



Otosclerosis: Etiology and Prognostic Factors

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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ABSTRACT

Otosclerosis is a temporal bone osseous dyscrasia that causes auditory impairment over time. Aberrant growth of a new bone in the middle ear eventually immobilizes and stops the stapes from oscillating in reaction to sound vibrations, resulting in gradual and persistent hearing impairment. Both ears are generally affected by the disease. Otosclerosis is a rapid occurrence of unexplained facial nerve palsy. With just an estimated prevalence of 15-30 per 100,000, it would be the most prevalent reason for lower motor neuron facial nerve paralysis. There are broadly two types of Otosclerosis, namely histological and clinical Otosclerosis. A minor incidence of Bell's palsy usually goes away in about a fortnight. It takes to recover from a more severe instance of complete paralysis varies. Among the potential complications are. Multiple sclerosis, Miller-Fisher syndrome, Guillain-Barré syndrome, and autoimmune encephalitis are autoimmune diseases. In patients with otitis media, malignant otitis externa damage to your facial nerve is irreversible. Nerve fiber regrowth is abnormal. When you try to move others, specific muscles may contract involuntarily (synkinesis) – for example, when you grin, the eye on the afflicted side may shut. Due to prolonged dryness and scratching of the transparent protective coating of the eye (cornea), the eye becomes wholly or partially blind. Patients report fast and increasing symptoms over a day to a week, with a 72-hour maximum severity. One-half of the face will be weak, resulting in eyebrows, forehead, and mouth angle instability. The incapacity to shut the afflicted eyelid or lip on the affected side is a common symptom. A partial or total weakening of the forehead is the most apparent physical exam finding. If the integrity of the forehead is preserved, a significant reason should be looked into. Most of the

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cases of Otosclerosis have idiopathic or unknown causative agents. In some patients with Otosclerosis, a sponge-like bone grows abnormally in the middle ear chamber. Because of this expansion, the ear bones cannot vibrate in response to acoustic pressure.

Keywords: Otosclerosis; conductive hearing loss; impairment; stiffening; tympanic membrane.

1. INTRODUCTION

The name otosclerosis comes from *oto*, which means "auricular," and *sclerosis*, which means "unusual stiffening of a tissue." It is a temporal bone osseous dyscrasia that causes auditory impairment over time. A series of events translate audio pressure waves into electrical and chemical signals within the ear, resulting in functional hearing. Similar impulses are subsequently carried to the cortex by the auditory nerve. The aberrant format neocartilagertilage in the eustachian tube finally immobilizes it and stops the stapes from oscillating in reaction to sound vibrations, resulting in gradual and persistent hearing impairment. Both ears are generally affected by the disease. This is a prevalent ailment, especially among young females. It might be brought on by pregnancy [1].

2. HISTORY

The disease of Otosclerosis was first researched and described by Antonio Maria Valsalva in 1704. In addition, he is attributed with being the one to describe spinous processes localization as a mechanism of hearing impairment. In his research, he discovered that bone ankylosing of the spinous processes to the fenestration flocculation was the most common cause of conductive hearing loss. Schwartze described Schwartze sign in 1873, which described the reddish-blue discoloration on the cochlear promontory of patients suffering from Otosclerosis. Further descriptions about the disease were given by Siebenmann, Toynbee, and Adam Pulitzer. Prosper Ménière temporarily elevated his auditory capacity by mobilizing Stapes with a gold rod [2].

This gave rise to the modern era of stapes surgeries, which included the antibiotic, which was before the era, the facade era, the mobilization epoch, and a modern stapedectomy era. Samuel Rosen accidentally introduced the world to stapedectomy surgery. He performed the surgery under local anesthesia, and the patients got immediate results after the surgery was completed. The state of present surgeries of Otosclerosis has been a long and tortuous one.

Many pioneers contributed to its modern form as we see it today. These methodologies will certainly guide surgeons in the future trends for the treatment of Otosclerosis [3].

3. EPIDEMIOLOGY

Otosclerosis is a rapid occurrence of unexplained facial nerve palsy. With just an estimated prevalence of 15-30 per 100,000, it would be the most prevalent reason for lower motor neuron facial nerve paralysis. This study aims to look at the occurrence and treatment of Bell's palsy in Sudan [3]. A retrospective cross-sectional study was conducted at Sudanese Academic Dental Hospital and Sudanese National Medical Center. The data and files of 698 Bell's palsy patients were analyzed about age, gender, location, risk factors, season, and kind of therapy. As a consequence, a total of 746 complaints were reviewed. Fifty-five percent were females, and 45% were guys, while approximately 38 percent fitted into the 21-40 year age cohort. The right-hand side of the face was injured in 57 % of the participants. December was the most prevalent month for onset, accounting for approximately 53.5 percent of all occurrences. Corticosteroids seem to be the most widely prescribed medicines, accounting for 47.3 percent of all cases [4].

4. ETIOLOGY

Otosclerosis is a bone remodeling disorder that affects the otic membrane of the adult or senior temporal bone [5]. This does not affect the temporal bone, unlike other related bone illnesses. These symptoms start with strong otic capsular bone erosion in adulthood, preceded by a healing process with bone synthesis. Although genetic elements contribute to this condition, typical measles and autoimmune disorders may also play a significant role [6].

Most of the cases of Otosclerosis have idiopathic or unknown causative agents. In some patients with Otosclerosis, a sponge-like bone grows abnormally in the middle ear chamber. Because of this expansion, the ear bones cannot vibrate in response to acoustic pressure [7].

Otosclerosis is a disease of the senior age group. Men are less susceptible than women as the latter may be prone to the disease more during pregnancy. Almost ten percent of cases are reported in Caucasian patients. Blacks and Asian races are less susceptible to it. Infections like Measles can manifest as otosclerosis in older patients. Some autoimmune diseases are also attributed to it. Genetic predisposition has been researched thoroughly. Further investigations and research should be conducted to close the gap of idiopathic causes [8].

5. CLASSIFICATION

There are broadly two types of Otosclerosis, namely histological and clinical Otosclerosis. Histological otosclerosis is when the patients do not experience significant or minor symptoms. The diagnosis can only be made by biopsy or post mortem examination. The other type is clinical otosclerosis, in which the patients experience symptoms. This group is subdivided into Stapedial, Cochlear, and Mixed Otosclerosis. Ankylosis of bones in the middle chamber occurs in Stapedial Otosclerosis. In cochlear otosclerosis, the ankylosis encroached the membranous labyrinth, producing a sensory neural hearing impairment. In mixed otosclerosis, there are symptoms of both stapedial and cochlear otosclerosis.

Another classification called the Veillon Classification subgroups otosclerosis by its site and extends into six different grades [9].

Chart 1. Veillon classification

Grade	Descriptive assessment
Ia	localized augmentation of the caused by a crop of the running board
Ib	a single 1 mm front assets of lower density (AFH)
II	>1 mm superior wearables of lower density localized (AFH)
III	>1 mm upstream wearables lower density; lower density continues to the cochlea's exoskeleton of crustaceans
IVa	Pre Cochlear lower density (PH) and infraorbital tube hypodensity are unit cells locations in the whole central portion of the esophageal cul
IVb	The whole otic membrane is involved, including the entryway and medulla oblongata.

6. PATHOPHYSIOLOGY

A series of events translate audio pressure waves into electrical and chemical signals within the ear, resulting in functional hearing. Similar impulses are subsequently carried to the cortex by the auditory nerve. Sound vibrations initially hit the outer ear and travel via the ear canal to the tympanic membrane. The inbound sound waves cause the tympanic membrane to vibrate, and the vibrations go to the malleus, incus, and stapes, three small structures in the middle ear. The cochlea, a liquid organ coiled like a snail in the inner ear, amplifies audio vibrations and sends them to the middle ear bones. A stretchy basilar membrane separates the bottom and the top regions of the cochlear apparatus and acts as the basis for essential auditory components. The liquid inside the cochlear apparatus ripples due to arriving acoustic energy, and wave propagation forms along the basilar membrane. Hair cells on the membrane's surface "float on the wave and move up and down with it. The hair cells' spiky components then collide with an overlying layer, causing the filaments to lean to one point and open pore-like passages. Specific molecules flood in, causing electrical impulses to be sent to the cortex via the auditory nerve. The ultimate product is a sound that can be recognized. More considerable pitched noises, such as with a cellular ringing phone, are detected by hair cells towards the base of the cochlear apparatus. In adulthood, such lesions start with the erosion of solid otic chamber bone, followed by a healing period with bone synthesis [8].

7. CLINICAL FEATURES

Patients report fast and increasing symptoms over a day to a week, with a 72-hour maximum severity. One-half of the face will be weak, resulting in eyebrows, forehead, and mouth angle instability. The incapacity to shut the afflicted eyelid or lip on the affected side is a common symptom. A partial or total weakening of the forehead is the most apparent physical exam finding. If the integrity of the forehead is preserved, a significant reason should be looked into. Patients may also experience changes in taste, hypersensitivity to hearing, tearing and salivation behaviors, and otalgia. Upper eyelid contraction, lagophthalmos, corneal protrusion, loss of nasolabial crease, eyebrow droop, decreased tear flow, and paralyzing ectropion of the lower lid are some of the ocular characteristics [9].

8. DIAGNOSIS AND INVESTIGATIONS

Idiopathic means that there is no known reason for the paralysis. Positive PCR to VZV in CSF in Ramsay-Hunt Syndrome.

Neoplasia: intracoronary, intradermal, or meningeal neoplasm with anatomical proximity to the facial nerve or its nucleus, tumor cells in CSF, mastoid or parotid gland neoplasia, or characteristic herpetic efflorescence in the external auditory canal.

A positive *Borrelia* antibody index diagnoses Lyme neuroborreliosis in the CSF or a positive CXCL13.

Pleocytosis >20 leukocytes/L without identification of causal pathogen in viral/bacterial CNS illness other than HSV, VZV, and *Borrelia*.

Other uncommon causes of peripheral facial palsy, such as Brucellosis and Sarcoidosis, are uncommon etiologies.

Multiple sclerosis, Miller-Fisher syndrome, Guillain-Barré syndrome, and autoimmune encephalitis are autoimmune diseases.

Patients with otitis media, malignant otitis externa, mastoiditis, or cholesteatoma are autogenous [10].

9. COMPLICATIONS

Otosclerosis, A minor incidence of Bell's palsy, usually goes away in about a fortnight. It takes to recover from a more severe instance of complete paralysis varies. Among the potential complications are:

Damage to your facial nerve is irreversible.

Nerve fiber regrowth is abnormal. When you try to move others, specific muscles may contract involuntarily (synkinesis) – for example, when you grin, the eye on the afflicted side may shut.

Due to prolonged dryness and scratching of the transparent protective coating of the eye (cornea), the eye becomes wholly or partially blind [10].

10. TREATMENT

No test can definitively determine whether or not you have Bell's palsy. Doctors frequently find out through a "judgment of limitation," as they phrase

it. That implies that they won't identify Bell's palsy throughout most situations until all other possibilities have been eliminated. A physician will begin by performing a thorough physical examination. If they suspect you have Bell's palsy, they'll attempt to close your eyelid on the affected side of your face. If that doesn't close, the patient suffers from "Bell condition," as physicians refer to it. When people try to close their eyes, it moves upward and outward due to this problem [11].

After that, the doctor will rule out any other possible causes. They'll probably put his hearing and balance to the test. They may also request various tests, including X-rays of the head, a computerized tomography (CT) scan, or imaging (MRI). Electromagnetic testing could assist you in figuring out what's wrong. It may also aid them in predicting how wholly and quickly you will heal. There isn't a specific individual who can halt it. If your doctor suspects the shingles (herpes zoster) or herpes virus (herpes simplex 1) is causing your symptoms, they may prescribe an antiviral medicine such as acyclovir. However, there is no evidence that these drugs help with Bell's palsy symptoms. The doctor may also recommend corticosteroids for a brief time (prednisone). The strategy is to create your facial nerveless swollen. This might help Bell's palsy symptoms last a short duration [12].

Likewise, the physician would advise you to take precautionary measures to preserve the damaged vision. Because you won't wink, they may advise you to wear an eye patch. If you notice that your eyes are not weeping as much as they should, you may need to use eye drops to keep them from burning out. Furthermore, the doctor might recommend you to have your face muscles massaged. They may recommend surgery to relieve pressure on the nerve in scarce circumstances if conditions are not favorable after some time [13].

Curative treatment of malignancies like acoustic neuromas and ocular schwannomas is standard. Oculoplastic surgery may be recommended for patients at high risk of developing a corneal ulcer. A variety of surgical treatments may be performed for individuals with dense facial palsy and no nerve function. These can be classified into the following categories: Nerve grafts or anastomosis are used in facial reanimation operations. Muscle translocation operations are used in facial rejuvenation procedures. Stationary operations (e.g., cosmetic surgery)

improve uniformity at rest but not movements [14].

Assess to see if individuals are correctly managing their afflicted eyes. Patients with facial palsy are more prone to experience dry eyes in the early weeks and months since the facial nerve is crucial for producing lubricant for the cornea. As a result, they risk getting corneal ulceration, resulting in vision loss in the afflicted eye. If another medical staff has not done so, the therapist should educate the patient about dry eye treatment. An immediate reference to ophthalmology is necessary if the eye seems red or the patient describes frequent episodes of redness. Otherwise, patients should be directed to an emergency room at a medical center. We offer data on the performance and tolerability of the following interventions in this literature review: antiretroviral medication, prednisolone (alone in combination with antiretroviral therapy), oxygen therapy, and face re-training [15].

A primary reason for sudden, unilateral denervation is Bell's palsy. Although the specific cause is unknown, it is assumed that the HSV-1 virus causes facial edema and, as a result, neurological problems. Luckily, the bulk of Bell's aphasia individuals regains facial functionality independently. Oral corticosteroids are recommended as initial therapy for people with Bell's palsy to recover facial muscle. Antiviral therapy is contentious. However, it is indicated when patients have lost all oral functionality [16].

11. PROGNOSIS

Otosclerosis Adults with Bell's palsy have a typically fair expectancy. The level of nerve injury determines the healing time. Most people do better within two weeks of the commencement of complaints, whether they get therapy or not, and most people regain some or complete facial expression within six months, whether they get medication or not. Some people may experience mild to severe adverse effects. Residual muscular weakening can continue or be irreversible in some circumstances [13]. People with Bell's ataxia have a typically fair outcome. In 85 percent of instances, the medical evidence of healing appears within three weeks, and most people will eventually regain neutral facial functionality. Some people may have slight residual facial impairment or have moderate to severe impairments. Bell's palsy can occur from a previous injury or illness, such as spontaneous jaw motions while blinking or inadequate

recovery of facial muscular paralysis, causing speech problems or articulating words (dysarthria) [14-21].

12. DISCUSSION

Otosclerosis is a temporal bone osseous dyscrasia that causes auditory impairment over the unusual rant growth of a new bone in the middle ear eventually immobilizes and stops the stapes from oscillating in reaction to sound vibrations in gradual and persistent hearing impairment. Both ears are generally affected by the disease. Otosclerosis is a rapid occurrence of unexplained facial nerve palsy. With just an estimated prevalence of 15-30 per 100,000, it would be the most prevalent reason for lower motor neuron facial nerve paralysis.

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Patients with otitis media, malignant otitis externa damage to your face nerve is irreversible.

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13. CONCLUSION

Most of the cases of Otosclerosis have idiopathic or unknown causative agents. In some patients with Otosclerosis, a sponge-like bone grows abnormally in the middle ear chamber. The ear bones cannot vibrate in response to acoustic pressure because of this expansion.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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