RESEARCH ARTICLE

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Otosclerosis in Iraq - is it rare disease?

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ABSTRACT

Background: A common form of conductive hearing loss in adults is otosclerosis in otosclerosis there is deposition of new spongy bone in the footplate of stapes and it is more common in females and it is presented mainly with hearing loss, although tinnitus and vertigo may occur.

Objectives: Early detection of the disease to improve the patient disability and prevent its progression and, Highlight that otosclerosis is not a rare disease in Iraq.

Methods: A retrospective study was carried out between 2019-2022 in private center of hearing and balance in Baghdad including persons who presented with hearing loss and their ages were above 20.

Results: There are 8683 persons included in the study, 52 of them were diagnosed with bilateral otosclerosis with prevalence of 0.56 for each 100 patients.

Conclusion:

1. Otosclerosis is not rare disease in Iraq.

2. Most cases presented with moderate conductive hearing loss

3. Any patient presents with conductive hearing loss should be subjected to further audiological and radiological investigations.

4. Further measures are needed to educate the population about the disease to discover it early.

Keywords: Hearing loss, Otosclerosis, Audiological, Radiological investigations, Bilateral otosclerosis.

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INTRODUCTION

Otosclerosis regarded as common problem of conductive hearing loss in adults with progressive nature.The pathology starts as spongification of the otic capsule bone in active form[1], and normal dense endochondral layer of the bony otic capsule is replaced by spongy bone laid in irregular manner,then followed by bone remodeling and sclerosis, the stapedio_vestibular interface usually involved by this remodeling process[2, 3].

The mostly affected part by such process is fissula ante fenestrum, where remnants of embryonic cartilage persist and stapedial footplate fixation may result[4].

Positive family found in more than half of patients with otosclerosis, history earlier onset of otosclerosis occurs in patients with positive family history. As shown by genetic studies the otosclerosis is (autosomal dominant pattern of inheritance) , with reduced penetrance (40%)[5-8].

Clinically, otosclerosis is occurs with higher prevalence in women than men suggesting, the hypothesis that (sex hormones) may lead to the development of disease[9- 13]. And it is more common in Whites than Black population[14, 15].

More commonly hearing loss is found between the second and the third decades[16].

In addition to the (genetic component) of otosclerosis ,other causes may have a role in its pathogenesis like measles –virus infection[17-23],fluoride in drinking water[24] and co-incidence with certain systemic connective tissue diseases[25,26].

The most frequent presentation is hearing impairment, though tinnitus and vertigo can occur.

Bilateral Hearing loss is an usual presentation that gradually worsening over many years, the typical pattern is to it starts in one ear and then progressed until it involve the other ear, the patient also may report that hearing in noisy environment(paracusis willisii) is better, though it is not specific for otosclerosis only. The patient may speak in a monotonous, low volume voice.

Tinnitus may become worser as the disease progress and becomes more severe and extensive, usually there is mild dizziness and may deteriorate with progression of the disease, resembling Meniere's disease[27].

Tympanic membrane may appear normal on examination but occasionally it has flamingo color due to promontory hyper vascularity due to active lesion this is termed Schwartz sign[28].this sign is seen in about 10% of cases[29,30].

Examination by tuning fork shows a negative Rinne test with lateralization to the ear with worse conductive hearing loss .Pure tone audiometry shows air conduction loss at low frequency, and bone conduction is normal.

Carhart notch represents a dip of the conductive bone thresholds of 20-30 db. at 2kHz ,it is not specific for otosclerosis because it may be seen in other conditions like detachment of incudostapedial joint and fixation of incus or malleus, normal discrimination score shown by speech audiometry[31].

Tympanometry is usually normal in early stage but at severe cases it is of low compliant type (As).High resolution CT-scan of the temporal bones is the imaging of choice to reach the diagnosis[32]. It helps to recognize and exclude other causes of hearing loss; the otosclerotic focus can be detected in more than 80% of cases in addition to other findings like footplate thickening and round window involvement[33].

MATERIALS AND METHODS

A retrospective study was carried out between2019-2022 in private centre of hearing and balance in Baghdad including persons who presented with hearing loss and their age were above 20 ,those with history of discharging ear or ear trauma or Eustachian tube obstruction were excluded from the study in addition to unilateral questionable conductive hearing loss.The study had been approved by the ethical and scientific committee of the institution(no.366 at 8-11-2022.

The criteria for diagnosis of otosclerosis include

1. History of recent diagnosis of conductive hearing loss.

2. Presence of air-bone gab more than 10 db. At time of presentation.

3. Low –compliant impedance curve (As) or normal (A).

4. Absence of acoustic reflex.

There are 8683 persons included in the study, 52 out of them were diagnosed with otosclerosis

Complete ENT examinations were done for them including endoscopic examinations.

All persons were subjected to hearing assessment tests including tuning fork tests, tympanometry and pure tone audiometry.

Radiological studies including CT scan were done for cases confirmed to have otosclerosis as a preoperative step for those who need surgery to exclude any associated congenital anomalies which may make the surgery difficult and not to diagnose otosclerosis.

RESULTS

Fifty two cases of otosclerosis cases were included in this study, all cases attended private otolaryngology and audio-vestibular center in Baghdad from 1st of January 2019 to 31 of December 2021 and were diagnosed for the 1st time as otosclerosis based on history , clinical examination and characteristic audiological results. Known cases of otosclerosis and cases visited the same center and they were having different diagnosis were excluded from the study, total number of patients visited the centre

in the study period was 8683 patients prevalence of otosclerosis was 0.56 for each 100 patients .

Age of the included patients was 23-48 years, mean age was 33.7; 36 female and 16 male; M/F ratio was 1:2.25

Accordingly from 52 cases included 104 ears were enrolled in this study.

(Figure 1) shows that 67 out of 104 ear (64%) had Carhart's notch in PTA while in 37 out of 104 ear (36%) ,no Carhart s notch recorded.

Audiometric analysis and findings:

From the 104 ears included in this study; only 27 ears showed mild hearing loss as first presentation of this disease; while the majority of the ears (65ears) showed moderate hearing loss and only 12 ears showed moderately severe hearing loss at first presentation (Figure 2).

Tympanometric analysis and findings:

In the majority of ears included in the study (66 ears – 63%) impedance curve was normal A; while 38 ears (37%) the curve was shallow type as (Figure 3).

Otoscopic analysis and findings:

No Schwartz's sign was identified in any ears from the 104 ears included in this study.

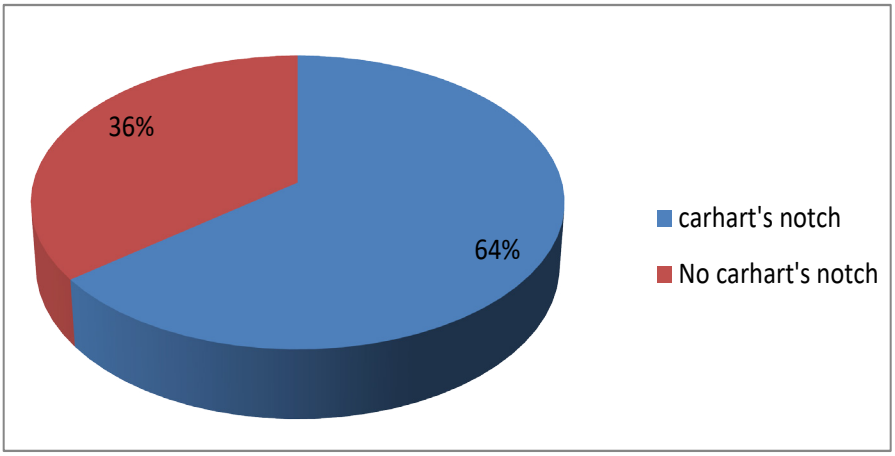

Figure 1: Shows percentage of the ears included in this study with and without Carhart's Notch in pure tone audiometry.

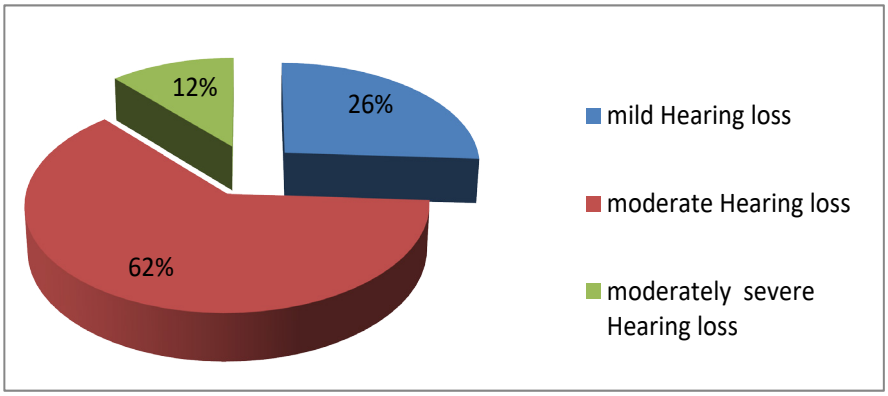

Figure 2: Percentage of ears included in this study presented with various categories of hearing loss.

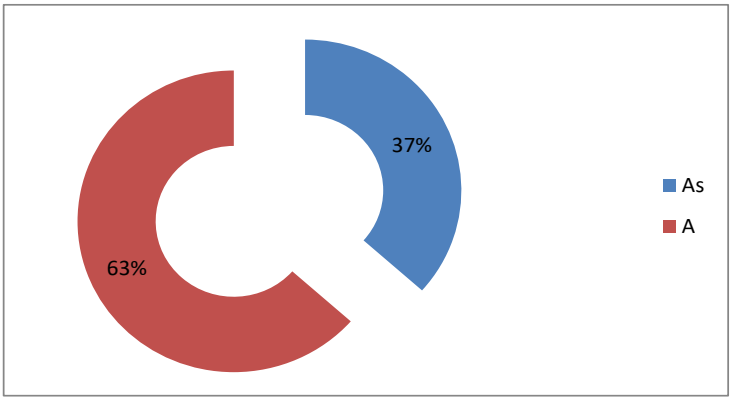

Figure 3: Percentage of impedance curve findings of the ears included in this study.

DISCUSSION

Otosclerosis is one of the cause of conductive hearing loss more common in middle age female. Different tools utilized to diagnosed otosclerosis including pure tone audiometry, traditional tympanometry, multifrequency tympanometry and high resolution CT-scan and using this tools also to differentiate otosclerosis from other cause of conductive hearing loss like middle ear pathology, tympanosclerosis, ossicular disconnection, ossicular fixation, superior semicircular canal dehiscence and other causes. In this study all patient after taking history and otoscopic examination examined by pure tone audiometry and traditional tympanometry. 8683 persons included in the study, 52 out of them were diagnosed with otosclerosis.

Age of the included patients was 23-48 year, mean age was 33.7, 36 female and 16 male, M/F ratio was 1:2.25.

Other studies reported the age of onset of the disease is at the 3 and 4 decades (62), with age of onset above the age of 50 (4%) or below the age of 10 (6%) being rare[34-36].

From the 104 ears included in this study; only 27 ears showed mild hearing loss as first presentation of this disease; while the majority of the ears (65 ears) showed moderate hearing loss and only 12 ears showed moderately severe hearing loss at first presentation.

In other studies, audiological data was obtained from 154 subjects the mean Pure-Tone Average (PTA) Air Conduction (AC) threshold was 57 dB HL, and the Bone Conduction (BC) was 28 dB HL[37, 38] and the average age of onset is (36 years)[39], the range of age involving children as young as 12 years[40], leading to progressive conductive hearing loss[41].

Otosclerosis could also cause tinnitus, and sensorineural hearing impairment with or without vestibular symptoms[42].

In the majority of ears included in the study, 66 ears (63%) impedance curve was normal A; while 38 ears (37%) the curve was shallow type As.

In other studies, the conventional type (226 Hz) tympanometry is usually not adequate for distinguishing an ear with otosclerosis from a normal middle and to differentiate otosclerosis from other middle ear pathologies[43, 44, 45].

In last years, multifrequency tympanometry type (MFT), has been found to have best diagnostic value and more accurate to differentiate patients with otosclerosis proven by surgery from other middle ear diseases[46, 47].

Accordingly from 52 cases (104 ears) were enrolled in this study, 67 out of 104 ear (64%) had a Carhart's sign, while 37 out of 104 ear (36%) hadn't.

In other studies, Carhart's sign was observed in (53.1%) of the (931) audiograms done preoperatively, mainly at (2 kHz)[48].

The presence of a (Carhart's sign) in a patient with conductive hearing impairment is not pathognomonic for otosclerosis pathology and can also occur in different middle ear pathology such as otitis media with effusion , tympanosclerosis , or ossicular deformity and malformations[49].

No Schwartz's sign was identified in any ears from the 104 ears included in this study.

CONCLUSION

Otosclerosis is not rare disease in Iraq. Any patient presents with conductive hearing loss especially with positive family history should be subjected to further radiological studies to reach the final diagnosis. Any patient presents with conductive hearing loss should be subjected to further audiological and radiological investigations. Further measures are needed to educate the population about the disease to discover it early.

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