VIEWS AND REVIEW

Somatosound: A review with report of 9 cases with long-term follow-up

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Abstract

Somatosound (somatic tinnitus) is associated with vascular, musculoskeletal, respiratory, or temporomandibular joint disorders. Several studies of its management have been widely reported, but only few presented long-term follow-up results. The purposes of this paper are to review the causes and management, present cases with long-term follow-up, together with previously reported cases in literatures. We treated nine patients with somatosound of vascular, hematologic, endocrinologic, muscular, and cervical origin. Follow-up were conducted routinely, and the final results were collated in 3 to 11 years. Patients with non-life-threatening causes were given counseling and palliative management. Their tinnitus becomes tolerable, gradually decreased, and even disappeared. Patients with life-threatening causes were treated immediately. Among all, there were three cases which to the best of our knowledge, are the first reported of its causes. One case was caused by a compensatory of increasing blood flow in internal carotid artery (ICA) secondary to contralateral ICA stenosis. Another had a combination of anemia and an ipsilateral jugular bulb diverticulum. The tinnitus disappeared after the anemia treated. The last was patient with hyperthyroidism. The tinnitus disappeared by controlling the condition. Even when the causes are benign and the available treatments may carry risks, the patients should not be left unmanaged. Symptomatic treatment should be given, such as counseling, sound therapy, and palliative management. Our long-term observation indicated that overall outcomes are positive when the etiologies are identified early and managed properly.

Keywords: Somatic tinnitus, somatosound, pulsatile tinnitus, vascular origin tinnitus, long-term follow-up

INTRODUCTION

Somatosound is defined as a perception of a sound which appears to be preceded or strictly linked to a somatic disorder.¹⁻³ The terms “somatic tinnitus” and “somatosound” are used interchangeably. Its origin is usually vascular, muscular, skeletal, respiratory, or in the temporomandibular joint (TMJ). Somatosounds thus have an internal acoustic source^{4,5}, and the source could be detected with the appropriate sensing equipment.⁴ The reported incidence of somatosound among all kinds of tinnitus varies, for example, 17.1%⁶, 12% to 43%⁷, 5% to 10%⁸, 16% and 83%.⁹ The variations might be explained by the differences in diagnostic criteria, lack of any agreed standards

for clinical assessment⁹, study settings⁷, and its heterogeneity.¹⁰

Somatosound can be classified into vascular and non-vascular by origin⁶, and pulsatile and non-pulsatile according to the sound characteristics.¹¹

Vascular tinnitus is reported to have a 4% incidence¹² among all types of tinnitus. Among somatosounds, vascular tinnitus accounts for 7.6%, muscular origin 4.3%, and patulous eustachian tube 5.2%.⁶ In pulsatile tinnitus, venous origin is more common than arterial.¹³ Anemia-related tinnitus is infrequent and reported as 0.68% among pulsatile tinnitus.¹⁴

Among non-pulsatile tinnitus, cervicogenic somatic tinnitus (CST) arising from cervical spine disorders has a reported incidence of 43%.¹⁵

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Pulsatile tinnitus is a tinnitus which the sound is fluctuates in synchrony with the heartbeat.^{13,16} It is usually unilateral but may be bilateral in systemic disease or a high cardiac output state such as anemia or hyperthyroidism, or with the presence of a midline vascular lesion.^{16,17}

The plausible mechanisms of pulsatile tinnitus include: (1) changes in the laminar blood flow, or accelerated blood flow, resulting in audible local turbulence via the inner ear, or (2) normal blood flow sounds that are perceived more intensely due to increased bone conduction or sound conduction in the middle ear bypassing the masking effect of external sounds.^{18,19} Therefore, the causes of pulsatile tinnitus can be divided into vascular and non-vascular.²⁰ Vascular causes are arterial, venous, or arteriovenous in origin.^{19,20} The arterial causes are atherosclerosis plaques, aneurysms of the internal carotid artery (ICA) or vascular tumors, arterial dissection, fibromuscular dysplasia, arterial atherosclerosis stenosis. Venous causes are malformations of jugular bulb (JB), transverse-sigmoid sinus aneurysm, venous hum, idiopathic intracranial hypertension, high flow through local emissary veins, or local venous stenosis.^{12,13,17–19,21} Arteriovenous causes include dural arteriovenous fistula (DAVF) and malformations, capillary hyperemia, and highly vascularized skull base tumors.^{18,19} Non-vascular causes include neoplasms like a paraganglioma, osseous pathology, Paget's disease, endolymphatic sac tumor, and systemic disease,^{17,18} for example, hematologic or endocrinologic disorders.

Non-pulsatile tinnitus is caused by TMJ and cervical spine (neck) disorders,¹ eustachian tube dysfunction,^{4,5,11,22,23} or neurologic disorders such as palatotomyoclonus and idiopathic stapedial muscle spasm, which are often associated with other neurologic disorders such as brain-stem tumor, infarction, or multiple sclerosis.²⁴

Identification of the underlying cause is essential in providing appropriate treatment and determining the prognosis. The management of somatosound depends on the cause and varies from counseling, palliative, or conservative management, to aggressive intervention in life-threatening hematologic or vascular causes.

Studies of management outcomes usually involved relatively short-term follow-up periods, with the longest follow-up being up to 3 years. This review article is based on clinical characteristics and management evaluation of our nine patients with somatosound with long-term follow-up ranged from 3 to 11 years, and selective search of the literature included review articles and case

reports without restriction on date of publication. Not all types of somatosound are covered, but the common types are included. Table 1 shows the record of our nine patients' cases while Table 2 shows some example of somatosound cases we collected from literatures.

CLINICAL EXAMINATION

Evaluation of the tinnitus patient begins with a thorough history, auscultation, otoscopy, and neurological examination. It should be determined whether the tinnitus is pulsatile or non-pulsatile, and if it is unilateral or bilateral. For vascular tinnitus, palpation, provocation, and rotation maneuvers may differentiate the origin.¹⁹ In venous tinnitus, light compression of the ipsilateral jugular vein will alleviate the tinnitus, while compression of the carotid artery has no effect. The Valsalva maneuver and rotation of the head toward the ipsilateral side will decrease the tinnitus. Contralateral venous compression, the Muller maneuver, and rotation of the head to the opposite side will increase it.^{12,17,19,25} In tinnitus of arterial origin, strong compression of the carotid artery will decrease or eliminate the tinnitus. Slight compression on the ipsilateral veins, the Valsalva and Muller maneuvers, and rotation of the head toward the ipsilateral or opposite side has no effect. Slight compression on the contralateral veins also has no effect or rarely decreases the tinnitus.^{17,19}

If there are no other abnormalities, venous tinnitus is more frequently right-sided because the right jugular vein is dominant in 70%–80% of cases and often triggered by physiological conditions.¹⁹

PULSATILE TINNITUS

Vascular origin

JB abnormalities, of venous origin, are commonly encountered causes of pulsatile tinnitus. JB abnormalities include: 1) high riding jugular bulb (HRJB), when it extends above the inferior bony annulus, if it rises to the level of the basal turn of the cochlea, or if the dome of the bulb encroaches to within 2mm of the floor of the internal auditory canal; 2) jugular bulb diverticulum (JBD) which is defined as an irregular outpouching that may project into the middle ear cavity, mastoid cavity, or medially toward the petrous apex, and 3) dehiscent JB, when there is a thin or absent bony septum (sigmoid plate) between the JB and middle ear space.^{12,26–28} JB is not present at birth.²⁶ It forming after 2 years. Prevalence

Table 1: Clinical case reports

Patient	Sex/ Age	Diagnosis	History	Examination	Management	Follow-up period	Tinnitus outcome
1	F/60	HRJB with JBD	2-year of pulsatile tinnitus, right ear.	Tinnitus diminished by pressing just posterior to the right mandibular angle. TBCT: HRJB & JBD on right side.	Counseling to perform habituation.	9-year	Slowly reduced over time and disappeared.
2	F/67	Dehiscent HRJB with JBD	5-month of pulsatile tinnitus, left ear, headache, no obvious cause.	Tinnitus reduced by light compression just posterior to the left mandibular angle. No hearing loss. Neurological & otoscopy: normal. TBCT: HRJB & JBD on left side, with bony dehiscence.	Counseling and oral medications.	3-year	Slowly reduced and disappeared.
3	M/35	Venous hum	1-year of occasional pulsatile tinnitus, right ear, had disappeared but started again 1 day before visiting. Same severity.	Tinnitus reduced by compression of ipsilateral jugular vein. No hearing loss. Neurological & otoscopy: normal. Blood tests, TCD, and TBCT: normal.	-	4½-year	Disappeared completely 2 months after initial visit.
4	M/69	Increased blood flow in ICA	Long-standing hypertension & diabetes, 1-year of pulsatile tinnitus, left ear, sleep disturbances.	Auscultation: no carotid bruits, both carotid arterial pulses palpated. Carotid ultrasound: left ICA blood flow velocity (peak velocity 61.9cm/s) faster than right ICA (peak velocity 27.7cm/s). Intracranial arteries MRA: right terminal ICA stenosis.	Atenolol to decrease heart rate, antiplatelet to prevent stroke. Pulse rate decrease from 80bpm to 60bpm. Alprazolam to enhance sleeping. Advise: listen to natural sounds whenever bothered by the tinnitus.	10-year	Reduced.
5	M/44	DAVF	Falling accident leading to left occipital head injury 5 months before visiting, pulsatile headache began around the injured site and persisted. 2 months before visiting, pulsatile tinnitus started abruptly, left ear.	Tinnitus reduced by compression just anterior of left mastoid process. Auscultation: vascular bruits in front of left auricular apex. DSA: DAVF between left OA and TS.	Endovascular (partial embolization).	7-year	Persisted but tolerable.

6	F/26	Anemia & HRJB with JBD	1-month of pulsatile tinnitus, right ear.	Auscultation: no carotid bruits, both carotid arterial pulses palpated. TBCT: HRJB & JBD on right side, no bony dehiscence. TCD: increased blood flow in both MCA and BA without stenotic features. Blood tests: Hb 6.9g/dl, Ht 20.9%, WBC 25,790/ μ L, platelets 18,000/ μ L. Peripheral blood smear: mild anisopoikilocytosis with normocytic, normochromic RBC, increased WBC count with many immature cells and blast forms, decreased platelet count.	Anemia management.	5-year	Disappeared after the anemia treated.
7	F/72	Iatrogenic hyper-thyroidism	3-month of pulsatile tinnitus, both ears. 10-month of thyroid hormone replacement therapy after thyroidectomy.	Auscultation: no carotid bruits, both carotid arterial pulses palpated. TCD: increased blood flow in both MCA and BA without stenotic features. Blood tests: hyperthyroidism.	Thyroid hormone management.	10-year	Disappeared immediately after the thyroid hormone level corrected.
8	M/16	Essential palatal myoclonus	“Clicking sound” like a wristwatch, left ear. can be heard by others when listening close to the left ear, sound also present during sleep. On Levettiracetam 500mg twice/day since childhood to treat complex partial seizures.	PE: oral & oropharynx: rhythmic involuntary movement synchronous with the tinnitus. MRI, MRA, EEG: normal.	Carbamazepine, Clonazepam, Flunarizine, Divalproate, Trihexyphenidyl, Valproic Acid.	11-year	Persisted but tolerable.
9	F/65	CST	3-day of low-tone tinnitus, left ear.	1 st visit: PE: normal hearing, lump of earwax lodged in left ear canal. Pressure of left neck: dull pain around the pressed site. Turning neck to the left: mild pain on the left side. Turning neck to the right: no pain. Tinnitus become stronger when pushing against the given pressure on the left head. 2 nd visit: PE: hearing loss, earwax blocking the ear canal. Rinne test: conductive hearing loss.	1 st visit: pain medications, advice on sufficient sleep, earwax removal appointment. On the next day, tinnitus decreased, but sudden hearing loss after showering. 2 nd visit: earwax removal, hearing recovered.	5-year	Disappeared.

HRJB=high riding jugular bulb; JBD=jugular bulb diverticulum; TBCT=temporal bone computed tomography; TCD=transcranial doppler; ICA=internal carotid artery; MRA=magnetic resonance angiography; DAVF=dural arteriovenous fistula; DSA=digital subtraction angiography; OA=occipital artery; TS=transverse sinus; MCA=middle cerebral artery; BA=basilar artery; Hb=hemoglobin; Ht=hematocrit; WBC=white blood cell; RBC=red blood cell; PE=physical examination; MRI=magnetic resonance imaging; EEG=electroencephalography; CST=cervicogenic somatic tinnitus.

Table 2: Previously reported cases of Somatosound in literatures

Patient	Sex/ Age	Diagnosis	History	Examination	Management	Follow-up period	Tinnitus outcome
A ²⁹	F/58	JBD	10-year of pulsatile tinnitus, right ear.	Carotid ultrasound: normal. MRA & DSA: 2 JBD on right side.	Long-term aspirin, stenting & coiling.	10-month	Disappeared.
B ¹³	M/28	HRJB & prominent venous plexus	2-month of pulsatile tinnitus, right ear.	No hearing loss. Otoscopy: normal. Tinnitus audibles louder on auscultation at left side of posterior neck by head-flexing and turned to right side, but disappeared when the head turned to left side.. TBCT: HRJB on left side, no bony dehiscence. MRI & MRA: abnormal vascular structure in the left side of posterior upper neck & prominent paravertebral venous plexus.	Medical treatments or surgical interventions were refused.	7-month	No change.
C ²⁰	F/45	Dehiscent HRJB with JBD & sinus stenosis	2-year of pulsatile tinnitus, left ear, recently become incapacitating.	Tinnitus disappeared by compression of ipsilateral jugular vein. ENT & neurological: normal. TBCT: HRJB with JBD on left side, with bony dehiscence. MRA & DSA: hypoplasia of right TS & stenosis in a dominant left transverse SS transition.	Stenting for sinus stenosis, stent-assisted coiling for JBD because of the wide neck. JBD totally occluded.	16-month	Immediate complete disappeared.
D ³¹	F/37	Venous hum	5-year of pulsatile tinnitus, right ear, referred to psychiatrist due to insomnia and followed for 3 years but no improvement.	Tinnitus reduced by turning the head to the same side, and increase by turning to the other side. All tests: normal (unspecified).	IJV ligation.	1-year	Disappeared.
E ¹⁹	F/81	DAVF	5-month of pulsatile tinnitus, right ear, unbearable, sleep disorders, consulted several specialists but no conclusion, leading to attempted suicide.	Tinnitus detectable on auscultation behind the right ear. Tinnitus ceased by arterial compression in the right side of the neck and right mastoid process. MRA: subtle alterations as result of atypical flows in the right TS. DSA: DAVF fed by right OA. Venous drainage occurring only orthograde through the right TS/SS into right jugular vein.	Symptomatic endovascular treatment - embolization of the fistula vessels.	6-month	Disappeared.

F ⁴²	F/64	Pernicious anemia	Few months of pulsatile tinnitus, both ear.	Otосcopy: normal. Auscultation: bilateral carotid bruits, tinnitus reduced in either ear by light pressure over the carotid. Blood tests: Ht 21% with macrocytic indices, folate normal, vitamin B12 quite low. Bone marrow: increase in megaloblasts. Schilling tests: inadequate absorption of vitamin B12.	Parenteral vitamin B12, Ht: 38% in 3 weeks.	5-year	Disappeared after the anemia treated.
G ⁴⁵	F/36	Essential palatal myoclonus	Noise similar to “bubbling water” and “popping bubble wraps” for the past year, left ear, distressed from lack of sleep.	Otосcopy, audiogram, tympanogram: normal. PE: oral & oropharynx: rhythmic contraction of both side of soft palate asynchronous with her pulse. Auscultation: no carotid bruits. Brain MRI: normal.	Initial: diazepam but no relief. Botulinum toxin type A injection, 4 time over 2 years.	2-year	Asymptomatic.
H ⁴⁴	F/12	Essential palatal myoclonus	2-week of clicking sound, following an episode of upper respiratory tract infection, same quality in both ears, heard by others by listening near the head, absent in sleep, not synchronous with her pulse, can not be stop or altered by her own.	The sound audibles up to 50cm from the ear. PE: ear & nose normal, oral & oropharynx: involuntary regular movement of soft palate, absent during swallowing. CNS normal, no carotid bruits. PTA: normal hearing, high pitch tinnitus Blood tests, throat swab, EEG, MRI, cerebrum, cerebellum, brainstem, MRA: normal.	Clonazepam 0.5mg once daily for 3 weeks	1-year	Disappeared in 3 weeks, no recurrence.

JBD=jugular bulb diverticulum; MRA=magnetic resonance angiography; DSA=digital subtraction angiography; HRJB=high riding jugular bulb; TBCT=temporal bone computed tomography; MRI=magnetic resonance imaging; ENT=ear, nose, & throat; TS=transverse sinus; SS=sigmoid sinus; IIV=internal jugular vein; DAVF=dural arteriovenous fistula; OA=occipital artery; Ht=hematocrit; PE=physical examination; CNS=central nervous system; PTA=pure tone audiometry; EEG=electroencephalography.

of JB abnormalities increases during the first four decades of life and stabilizes thereafter.²⁹ The patients may be asymptomatic^{19,26,29}, or may experience tinnitus, vertigo, hearing loss, and other symptoms which vary based on the extent, position, and the affected structures.²⁶

For HRJB, initial treatment can be tinnitus retraining therapy, medication, or sound therapy.¹² Surgical treatment should be considered only when initial treatment fails. This includes jugular vein ligation, surgical lowering, and transcatheter endovascular coil embolization. Surgery has complications such as venous flow disruption, intracranial hypertension, facial nerve injury, and conductive hearing loss.^{12,27}

In JBD, the diverticula may cause pulsatile tinnitus through turbulent flow or by expansion into adjacent structures.²⁹ Endovascular treatment is a valid, safe method to treat the underlying cause^{20,29} with total amelioration of the tinnitus. For patients who do not want an intervention, counseling is an option as long the underlying causes are not life-threatening.

For a dehiscent JB, several treatment options are mentioned including jugular vein ligation, transcatheter endovascular coil embolization, and surgical covering, reinforcement, or reconstruction.³⁰ However, as mentioned, there are several potential complications. Thus, both outcomes and risks should be evaluated properly.

Among our nine patients with somatosound, patient 1 and 2 had JB abnormalities which were given only counseling to perform habituation and oral medications. The tinnitus had been successfully managed and slowly disappeared. The temporal bone computed tomography (TBCT) of both patients and illustration of digital compression examination are presented. (Figure 1 A-C) The comparative cases which we found in literatures were Patient A²⁹, B¹³, and C²⁰. Patient A had JBD which treated by endovascular intervention and remains asymptomatic. Patient B is the case example of patient with HRJB, presented together with prominent venous plexus, who refused any medical treatments or surgical interventions. On follow-up 7 months after admission, no symptomatic changes on the characters of his tinnitus have been noted. In this case, it might worthy to try different approach such as counseling, sound therapy or others, and follow it over a longer period of time, with the expectation to, at least, reduce the nuisance of the tinnitus. Patient C reported to have a dehiscent HRJB with JBD associated with transverse-sigmoid sinus stenosis. Considering that the

JBD had a wide neck, endovascular management was performed with total occlusion of JBD, and the patient experienced immediate complete regression of the tinnitus.

Venous hum, which is an idiopathic vascular tinnitus, thought to be caused by turbulent blood flow in the ipsilateral internal jugular vein (IJV). When a venous origin is suspected, a TBCT should be done to rule out JB and other venous anomalies. The diagnosis of a venous hum can only be made after the exclusion of other disorders.¹² Venous hum is usually manageable by counseling and facilitating habituation. However, ligation of the IJV may be warranted if the venous hum is loud enough to disturb sleep or hearing.^{31,32} The relief of tinnitus is immediate and usually permanent. Our patient 3 presented with venous hum which the tinnitus had disappeared in 2 months completely without any intervention. Meanwhile, venous hum in patient D³¹ had caused her a bothering tinnitus and insomnia which she had been referred to psychiatrist but no improvement. In this situation, further intervention should be taken. Ligation of the IJV was performed and the tinnitus disappeared.

Of arterial origin, atherosclerotic plaques and stenosis are the most common cause of pulsatile tinnitus in the elderly. It accounts for approximately 8%–20% of all pulsatile tinnitus cases.³³ The pathophysiological mechanism producing the pulsatile sound is turbulent blood flow, either secondary to stenosis or an increased inflow.⁸ The tinnitus may be ipsilateral, secondary to the turbulent flow, or the closure of a vessel on one side of the body may lead to a compensatory acceleration inflow in the contralateral vessel, which then becomes symptomatic and leads to contralateral tinnitus.¹⁹ Our Patient 4 had pulsatile tinnitus originating from an increased blood flow in the ICA, (Figure 2A) which was a compensatory acceleration secondary to contralateral ICA stenosis. (Figure 2B) Even though the mechanism has been previously postulated¹⁹, no case has reported. To the authors' knowledge, ours is the first case report of this kind of somatosound.

Of arteriovenous origin, DAVF, counted as a rare cause of tinnitus, causing 2%–20% of pulsatile tinnitus cases.³⁴ It is usually an acquired, abnormal connection between an artery and vein without an intervening nidus, located along the dura or within a dural sinus.¹⁷

There are 2 classification systems for DAVF. The most commonly used, the Borden-Shucart classification, categorizes the fistulae

Figure 1. (A) HRJB with JBD (Patient 1). Axial image of temporal bone CT showed a HRJB with JBD at the right side (arrow). No sign of bony dehiscence. (B) An illustration picture demonstrated light digital compression of ipsilateral jugular vein which will decrease or diminish tinnitus in venous origin. (C) A dehiscence HRJB with JBD (Patient 2). Axial image of temporal bone CT showed a HRJB with JBD and bony dehiscence at the left side (arrow). (D) Anemia and HRJB with JBD (Patient 6). Axial image of temporal bone CT showed HRJB with JBD at the right side (arrow) without bony dehiscence. (E) Patient 6. TCD test showed increased blood flows of the both middle cerebral arteries and basilar artery without stenotic features. (F) Patient 6. After remission of her leukemia, the blood flows became normalized.

into types I, II, and III, based on venous drainage characteristics.^{35,36} The Cognard classification divides DAVF into types I–V and creates subclassifications based on the shunt location, venous drainage characteristics, and venous outflow angioarchitecture.^{35,37} Detailed descriptions of the classification can be found elsewhere in the literatures.^{35,36,37}

Clinical manifestations of DAVF include pulsatile tinnitus, headaches, visual symptoms, and insomnia. There may be more serious symptoms, such as intracranial hemorrhage.⁸ Depending on the location and venous drainage pattern, DAVF arises mostly from the transverse, sigmoid, and cavernous sinuses.³⁴ Arterial inflows are mainly from dural branches of the carotid artery, most frequently the occipital artery (OA).¹⁹ Compression of the OA against the mastoid process, therefore often reduces tinnitus.¹⁹

The evaluation of flow dynamics is limited on magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), or conventional computed tomography angiography, therefore, digital subtraction angiography (DSA) is considered as the gold standard for detecting and evaluating DAVF.¹⁹ The current standard treatment consists of endovascular embolization and/or neurosurgical destruction when necessary.^{19,34}

Our patient 5 had a DAVF which only be treated partially by embolization due to its complex location. The tinnitus still persists but tolerable. The illustration of compression examination and DSA result are presented. (Figure 3 A–C) Patient E¹⁹ is a similar case to Patient 5. The tinnitus became too much affected her quality of life even led to attempted suicide. In this case, complete fistula obliteration was not indicated since the patient showed no risk of neurological complications.

Figure 2. (A) Increased blood flow in ICA (Patient 4). Carotid ultrasound revealed that peak blood flow velocity in the left ICA (61.9cm/s) was faster than the right ICA (27.7cm/s) in the cervical area. (B) Patient 4. Stenosis in the right intracranial ICA. The MRA of his intracranial arteries showed stenosis in the right intracranial ICA (arrow). (C) Iatrogenic hyperthyroidism (Patient 7). Increased blood flows of the both middle cerebral arteries and basilar artery without stenotic features were observed by TCD exam.

Instead, only symptomatic endovascular treatment was performed, which was sufficient to resolve the unbearable tinnitus.

Non-vascular origin

Hematologic and endocrinologic conditions such as anemia and hyperthyroidism can cause hemodynamic alterations. Increased cardiac output and blood flow create turbulence, resulting in pulsatile tinnitus^{12,14,38}, which is often bilateral.^{17,39-41}

Our Patient 6 is a unique case in which she had both anemia and JBD, and the pulsatile tinnitus was ipsilateral to the JBD. (Figure 1 D) Transcranial Doppler (TCD) demonstrated increased blood flow in her initial visit (Figure 1 E), which became normalized on follow-up 5 years later. (Figure 1F) Her tinnitus disappeared after recovered from the anemia following her treatment. Therefore, anemia played the key role in her pulsatile tinnitus. We can also postulate that obscured JBDs which do not present with tinnitus might be abundant. To the authors' knowledge, Patient 5 is the first case of somatosound reported with such a combination of causes and clinical course. Patient F⁴² had bilateral pulsatile tinnitus which concluded to be arises from her pernicious anemia condition.

The tinnitus disappeared right after her anemia resolved. For patient 7, hyperthyroidism appeared to be the underlying cause of her bilateral pulsatile tinnitus. Increased blood flow was demonstrated in her TCD result. (Figure 2 C) Management of her hyperthyroidism resolved the tinnitus, and she remained asymptomatic during long-term follow-up. Although hyperthyroidism is widely described in the literature as a cause of tinnitus, to the authors' knowledge, our Patient 7 is also the first case report of this kind.

Considering these cases, evaluation of patients with pulsatile tinnitus therefore requires a whole blood count and thyroid function tests to be done to eliminate hemodynamic causes.

NON-PULSATILE TINNITUS

Essential palatal myoclonus, characterized by involuntary rhythmic contraction of palatal musculature as well as pharynx⁴³⁻⁴⁵, had been reported to be one of the cause for non-pulsatile tinnitus, as appeared to our Patient 8. The structures around his pharynx, including the soft palate, uvula, and the base of the tongue, showed centrifugal rhythmic involuntary movement synchronous with the tinnitus (Figure 3D) (Supplementary data–Video 1)^a, it was also visible in his submental triangle area

(Figure 3 (E) (Supplementary data–Video 2)^b.

There are two types of palatal myoclonus, symptomatic and essential. Symptomatic palatal myoclonus is usually associated with hypertrophic degeneration of the inferior olivary nucleus, while essential palatal myoclonus has been described as either idiopathic, psychogenic, voluntary, central, or peripheral in origin, with no focal neurological lesions on imaging.^{44,45} Diagnosis is primarily made by clinical oropharynx examination. Brain MRI is needed to differentiate between the essential and symptomatic.⁴⁴ The sound can be perceived unilaterally and bilaterally depends on the cases. No definite cause of essential palatal myoclonus has been determined, therefore there is no standard treatment. Several methods have been described in the literature, including anticonvulsants and botulinum toxin injections.^{44,45}

Patient G⁴⁵ with essential palatal myoclonus was initially treated with oral diazepam but experienced no relief. After that, she was treated with botulinum toxin type A injection which then completely relieved the tinnitus. Meanwhile, patient H⁴⁴, who also diagnosed with essential palatal myoclonus, was treated with oral clonazepam 0.5mg once daily for 3 weeks and experienced total relieve of her tinnitus with no recurrence after 1 year.

As for the other cause of non-pulsatile tinnitus, a subgroup of patients can be defined where the tinnitus is related to the somatosensory system of the cervical spine. This is known as CST.^{15,46} Clinical diagnosis of CST⁴⁷ can be made when there is a history of at least one of the following before onset: (1) evident history of head or neck trauma, (2) some manipulation of the teeth, jaw, or cervical spine, (3) recurrent episodes of pain in the head, neck, or shoulder girdle, (4) temporal coincidence of the appearance or increase of pain and tinnitus, (5) an increase of tinnitus during certain postures when at rest, walking, working, or sleeping, and (6) intense bruxism during the day or night.

Other specific clinical criteria have been suggested⁴⁶: (1) neck pain, (2) impairment of cervical range of motion, in particular, rotation, (3) modulation of tinnitus by head and neck movement or posture, (4) tenderness of cervical-occipital muscles. A positive diagnosis of CST is made when all four criteria are met.

Among all criteria, the primary diagnostic is the temporal coincidence of onset or increase of neck pain and tinnitus.¹⁵ This was the case with our Patient 9. It remains unclear if the earwax had also contributed in triggering her tinnitus.

Figure 3. (A) DAVF (Patient 5). Digital compression anterior to the left mastoid process reduced the tinnitus. (B) Patient 5. Compression using knuckles together with body position gravitation against the knuckles pressure facilitated the compression to the occipital artery thus diminished the tinnitus. (C) Patient 5. Left ECA angiography showed transverse sigmoid sinus DAVF (arrow) fed by OA. (D) Essential palatal myoclonus (Patient 8). The structure around the pharynx including soft palate, uvula, and tongue showed centrifugal rhythmic involuntary movement synchronous to the clicking sound. The lips, larynx, and diaphragm were not involved. (Supplementary data -Video 1)^a (E) Patient 8. The rhythmic involuntary movement also shown on the submental triangle area (arrow). (Supplementary data -Video 2)^b

^aVideo 1 can be seen on Do Neurology YouTube channel <https://youtu.be/ZjmnFXtixgg>, also can be accessed on YouTube using keywords: “palatal myoclonus AND Do neurology.”

^bVideo 2 can be seen on Do Neurology YouTube channel <https://youtu.be/cCBKa2YBT08>, also can be accessed on YouTube using keywords: “palatal myoclonus AND Do neurology.”

However, the diagnosis for underlying cause as CST was made based on the increased neck pain and tinnitus on resisting applied pressure, consistent with that primary diagnostic criteria also with the clinical diagnosis criteria suggested by Sanchez *et al.*

Several treatments had been proposed for treating CST, including cervical physical therapies such as exercise, transcutaneous electrical nerve stimulation, manipulation of the cervical spine and trigger point deactivation by ischemic compression, steroid and lidocaine injections, mechanical treatments focused on normalizing cervical spine mobility through repetitive movements, joint mobilization, and soft tissue massage, which shown to be effectively improved the tinnitus.⁴⁸

CONCLUSION

Although this case report does not cover all types of somatosound, the common types are included. Our impression based upon long-term observation is that overall outcomes are good if the etiology is determined and treated early.

Life-threatening underlying causes should be treated urgently. However, benign or non-life-threatening causes might be left untreated, but definitely should not be neglected. Even though the causes are benign, and one might conclude that there are no risks necessarily taken to treat them, these patients should not be left unmanaged. Such patients can be offered symptomatic management. Counseling is very important to reduce the nuisance of tinnitus. Facilitating habituation with sound therapy also play a significant role. Palliative management for sleeping problems, anxiety, or decrease heart rate are options to reduce the nuisance of vascular tinnitus. Such management should be offered to relieve the patient's distress.

The management of somatosound varies depending on the causes, the options available to the therapists, and the patient's preferences. The long-term follow-up results of our patients showed that somatosound could disappear or weaken with time, often achieved through habituation imposed by the patients themselves. The authors hope that this paper will give therapists an overall understanding of somatosound management.

DISCLOSURE

Conflict of interest: None

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