a dilated left PSS, with no other vascular abnormality seen in the posterior fossa or jugular venous system. Other reports have focused on the potential role of the PSS as an alternative venous drainage pathway for the posterior fossa towards the EJVS system, on its potential clinical significance during otological surgical approaches, or on its implication in the potential spread of septic thrombosis in the otological infections [6]. The possibility of persistent PSS running through the mastoid should be considered if LS thrombosis without marked inflammation and bone erosion is noted, like in our female patient [16]. A disruption in venous drainage through the thrombosed sigmoid sinus and the IJV, speculatively secondary to a venous thrombosis, like in our case, could have led to the anomalous redirection of venous drainage from the partially thrombosed transverse sinus towards the EJVS system through the PSS. Chauhan *et al.* [20] presented multiple emissary veins of the posterior fossa and unusual origin of the left PSS from a dilated mastoid emissary vein.

## ✉ Conclusions

The principal different point of our case from the previous reports is that the venous PT was associated with a single large EV (left PSS), with no other vascular, skull base, and/or middle ear abnormalities/infections present. The early initialization of an efficient medical therapy had a great impact on her favorable evolution.

## Conflict of interest

The authors state that there is no conflict of interest.

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