Non Otologic Causes of Tinnitus – A Review

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Abstract

Tinnitus or ringing in the ear can be caused by variety of causes, the otologist must be aware of causes of tinnitus outside the ear. This article focuses on such causes of tinnitus, their examination, diagnosis and possible mechanisms of their generation.

Keywords: Costen's Syndrome, Dehiscent jugular bulb, Palatal Myoclonus, Vibratory Tinnitus

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Introduction

Tinnitus may be defined variously as 'a sound perceived for more than five minutes at a time, in absence of any external acoustical or electrical stimulation of the ear and not occurring immediately after exposure to loud noise,' phantom auditory perception' or 'head noise'. In the light of current understanding, tinnitus may be defined as an auditory perception due to aberrant spontaneous activity, arising from an altered state of excitation or inhibition within the auditory system[1]. Thorough history of Tinnitus and its characteristics can lead us to a diagnosis. Virtually any lesion that causes a conductive or sensorineural hearing loss, alters the normal local or regional blood flow, or changes local muscular activity may cause tinnitus.

Etiology of Tinnitus

Sr. No	Affected sites	Name of diseases
	External Auditory Canal	
1	Foreign Body Cerumen	Impaction
2	Infection	Otitis externa (acute, chronic, fungal, necrotizing)
3	Tumor	Benign (osteoma, exostosis) Malignant (squamous cell carcinoma, basal cell carcinoma, ceruminoma)
4	Atresia	Congenital (osseous, nonosseous) Traumatic
5	Tvmpanic Membrane	Perforation Atelectasis
	Middle Ear	

6	Effusion	Blood Cerebrospinal fluid(purulent, serous, mucoid)
7	Ossicular	Fixation Discontinuity
8	Tumour	Cholesteatoma Glomus tumor Facial nerve neuroma Hemangioma Carcinoma
9	Vascular	Aberrant vessels (internal carotid artery,dehiscent jugular bulb) Persistent vessels (stapedial artery)
10	Neuromuscular	Myoclonus (palatal, tensor tympani, and stapedial muscles)
Cochlea		
11	Virtually any disorder of the cochlea may result in a sensorineural hearing loss.	

Retrocochlear		
1	Internal Auditory Canal and Cerebello-pontine Angle	Acoustic neuroma Cholesteatoma Hemangioma, Facial nerve neuroma Meningioma Vascular loops on auditory nerve
2	Central Nervous System	Tumors, inflammatory and vascular lesions
3	Miscellaneous	Patulous Eustachian Tube Temporomandibular Joint Dysfunction Head Trauma Extracranial Aneurysm Arteriovenous Malformations Venous Hum

Physical Examination

A thorough otoscopic examination of the ears should also include pneumatic otoscopy and tuning fork tests. Examination of the nose, nasopharynx, oral cavity, larynx, and neck should also be included. A neurologic evaluation with special attention to cranial nerves and cerebellar function may reveal findings compatible with intracranial pathology. If

the history suggests a pulsatile tinnitus, auscultation of the systemic blood pressure and the many vessels of the head and neck may uncover the source of the head noise. The neck should be ausculated for bruits associated with carotid body tumors, arteriosclerotic vascular lesions, and the crescendo-decrescendo murmur of a "venous hum." The intracranial area can be ausculated by placing a stethoscope over the eyes, the temporal-parietal region, and the mastoid process. A bruit or murmur may suggest an arteriovenous malformation (AVM) or aneurysm. Extracranial arteriovenous malformations and aneurysms may also occur, and can usually be detected by careful examination of the skin in the region of the ear and overlying the skull.

Laboratory Examination

Audiometry for pure tone air and bone conduction. Impedance audiometry with acoustic reflex decay, BERA, OAE should also be part of the standard battery of tests for patients complaining of tinnitus. Tinnitus matching should be performed to obtain for the tinnitus profile. Additional tests are obtained as indicated by the results of the history and physical examination.

Non-Otologic causes of Tinnitus (Vibratory)

This group includes pulsating clicking sounds having a mechanical basis and arising in or near the ear these head noises have been further divided into those that can be heard by interested listeners (objective) and those heard only by the patient (subjective). Although some sounds emanating from the head are audible without amplification, others are heard only with the aid of a stethoscope or *Toynbee* ausculation tube.

Vascular

Local or regional vascular disorders are a common cause of vibratory tinnitus. Characteristically the head noise is pulsatile and synchronous with the hearbeat. Unless the cause is obvious, every patient with pulsatile tinnitus deserves an intravenous contract enhanced CT scan of the temporal bones and intracranial structures. This one test should rule of most of the following causes of pulsatile tinnitus or demonstrate an abnormality which would lead to further diagnostic evaluation.

Arteriovenous Malformations

An arteriovenous malformation (AVM) is a tangled, interconnected network of vessels in which arterial blood passes directly into venous channels without the normal intervening capillaries. Intracranial AVM's are most common in the posterior fossa and occur mainly between terminal branches of the external carotid artery and the transverse sinus. Up to 50 percent of

these patients will have nonlocalizing neurologic deficits.

Patients frequently present with a subjective or objective continuous head murmur with systolic intensification (louder with the heartbeat). Ausculation over the carotid bifurcation may reveal a bruit; however, in posterior fossa AVM's the sound is loudest over the mastoid process or occipital artery and in anterior and middle cranial fossa AVM's the pulsations are maximal over the eye or temporal region *Arenberg and McCreary* (1972)[2]. The tinnitus can often be eliminated or decreased by carotid artery compression. Extracranial AVM's causing tinnitus occur-over the scalp or in the region of the ear and are usually obvious on -physical examination. These dilated, 'easily - compressible, bluish masses underlying the skin will often have a palpable vibration with systolic intensification [3]. CT or MR angiography may be diagnostic.

Carotid - Cavernous Fistula

This condition is an abnormal communication between the internal carotid artery and the cavernous sinus. Post-traumatic fistulae are three times as common as those-occurring spontaneously. Clinical findings in this disorder include exophthalmos, ocular pulsations; orbital and/or cephalic bruits, headache, chemosis, extraocular palsies and loss of vision. The bruit increases in intensity when the patient is reclining and the environment is quiet, often making sleep difficult. It may be audible over the orbit, temporal region, frontal area, or great vessels of the neck and is occasionally heard without a stethoscope. A bruit or pulsatile tinnitus is the most common initial symptom of this disease and is ultimately present in almost 80 percent of cases.

Angiography is the definitive diagnostic procedure and must be performed to plan the operative approach. Life-threatening intracranial hemorrhage occurs in about 3 percent of untreated patients, although spontaneous remission may occur in up to 10 percent of patients. Operative treatments include carotid artery ligation and various other techniques *Day and Rhoton* (1982)[4].

Aneurysm

Middle ear aneurysms of the internal carotid artery are rare. The aneurysm is usually anterior or anterior inferior in the middle ear space, and on physical examination presents as a pale pink to reddish mass behind an intact tympanic membrane. An aberrant or tortuous internal carotid artery that is buckled into the middle ear cavity will have the same appearance. If the artery is in contact with the tympanic membrane or ossicles, or sufficient turbulent blood flow occurs, a

pulse synchronous sound may be heard with the Toynbee tube.

Though these are extremely rare diagnosis of an aneurysm is by angiography. Treatment of extracranial aneurysms involve ligation of the artery. Unless an aneurysm or aberrancy of the internal carotid artery in the middle ear is symptomatic (bleeding, enlarging, or destroying hearing by impinging on the ossicles), no treatment is necessary. The potential mortality and morbidity of primarily clamping an internal carotid artery should temper thoughts of such a heroic procedure performed solely for the symptom of tinnitus. Intracranial aneurysms are surgically treated because of ever present risk of intracranial hemorrhage and consequent stroke or death.

Venous Hum

Venous hum is also known as a cephalic murmur or essential objective tinnitus and is caused by eddy currents in the internal jugular vein, jugular bulb, or sigmoid sinus *Iglaver* (1931)[5], *Hentzer* (1968)[6]. Although some cases have been reported following trauma of the head and neck, the majority of cases are idiopathic. This condition also occurs in high cardiac output states such as severe anaemia, thyrotoxicosis and pregnancy *Iglaver*, (1931)[5], *Ward* et al., (1975)[7]. It is 2 to 4 times more common in females and is most common in the third and fourth decades of life.

The patients usually presents with subjective and objective tinnitus that is synchronous with the pulse and has systolic intensification. The tinnitus is usually maximal with the head facing forward and decreases in intensity or is eliminated by turning the head toward the side of the tinnitus or exerting light pressure upon the lower anterior neck. Although turning the head to the opposite side either has no effect or exaggerates the tinnitus, turning to the extreme opposite side will decrease the intensity of the pulsations. Performing the Valsalva maneuver will decrease and deep breathing will increase the tinnitus of venous hum. It should be confirmed by angiography and venography to rule out the presence of an AVM, aneurysm or vascular tumor. Hematologic studies may be indicated if anemia or thyrotoxicosis are suspected. If associated with pregnancy, the tinnitus will resolve when the uterus empties. Treatment is usually not required, however, if the tinnitus persists and is a major problem for the patient, ligation of the jugular vein usually effects a cure.

Dehiscent Jugular Bulb

The normal jugular bulb is in the floor of the hypotympanum, separated from the middle ear space by an average of 0.5 to 5 mm of bone. Congenital or traumatic

absence of this bony separation may allow the jugular bulb to enlarge and subsequnt1y enter the middle ear from inferiorly. Extension to the tympanic membrane or ossicles produces a pulsatile tinnitus with the same characteristics as venous hum. Bluish mass can be identified inferiorly behind an intact tympanic membrane. Audiometry may be normal or reveal a conductive hearing loss. Impendence studies are variable and may demonstrate findings compatible with normal middle ear function. The diagnosis is confirmed by radiologic evaluation (CT scan or jugular venography). If the tinnitus or hearing loss is both of the jugular vein and sigmoid sinus may be considered. Usually, however, no treatment, no treatment is required.

Persistent Stapedial Artery

In the human embryo, an artery courses from the second aortic arch (stapedohyoid artery) through the stapes crura (Obturator foramen). Persistence of this artery into adult life is rare and usually asymptomatic,however, pulsatile tinnitus may be produced due to the movement of the stapes suprastructure with each arterial pulsation.

Although tympanometry may show tympanic membrane movement synchronous with the pulse, the diagnosis of this condition is difficult. Depending upon the size of the artery, angiography may be of benefit. Unless extremely symptomatic, diagnosis by exploratory middle ear surgery is unwarranted, since attempts at removal of the artery are not without risk to the hearing.

Eagle's Syndrome

This disorder is caused by an elongated styloid process or calcified stylohyoid ligament. It is usually characterized by a dull or nagging pain in the throat which may radiate up to the ear, producing otolgia. The pain is elicited by palpation of the tonsillar fossa and not consistently relieved by topical or local anesthetic *Christiansen, Meyerhoff and Quick* (1975)[8]. Radiographic demonstration of either an elongated styloid process or a calcified stylohyoid ligament is essential for diagnosis. Bruits or objective tinnitus due to this anatomic abnormality are rare but may be relieved by resection of the styloid process *Cordier, Garnier and Durupt* (1970)[9].

Glomus Tumor

Glomus tumors arise from small glomus bodies located along Jacobson's nerve on the promontory of the middle ear and in the adventitia of the anterior part of the dome of the jugular bulb. These tumors spread by direct extension, destroying bone and invading soft tissue. They are multicentric in origin in 5 to 8 percent of cases and although histologically benign, there have been rare reports of metastases.

Glenner and Grimley, (1974)[10], *Glasscock, Harris and Newsome,* (1974)[11].

Glomus tumors cause pulsatile tinnitus which is usually subjective and occasionally objective. Continued growth of the tumor causes impingement on the tympanic membrane or ossicles resulting in a conductive hearing loss. Growth through the tympanic membrane results in infection and recurrent hemorrhage from the ear. The tumor may invade the facial nerve, cochlea (via the round window), internal jugular vein, carotid artery canal, cranial nerves IX, X, Xl, XII, and extend intracranially. Death may occur from intracranial extension.

On physical examination, a reddish mass is usually seen behind an intact tympanic membrane. Because of the potential morbidity of this tumor, every case of pulsatile tinnitus must be considered a glomus tumor until proven otherwise. CT scan, angiography, and venography are required to determine the location and extent of a particular tumor.

Hypertension

High blood pressure may produce a pulse synchronous murmur in the head. Every patient with pulsatile tinnitus should have a recording of the pressure. Hypertension is usually readily controlled with oral medications.

Palatal Myoclonus

Myoclonus is an involuntary shock-like contraction of a muscle in which the jerks may be single or repetitive for varying periods of time. The palatal movements are upward and downward at a rate of 40-200/minute and are often associated with contraction of the tonsillar pillars. The average rate of contraction is approximately 125/minute, but this may vary considerably from day to day. There are frequently other associated movements of the tongue, larynx, pharynx, facial muscles, diaphragm and temporalis, masseter and sternocleidomastoid muscles. The movements often cease when the mouth is open or the, patient is speaking or swallowing but do not disappear with sleep or coma, *Swanson, Luttrell, and Magladery* (1962)[12]. The sound is the same as the noise caused by the clicking together of two fingernails and is be due to the walls of the eustachian tube snapping shut *Pulec and Simonton*, (1961)[13].

Most reported cases of palatal myoclonus have had abnormalities of the pons or cerebellum. Although the principal causative factor seems to be acute brainstem infarction, this disorder may be due to multiple sclerosis, trauma, tumor, brainstem angioma, progressive bulbar palsy or encephalitis *Swanson*, et al., (1962)[12].

Diagnosis is frequently made from the history with visual inspection of the palatal excursions confirming the diagnosis. Electromyography of the soft palate will provide objective proof of the myoclonus. Treatment is often unsuccessful. Various surgical procedures have been utilized to denervate the tensor veli palatine muscles of the soft palate or alter the ultimate direction of pull the palatal muscles. Carbamazepine has been used with limited success in several patients *Rahko and Hakkinen,* (1979)[14].

Stapedial muscle spasm

Tinnitus from stapedial muscle spasm is initiated by loud sounds and is exacerbated by prolonged sound exposure. Thresholds of 70-90 dBHL initiate the tetany even though the stapedial contractions may intermittently recur after the sound is turned off *Marchiando, Per-Lee and Jackson,* (1983)[15].

Tinnitus of this type has also been described in post facial paralysis patients during eyelid closing. At exploratory middle ear surgery, eyelid closure correlated with stapedial muscle contraction and movement of the stapes. Incidence studies on these same post facial paralysis patients have documented compliance changes on eye blink, suggesting misrouting of regenerating orbicularis oculi facial nerve fibers to the stapedial muscle (*Williams*, 1980[16]; *Watanbe, Kamagami and Tsuda*, 1974[17]). Diagnosis is confirmed with impedance audiometry. Treatment is with selective lysis of the stapedial muscle tendon. If the patient does not desire surgery, a muscle relaxant may control the symptoms for variable periods.

Tensor Tympani Muscle Spasm

Although stapedial muscle spasm has been more widely documented, it is suggested that spasm of the tensor tympani muscle may produce similar symptoms. Impedance audiometry reveals medial movement of the tympanic membrane with tensor tympani muscle spasm and lateral or outward movement with stapedial muscle spasm. Treatment is lysis of the tensor tympani muscle tendon in the middle ear.

Temporomandibular Joint Dysfunction

This disorder (also known as Costen's Syndrome) is due to disease and dysfunction of the temporomandibular joint (TMJ) and is occur in the more than 20 percent of the adult population (Morgan, 1973). The resulting syndrome has a variety of seemingly unrelated areas of pain and muscle spasm and is characterized by snapping, popping, or clicking sounds and deviation of the mandible when the mouth is opened, crepitus (grating noise) over the joint, limitation of motion, low buzzing tinnitus, disequilibrium, hearing loss, otalgia, headache, and tenderness over one or more

of the muscles of mastication *Costen* (1934)[18], *Sharav, Tzukert and Refaeli (*1978)[19], *Myrhang* (1964)[20]. Acoustic symptoms, consisting of tinnitus, popping sensations in the ears and hearing loss, are present in over 25 percent of patients suffering from TMJ dysfunction *Myrhang* (1964)[20].

Electromyographic recordings in the and temporalis muscles are often abnormal, and occasionally the tympanic membrane can be seen to retract medially due to the pull of the tensor tympani muscle during clenching movements of the jaw. Treatment consists of the use of dental acrylic bite blocks to correct malocclusion, anti-inflammatory agents and local heat to relieve muscle spasm, surgery to correct the offending TMJ abnormality and psychologic counseling to try to correct the underlying psychological problem that resulted in the masticatory muscle spasm in the first place.

Patulous Eustachian Tube

The normal eustachian tube remains closed except during yawning and swallowing, at which time it opens to allow airflow from the nasopharynx to an area of relative negative pressure in the middle ear space. The problem frequently seems to arise with loss of peritubal fat around the medial cartilaginous region of the tube and not infrequently atrophic changes are present in the mucosa of the nose, nasopharynx and oropharynx. Many cases have occurred following marked weight loss.

The characteristic symptom of a soft, blowing murmur synchronous with respiration is often audible with the Toynbee auscultation tube. Examination of the tympanic membrane reveals inward and outward movements of the drumhead with each respiration. The patient may complain of a stuffy full feeling in the ears, and of autophony, in which his own voice seems to ring or echo in his head. Impedance audiomertry can be diagnostic.

Conclusion

So in cases of intractable tinnitus we need to be very attentive to the history of tinnitus given by patient and also look for the causes outside ear.

References

1. Scott Brown's Otorhinolaryngology, Head and Neck Surgery.7th ed 2008.
2. Arenberg, I: K. and El. S. McCreary: Objective Tinnitus Aurium and Dural Arteriovenous Malformations of the Posterior Fossa. Ann. Otol. 80: 111-120, 1972.
3. Hamburger, L. P.: Head Murmurs. Am. J. Med Sci. 181 : 756-762, 1931.
4. Day, A. L. and A. L. Rhoton: Aneurysms and Arteriovenous Fistulae of the Intracavernous`

Carotid Artery and Its Branches. In: Neurological Surgery, Vol. III (Ed. J. R. Voumans), W. B. Saunders, Philadelphia, pp. 1764-4783, 1982.

5. Iglauer, S.: Objective Tinnitus Aurium. Arch. Otolaryngol. 18 : 145-151, 1931.
6. Hentzer, E.: Objective Tinnitus of the Vascular Type. Acta Oto-Laryng. 66 : 273-281, 1968.
7. Ward, P. H. K. Babin, T. C. Calcaterra, and H. R. Konrad: Operative Treatment of Surgical Lesions with Objective Tinnitus. Ann. Otol, 84: 473-482, 1975.
8. Christiansen, T. A., W. L. Meyerhoff, and C. A. Quick: Styloid Process Neuralgia. Arch. Otolaryngol. 101: 120-122, 1975.
9. Cordier, G., H. Gamier, and L. Durupt: Les allongements anormaux de l'apophyse styloide temprale avec retentissement cartidien. La Presse Medicinale No. 30, p. 1547, June 21, 1970.
10. Glenner, G. G. and P. M. Grimley: Tumors of the Extraadrenal Paraganglion System (Including Chemoreceptors). In: Alias of Tumor Pathology, Second Series, Fascide 9. Armed Forces Institute of Pathology, Washington, D. C., 1974.
11. Glasscock, M. E., P. E Harris, and G. Newsome: Glomus Tumors: Diagnosis and Treatment. Laryngoscope 84: 2006-2032, 1974.
12. Swanson, P. U., C. N. Luttrell, and J. W. Maglader: Myoclonus - A Report of 61 Cases and Review of the Literature. Medicine 41: 339-356, 1962.
13. Pulec, J. L. and K. M. Simonton: Palatal Myoclonus: A Report of Two Cases. Laryngoscope 71: 668-671, 1961.
14. Rahko, T. and V. Hakkinen: Carbamazepine in the Treatment of Objective Myoclonus Tinnitus. Jnl. Laryngol. Otol. 93 : 123-127, 1979.
15. Marchiando, A., J. H. Per-Lee, and R. T. Jackson: Tinnitus Due to Idiopathic Stapedial Muscle Spasm. ENT Jnl. 62 : 8-13, 1983.
16. Williams, J.D.: Unusual but Treatable Cause of Fluctuating Tinnitus. Ann. Otol. 89: 239-240, 1980.
17. Watanbe, W., H. Kamagami, and T. Tsuda: Tinnitus Due to Abnormal Contraction of Stapedial Muscle. Otol. Rhinol, Laryngol. 36: 217-226, 1974.
18. Costen, J. B.: A syndrome of Ear and Sinus Symptoms Dependent

Upon Disturbed Function of the Temporomnandibular Joint. Ann Otol. 43: 1-15, 1934.

19. Sharav, Y., A Tzukert, and B. Refacli: Muscle Pain Index in Relation to Pain, Dysfunction, and Dizziness Associated with the Myofascial Pain-Dysfunction-Syndrome. Oral Surg. 46: 742-747, 1978.
20. Myrhang, H.: The Incidence of Ear Symptoms in cases of Malocclusion and Temporomandibular joint Disturbances. Brit. Jnl. Oral Surg. 2: 28-32, 1964.