Hearing Impairment in Vascular Disorders

Nadir Yıldırım

Abstract

Objective: Cochlea is provided with the arterial blood by the labyrinthine artery that is an end-branch of the vertebrobasilar system. Therefore, it is very susceptible to any compromise of the systemic and local circulation. During the last decades, a good proportion of the audio-vestibular disorders, that is previously labelled as “idiopathic” are proved to be of vascular origin through the advances in the study of microcirculation of the inner ear. This study aims to review the relevant literature on the relationship of inner ear pathologies, hearing loss in particular, and local and systemic vascular diseases in order to put together the current knowledge on the subject in a comprehensive manner.

Material and Methods: A literature search have been conducted through “Pubmed” and full texts of relevant literature have been obtained.

Results: A total of 82 landmark publications have been selected and used for the study.

Conclusion: In this literature review, characteristics of cochlear blood flow and its disturbances are summarized with their clinical correspondences as to include recent findings.

Key words: inner ear, cochlea, hearing, vascular

Overview of the Cochlear Blood Supply

Arterial blood supply to the cochlea is provided by a sole vessel of labyrinthine artery. In post-mortem studies, this artery has been found mainly arising from the anterior inferior cerebellar artery (AICA), a branch of the basilar artery in 83 % of the specimens, occasionally from the basilar artery itself (14%), or rarely its posterior inferior cerebellar artery (PICA) branch (1,2). Basilar artery is formed by two merging vertebral arteries (Fig.1).

Cochlear artery, a branch of the labyrinthine artery, enters internal acoustic canal following the cochlear branch of the VIIIth nerve, and gives off two branches within the cochlea: spiral modiolar artery and vestibulo-cochlear artery. Cochlear veins also follow the same route with the same names as arteries, e.g. spiral modiolar vein and drains into the vein of the cochlear aqueduct.

At the level of spiral lamina, arterioles divide into fine collaterals in radial fashion forming three distinctive group of capillary network, forming rich anastomoses with each other (Fig. 2) (3). In addition to this brief anatomical description, below given characteristics of the cochlear blood flow (CBF) are also found very important and highly relevant to central auditory or cochlear disturbances of vascular origin:

1. Cochlea is considered as an end-organ in terms of vascularization, as the blood is supplied mainly by a single artery, as it is the case for retina, heart and kidneys. This type of vascularisation makes the organ particularly vulnerable to any circulatory constriction (4). Insufficient cochlear blood supply may also increase susceptibility to noise induced hearing loss and accelerates natural going process of the cochlea (5).

2. There are well developed circulatory compensatory mechanisms for vital organs, which are activated when the oxygen supply is constrained. It has been shown that 30% decrease in hematocrite achieved 75% increase in cerebral blood flow, including of the brainstem, where auditory pathways and nuclei are located. Similar degree of compensation is estimated for the inner ear as well (6).

3. Cochlear blood flow is under the control of autonomic regulation that occurs along the basilar...
artery, the anterior inferior cerebellar artery and at the points of cochlear artery where the spiral modiolar artery branches off (7). Autoregulation of CBF has been demonstrated in guinea pigs (8). In this experiment, it was found that when this autoregulatory system is pharmacologically blocked, CBF responds proportionally to the changes in the systemic blood pressure. It has been shown through animal experiments that cochlear microvessels are rich in myofibrils that respond to this autonomic influence. This feature also offers therapeutic possibilities as to improve CBF by extrinsic agents, on the other hand,

4. There is no direct blood supply to the neuroepithelium of the organ of Corti. It receives oxygen and nutrients either from the spiral prominence vessels underlying basilar membrane or spiral limbus vessels, both branches of the spiral prominence vessels and adjacent to the organ (Fig. 2) (3).

5. Temporal bone histopathologic studies showed that small vessels are coursing freely through the perilymphatic space of the cochlea, especially in the apical turn (10).

6. Following parameters have been identified by the researchers studying (CBF) as the indicatives of significant circulatory disturbances (11,12,13):
   a. External artery constriction in the lateral wall,
   b. Irregularities within the vessel lumen,
   c. Red blood cell (RBC) aggregation,
   d. Decrease in RBC velocity,
   e. Changes in RBC density.
Investigation Methods of CBF

Vertebro-basilar system is well visualized through conventional arteriography up to the labyrinthine artery. The pathologies in the vessels of vertebrobasilar system as well as resultant ischemia, however, better studied through digital subtraction angiography (DSA), single photon emission computerized tomography (SPECT), magnetic resonance angiography (MRA) and computerized tomographic angiography (CTA) (14, 15). Microcirculation of the labyrinth, on the other hand, is assessed on research bases using videomicroscopy (7) and laser Doppler flowmetry (16, 17). Changes in the CBF have also been found to well correlate with the changes in the outer hear cells (OHC) electromicroscopically (18) and increase in the latency of the distortion product oto-acoustic emissions (DPOAE), both of which indirectly reflect the status of the microcirculation (19).

Local Vascular Diseases Effecting

Local primary vascular or other space occupying pathologies may cause compression or stenosis of the main supplying vessels of the labyrinth, such as internal carotid artery, vertebral artery, basilar artery or jugular vein, thereby indirectly effecting labyrinthine circulation without any intrinsic general or systemic vascular pathology of the inner ear. Sometimes, they cause ischemia and infarction of the end organs of cochlea and vestibule.

Experimental occlusion of the cochlear blood vessels themselves in animal model has been demonstrated to reduce the cochlear blood flow by 35%, resulting in drastically reduced cochlear oxygenation and auditory dysfunction that is well documented by means of electrocochleography and auditory brainstem response (ABR) measurements (20).

Reduced blood flow to the inner ear infrequently manifests with sudden hearing loss, either by directly affecting hair cells and/or stria vascularis or the VIIIth nerve. In a series of 392 patients with sudden hearing loss, 5% of the cases are attributed to pure vascular causes (21). Conductive deafness due to pathologies of the neighboring vessels has also been reported.
A. Pathologies of the adjacent vessels:

1. Congenital or idiopathic vascular disorders:
   
a. *Aberrant carotid artery* within the middle ear as a result of dehiscent hypotympanum may cause variable conductive or mixed hearing loss, however it is a very rare condition (22, 23, 24).
   
b. *Arterial loops of AICA* compress the VIIIth nerve on occasions, causing typical cerebello-pontine angle (CPA) mass findings with the signs and symptoms of sensorineural hearing loss (SNHL) of retro-cochlear origin (25, 26). Pulsatile tinnitus is also reported to be one of the hallmarks of this anatomic variation (27).
   
c. *Ectatic vertebral and basilar artery* has been shown to mimic CPA tumors, Ménière’s disease and other peripheral or central conditions with inner ear symptoms, by compressing brainstem and the VIIIth nerve (26, 28).
   
d. *Aneurysm of AICA* may also cause sudden hearing loss, although it is extremely rare, mostly originates from the loops around the internal acoustic meatus (IAM). Less than 50 cases have been reported so far. Signs and symptoms are indistinguishable from the CPA tumors (29, 30).
   
e. *High jugular fossa* is usually seen on the right side, encroaches upon labyrinth, effects cochlear aqueduct, vestibular aqueduct and erodes posterior semicircular canal. It typically causes low frequency SNHL as well as occasional conductive hearing loss (31, 32).
   
f. *Greatly enlarged jugular fossa* is also situated higher than normal level and accompanied by a giant sigmoid sinus and sometimes with a diverticulum. Symptomatology and aetio-pathogenesis is very similar to those of high jugular fossa. In addition, giant jugular fossa may affect cochlear circulation by causing turbulent flow and decreased venous return (31).
   
g. *Cavernous malformation of the internal acoustic canal* (33).
   
h. *Dissection of the vertebral artery* (34, 35, 36).
   
i. *Anomalous carotid artery* (37).
   
j. *Arterial anomalies of the middle ear* associated with stapes ankylosis (38).
   
k. *Aneurysm arising from the petrous portion of the internal carotid artery* (39).

I. *Infarction of internal auditory artery*: Isolated infraction of internal auditory artery has also been reported with the histopathologic correlates of degeneration in the vestibulo-cochlear nerve (40).

m. *Susac’s syndrome* is an idiopathic disorder characterized by the triad of encephalopathy, fluctuating hearing loss, and visual loss resulting from microangiopathy of the brain, cochlea, and retina (41).

2. Vascular tumors:
   
   
b. *Cavernous hemangioma of the IAC* (43).
   
c. *Glomus jugulare tumors* (paragangliomas) cause conductive hearing loss at early stages and may invade labyrinth, causing SNHL (44).
   
d. *Non-paraganglioma jugular foramen lesions* (45).
   
e. *Angiosarcoma of the temporal bone* (46).
   
g. *Jugular foramen schwannoma* (47).

B. General restrictions of vertebro-basilar system:

1. *Thrombosis of AICA*:

   Thrombosis of AICA affects almost all the structures in the brainstem, including auditory pathways and nuclei at varying degrees as well as the VIIIth nerve and labyrinth itself. Isolated vestibulo-cochlear damage, due to involvement of the cochlear and vestibular nuclei as a result of brainstem infarction also has been reported (48). However, it is more likely that vestibule-cochlear involvement is accompanied by ponto-cerebellar findings such as dysarthria, dysmetria and decreased facial sensation (49). Some cases of AICA infarction may also present with recurrent symptoms that mimic Ménière’s disease (50). There is a thrombus formation within the cochlear vessels as well, accompanied by widespread serous fluid collection and inflammatory cell infiltration both within the labyrinth and the myelin sheath of the VIIIth nerve. Sudden hearing loss that usually accompanies the pathology is due to cochlear involvement; whereas late auditory dysfunction tends to be caused by the residual brainstem lesion if the patient survives (51).

2. *Vertebro-basilar ischemia* is usually caused by atherosclerosis and may lead to sudden hearing loss by labyrinthine infarction with accompanying vertigo that could be bilateral (52, 53, 54). Although very rarely, infarction in the territory of PICA may also be associated with audio-vestibular symptoms (55).

**Systemic Cardio-Vascular Diseases**

There is a well established correlation between SNHL and systemic cardiovascular diseases. Patients with hearing loss of unknown etiology have been found 8 times more prone to have concomitant ischemic heart disease than their healthy peers. Susmano and Rosenbush
speculated that, this strong correlation may imply a possible genetic defect that is expressed in both coronary and labyrinthine arteries, which are both end-arteries (56). This theory may explain the co-existence of some congenital cardiac diseases with SNHL, such as Jarvell-Lange-Nielsen’s Syndrome, Romano-Ward’s Syndrome and valvular-pulmoner stenosis. Beyond this hypothetical co-existence, there is a plethora of studies reporting impaired cochlear blood flow and SNHL as a result of cardiovascular diseases. Bachor et al in their post-mortem studies of temporal bones found a statistically significant correlation between the congenital heart defects and abnormalities in the audio-vestibular vessels, which are more pronounced in the cochlea than the vestibule (10).

A. Intrinsic diseases of the cardio-vascular system:

Vertebral giant cell arteritis effects whole vertebro-basilar system including labyrinthine artery, presenting with Méniere-like symptoms (57).

Diabetes mellitus may cause low frequency hearing loss. However, it is argued that, the hearing loss seen in insulin-dependant diabetes mellitus patients is more related to peripheric neuropathy rather than microangiopathy as once had been thought (58). Recently, in diabetic rat inner ears, Liu et al have shown upregulation of nitric oxide (NO) that functions in the vascular tone, and increase in the expression of vascular endothelial growth factor (VEGF), that induces angiogenesis and plays a crucial role in diabetic microangiopathies (59).

Common systemic cardio-vascular diseases: Coronary heart disease, intermittent claudicatio and systemic hypertension are all associated with SNHL especially in elderly patients. They more likely accelerate the ongoing cochlear ageing process rather than causing sudden deafness, as well as causing retrocochlear hearing loss and central auditory dysfunction by way of predisposing cerebro-vascular accidents (58). Patients with hypertension showed deterioration of hearing thresholds at 8 kHz and, compared with normotensive subjects, a higher frequency of abnormal otoacoustic emissions (60). Likewise, stress, hyperlipidemia, glucose intolerance, renal apparatus and even gastroenteric diseases with a functional component can attribute to the development of hearing loss by constituting risk factors for systemic cardio-vascular diseases and/or hemodynamic imbalance (61, 62). Nevertheless, there is no obvious cause-effect relationship established between hearing loss and cardio-vascular insufficiencies. That is possibly due to varying degree of compensatory involvement of the above mentioned local auto-regulatory mechanisms.

4. Systemic vasculitis: Any systemic vascular disease that involves small arteries and arterioles can easily affect cochlear vessels. They include a broad spectrum of diseases which are characterized by disseminated blood vessel inflammation in different organs and systems, mostly by various immunopathological mechanisms. Systemic vasculitis usually causes sudden or progressive SNHL or mixed hearing loss by predisposing different forms of otitis media as well (63). Once they affect the inner ear, the damage is usually grave and irreversible. Long-standing systemic arteriolar insufficiencies tend to cause low frequency hearing loss, presumably due to resultant strial atrophy, whereas aforementioned cardio-vascular diseases typically cause or contribute to high frequency hearing loss by hair cell loss.
   a. Systemic lupus erythematosis (SLE): Typically causes severe vasculitis in the apical turn of the cochlea. Outer hair cells are lost by ischemic necrosis and microinfarctions. Clinical manifestation is typical of rapidly progressing SNHL (63, 65).
   b. Wegener’s granulomatosis: Commonly causes otitis media with effusion (35-47%), sometimes with chronic granulomatous changes in the middle ear (64). It may also cause cochlear damage and VIII th nerve damage by directly affecting their arterioles through necrotizing vasculitis (66).
   c. Polyarteritis nodosa (PAN) tend to cause and more severe inner ear changes, that sometimes leads to infarctions and fibrotic changes and less serious form of OME than Wegener’s granulomatosis (65).
   d. Cogan’s syndrome: Active vasculitis and fibrosis of the medium and small arteries are typical of this immune-mediated idiopathic disease. Audio-vestibular system is frequently involved with associated vertigo, tinnitus and hearing loss (67, 68).
   e. Tromboangiitis obliterans may cause sudden hearing loss (52).
   f. Systemic sclerosis (Scleroderma): In one particular study, 77% of the cases with systemic sclerosis showed abnormally low hearing levels (69).

B. Changes in the blood pressure:

It has been shown that, orthostatic fall in the blood pressure of the 10 mmHg in elderly may cause a 60% decrease in cerebral blood flow, because local humoral mechanisms are not quick or efficient enough to compensate for the insufficient supply and orthostatic hypotension is known as to cause TTS (temporary threshold shift) by means of cochlear hypoxia (70). Likewise, rapid reduction of the high blood
pressure has also been reported to cause temporary audio-vestibular symptoms (71). However, the relationship between the cochlear blood flow and hypoxia and measured auditory function is very inconsistent and erratic (12,20, 72,73).

Hemorrhagic hypotension has been demonstrated effectively reducing the blood flow in the apical turn of the cochlea in guinea pigs by Tyagi et al (74).

C Special circulatory conditions:

1. CBF rate: Yamasoba et al found an association between the sudden hearing loss and slow blood flow within the vertebro-basilar system (1). Hypoventilation has also been shown to be associated with the concurrent decrease in the CBF and endocochlear potential, suggesting a cause-effect relationship between these two entities (75). Cochlea is also found more susceptible to ischemia than the VIIIth nerve.

2. Increased blood viscosity also well correlate with hearing loss, adversely affecting RBC velocity and cochlear oxygenation. Incidence of sudden hearing loss in polycythemia and macroglobulinemia is found significantly higher in adults with SNHL than the controls of the same age group (6, 76, 77).

3. Decreased RBC deformability is characteristic of blood dyscrasia such as thalassemia and sickle cell anemia and associated with cochlear hearing loss. Its occurrence has also been shown in upper respiratory tract infections, respiratory diseases, diabetes, smoking and acidosis (6, 78).

4. Effect of noise on CBF has always been an area of controversy. However, there is controversial experimental evidence to suggest that, noise at non-physiological or potentially damaging levels, either continuous or intermittent, can produce constrictive effects on cochlear vessels, thus reducing CBF (5,73,79,80). It has also been demonstrated that acoustic overstimulation is capable of indirectly effecting CBF by angiotensin mediated systemic blood pressure elevating effect (81), which is an initial temporary increase of CBF followed by arterial constriction if noise exposure become chronic and continuous. A product of noise exposure, 8-isoprostaglandin F (2alpha), has also been demonstrated to reduce inner ear blood flow (82).

Conclusions

Vascular disorders, both local and systemic, can cause hearing loss by;

1. Creating a mass that interferes with the sound conduction,

2. Preventing the vessels of the vertebro-basilar system from providing cochlea and auditory pathways with a level of blood supply, that is quantitatively and qualitatively sufficient for their survival and normal functioning.

3. Making cochlear hair cells more susceptible to external effects, such as noise trauma.

Systemic cardio-vascular diseases tend to effect cochlear blood flow by one of several mechanisms, such as increased blood viscosity, thrombo-emboli and vasoconstriction and sometimes more than one of these factors are involved in the process.

Although there are local compensatory mechanisms, activated by ischemic conditions, their efficiency is very variable and reduced by age.

Damarsal Hastalıklarda İşitme Kaybı

Özet

Amaç: Koklea, vertebrobaziller sistemin bir uç dalı olan A. Labyrinthı tarafından tek başına kanlandılır. Bu nedenle de yerel ve sistemik damarlandırmak her türlü kistlanmaya duyarlıdır.

Son yıllarda iç kulakın mikro dolasımının incelemesinde yaşanan gelişmelerle daha önce “idiopatik” olarak sınıflandıran odyovestibüler hastalıkların önemli bir bölümüne özel hâlanın altında damarsal kaynaklı olduğu anlaşılmıştır. Bu çalışmadı, iç kulak patolojileri ve özellikle de işitme kaybı ile yerel ve sistemik damarsal hastalıkların ilişkisi üzerine mevcut literatürün araştırılması ve böylelikle konuya ilgili güncel bilgilerin anlaşılabılır bir şekilde bir araya getirilmesi amaçlanmıştır.

Gereç ve Yöntem: “Pubmed” aracılığı ile bu konuya ilgili literatür araştırılmıştır.

Sonuçlar: Mevcut araştırma için nirenliği oluşturan 82 öneleme çalışma seçilmiştir ve kullanılmıştır.

Sonuç: Bu derlemeye çalışmasında, son bulguları da içerecek şekilde, koklear kan akımının özellikleri ve bozuklukları klinik yansımaları ile özetlenmiştir.

Anahtar kelimeler: iç kulak, koklea, işitme, damarsal

References


2. Lownie SP, Parnes LS. Isolated vestibulocochlear dysfunction of central or peripheral origin. Laryngoscope 1991; 101:1339-1342.

3. Donaldson JA. Duckert LG. Anatomy of the ear


66. Yildirim N, Arslanoglu A, Aygun N. Otologic...


