Otosclerosis: Etiology And Prognostic Factors

ABSTRACT :
Otosclerosis is a temporal bone osseous dyscrasia that causes auditory impairment over time. There is an aberrant growth of a new bone in the middle ear that 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. The time it takes to recover from a more serious instance of complete paralysis varies. Among the potential complications are. Multiple sclerosis, Miller-Fisher syndrome, Guillain–Barré syndrome, or autoimmune encephalitis are examples of autoimmune diseases. Patients with otitis media, malignant otitis externa damage to your face nerve that is irreversible.
Nerve fibre regrowth that is abnormal. When you try to move others, certain 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 partially or completely blind.
Over the course of a day to a week, patients report fast and increasing symptoms, with a 72-hour maximum in severity. One-half of the face will be weak, resulting in instability of the eyebrows, forehead, and mouth angle. 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 by far the most obvious physical exam finding. If the integrity of the forehead is preserved, a central reason should be looked into. 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 are unable to vibrate in response to acoustic pressure because of this expansion.

KEYWORDS: Otosclerosis, Conductive Hearing Loss, Impairment, Stiffening, Tympanic Membrane.

INTRODUCTION

The name otosclerosis comes from the words 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 formation of neo cartilage in the eustachian tube, which finally immobilises 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]

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. Prospere Ménière temporarily elevated his own 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 era, the facade era, the mobilisation epoch, as well as a modern stapedectomy era. Samuel Rosen accidentally introduced the world to stapedectomy surgery. He performed the surgery under local anaesthesia 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]

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. The goal of this study is to look at the occurrence and treatment of Bell's palsy in Sudan.[3] At Sudanese Academic Dental Hospital and Sudanese National Medical Center, a retrospective cross-sectional study was conducted. The data and files of 698 Bell's palsy patients were analysed in relation to 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 fitting 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 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]

ETIOLOGY

Otosclerosis is a bone remodelling disorder that affects the otic membrane of the adult or geriatric temporal bone. This does not affect the temporal bone, unlike some other related bone illnesses. In adulthood, these symptoms occur to start with erosion of robust otic capsular bone, preceded by a healing process with bone synthesis. Although there are obviously hereditary elements that contribute to this condition, measles infection typically and autoimmune disorders may also play a significant role. [5]

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 are unable to vibrate in response to acoustic pressure because of this expansion. [6]
Otosclerosis is a disease of geriatric 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 carried out for closing the gap of idiopathic causes. [7]

CLASSIFICATION

There are broadly two types of Otosclerosis, namely histological and clinical Otosclerosis. Histological otosclerosis is that in which the patients do not experience any major or minor symptoms. The diagnosis can only be done 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 virtue of its site and extends into six different grades. [8]

Grade	Descriptive assessment
Ia	localised augmentation of the caused by crop of the running board
Ib	a single 1mm front assets of lower density (AFH)
II	>1mm superior wearables of lower density localized(AFH)
III	>1mm 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.

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, which goes 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 of the cochlear apparatus ripples as a result of arriving acoustic energy, and a wave propagation forms all 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. Certain 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 recognised. Larger 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 appear to start with erosion of solid otic chamber bone, followed by a healing period with bone synthesis.[7]

CLINICAL FEATURES

Over the course of a day to a week, patients report fast and increasing symptoms, with a 72-hour maximum in severity. One-half of the face will be weak, resulting in instability of the eyebrows, forehead, and mouth angle. 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 by far the most obvious physical exam finding. If the integrity of the forehead is preserved, a central reason should be looked into. Patients may also experience changes in taste, hypersensitivity to hearing, and 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. [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, tumour cells in CSF, mastoid or parotid gland neoplasia, or characteristic herpetic efflorescence in the external auditory canal.

Lyme neuroborreliosis is diagnosed by a positive Borrelia antibody index 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, or autoimmune encephalitis are examples of autoimmune diseases.

Patients with otitis media, malignant otitis externa, mastoiditis, or cholesteatoma are otogenous. [9]

COMPLICATIONS

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

Damage to your face nerve that is irreversible.

Nerve fibre regrowth that is abnormal. When you try to move others, certain 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 partially or completely blind.[9]

TREATMENT

There is no test that 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, throughout most situations, they won't identify Bell's palsy until all other possibilities have been eliminated off. 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 is suffering from "Bell condition," as physicians refer to it. When people try to close their eyes, it moves upward and outward due to this problem.[10]

After that, the doctor will try to rule out any other possible causes. They'll probably put his hearing and balance to the test. They may also request a variety of tests, including X-rays of the head, a computerized tomography (CT) scan, or imaging (MRI). Electromagnetic testing could be able to assist you figure out what's wrong. It may also aid them in predicting how quickly and completely 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 amount of time (prednisone). The strategy is to create your facial nerve less swollen. This might help the Bell's palsy symptoms last a little duration.[11]

Likewise, the physician would advise you to take precautionary measures to preserve the damaged vision. Because you won't be able to 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 extremely rare circumstances if conditions are not favourable after a period of time. [12]

Curative treatment of malignancies like auditory neuromas and ocular schwannomas is common. Oculoplastic surgery may be recommended for patients who are 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) is used to improve uniformity at rest but not movements.[13]

Assess to see if individuals are properly managing their afflicted eyes. Patients with facial palsy are more prone to experience dry eyes in the early weeks and months since the face nerve is crucial for producing lubricant for the cornea. As a result, they are at risk of getting a corneal ulceration, which can result in vision loss in the afflicted eye. If other medical staff have not

already 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 (either alone in combination with antiretroviral therapy), oxygen therapy therapy, and face re-training. [14]

A major 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 edoema and, as a result, neurological problems. Luckily, the bulk of Bell's aphasia individuals regain facial functionality on their own. Oral corticosteroids are recommended as an initial therapy for people with Bell's palsy in order to aid facial muscle recovery. Antiviral therapy is contentious, however it is indicated in extreme situations when patients have lost all oral functionality.[15]

PROGNOSIS

Otosclerosis Adults with Bell's palsy have a typically fair expectancy. The healing time is determined on the level of nerve injury. 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 serious adverse effects. Residual muscular weakening can continue a long time or be irreversible in some circumstances.[12] People with Bell's ataxia have a typically fair outcome. In 85 percent of instances, 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).[13-20]

DISCUSSION:

Otosclerosis is a temporal bone osseous dyscrasia that causes auditory impairment over time. There is an aberrant growth of a new bone in the middle ear that eventually immobilises 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.

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