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CLINICO-EPIDEMIOLOGICAL STUDY OF PATIENTS WITH OTOSCLEROSIS IN NEPALESE POPULATION: A TERTIARY LEVEL HOSPITAL BASED STUDY

ABSTRACT

Objective:

To analyze the clinico-epidemiological profile of patients with otosclerosis in a tertiary care center of Nepal

Materials and methods:

This was a retrospective cohort study conducted in the Department of ENT and Head & Neck Surgery, Tribhuvan University Teaching Hospital(TUTH), Institute of Medicine (IOM).Patients with surgically confirmed otosclerosis who had undergone stapes surgery were included in the study. Patient details from hospital record section with documented age of onset, family history of otosclerosis, laterality of disease, ethnicity and geographical distribution within Nepal were used for the outcome measures.

Results:

The age of onset of otosclerosis is commonly seen in adolescent and early adulthood. The incidence of unilateral and bilateral disease is 15.8% and 84.2% respectively. Higher prevalence was seen in women than men in the ratio of 1.4:1. A total of 19.7% of patients had positive family history.

Conclusion:

Otosclerosis is an adult onset disease with nearly equal incidence in both females and males mostly affecting bilateral ears rather than unilateral and approximately one-fourth with positive family history in our setting.

Keywords: Epidemiology, Otosclerosis, Stapes surgery

INTRODUCTION

Otosclerosis is a localized hereditary ailment of bone metabolism of otic capsule's enchondral bone. It is characterized by disordered resorption and deposition of bone.¹ The remodeling process often involves the stapedio-vestibular interface and can lead to fixation of stapedial footplate. Most commonly, the otosclerotic foci appear adjacent to the fissula ante fenestrum near the anterior oval window.^{1, 2} The development of otosclerosis is thought to be multifactorial, resulting from complex genetic and environmental factors. Several studies have suggested a link between the measles virus and otosclerosis.³ Among the clinical

varieties, stapedial otosclerosis is more common than the cochlear otosclerosis. The patients with the clinical type most commonly present with conductive type of hearing loss. Occasionally the lesion may involve only the cochlea, causing an isolated SNHL.⁴

On the management aspect various modality options of hearing aids and stapes surgery are offered to the patients. Among the surgical options of stapedectomy and stapedotomy; most of the clinicians prefer stapedotomy nowadays in which there is creation of a small circular fenestra in or near the center of the footplate. The limited

opening of the vestibule carries a lower risk of damage to the inner ear and resultant vertigo and/or sensorineural hearing loss.^{5,6} However, both of the interventions have shown successful outcome in improving the hearing loss to varying degree.⁷ Some studies have shown that only the piston surgery is not sufficient in moderate to advanced otosclerosis. One-stage piston surgery coupled with incus vibroplasty showed efficacy in restoring hearing loss in this group of population.⁸ treatment modalities in advanced otosclerosis with the sensorineural hearing loss (SNHL) Nowadays a small group of patients with far advanced otosclerosis are offered treatment of cochlear implantations also.

Regarding the epidemiology of the diseases, the prevalence of otosclerosis was estimated to be 2% in Caucasians and relatively less among Africans and Asians.^{9,10} The prevalence of otosclerosis in south India is 5%.¹¹ However there is no study that studies the prevalence of otosclerosis in Nepal till date. Hearing loss in females is aggravated by pregnancy thus making the prevalence of otosclerosis to be twice as frequent in females than in males.¹² Recent study in England however showed prevalence in females and males of 65% and 35% respectively.¹³ The incidence of bilateral disease is 75% and more common in females.¹⁴ the incidence of otosclerosis was 12.75%. Of the bones with otosclerosis, 56.1% belonged to men and 43.9% to women. The incidence of clinical and histologic otosclerosis was practically the same for men (44.7% to 55.3%) About 40% of the patients have positive family history of otosclerosis. Other rare causes of hearing impairment like congenital ossicular chain abnormalities and other diagnosis with incidence of 5% and 3% respectively are ruled out during the surgery.¹⁵ 437 cases (92.2%) The final standard diagnosis of otosclerosis is made during surgery by the assessment of the stapes fixation clinically.

This study was conducted in order to identify potentially distinct epidemiological profile of otosclerosis patients in context of Nepalese population visiting the tertiary center; due to variability in different study worldwide. This study further helps in application of screening test for early detection in prevalent group of population known from research which aid in early intervention and limited surgical procedure. To our

knowledge, this study in clinico-epidemiological profile of otosclerosis is the first study in Nepal.

MATERIALS AND METHODS

This study is a retrospective study done in patients who had undergone stapedotomy or stapedectomy surgery in the department of ENT and Head & Neck Surgery, Tribhuvan University Teaching Hospital in the last three years duration between April 2017 to April 2019 A.D.

All the patients with surgically confirmed otosclerosis who had undergone stapedotomy or stapedectomy were included in the study. The surgeries were performed by the senior surgeons of the department. All the surgeries were performed under local anesthesia which allowed the confirmation of hearing on table by evaluation of hearing improvement after piston prosthesis placement and also the possibility of the patients informing the surgeon if vertigo occurred per-operatively.

All the details about the patients' demography, clinical features and the surgeries were taken from hospital record section. The patients with incomplete information in hospital record were excluded from the study. The same patients with revision surgery or the patients with surgery of opposite ear in case of bilateral disease were entered for single time during data collection. All patients recruited in the study were born in Nepal and had spent the major part of their lives there. The variables are then entered in SPSS version 21.0 and analyzed.

The epidemiological evaluation further involved the assessment of the family history of otosclerosis in one or more family members concerning mainly on siblings and cousins or relatives. While taking family history, the extrinsic factor of hearing loss like noise trauma, otitis media, head trauma and miners disease had been ruled out by history. The other variables included were laterality of disease, ethnicity and geographical distribution within Nepal. The study was approved by Institutional Review committee (IRC) of Institute of Medicine, Nepal.

The data was analyzed using SPSS version 21.0 was used for the data analysis. Different demographics were compared using χ^2 test. Results of p values <0.05 were considered statistically significant.

RESULTS

Among the total of ninety-nine patients who underwent surgery for otosclerosis in 3 years duration, twenty-three patients were excluded from the study due to absence of complete information in the hospital record. Thus, finally 76 patients were analyzed in the study. The demographic characteristics of the patients were characterized in the table 1 and table 2 respectively.

Our study showed that on gender evaluation, otosclerosis was more common in cases of females (57.9%) than males (42.1%) with the ratio of females: males is 1.4:1. In the study it was found that the age of onset of the disease was in the 3rd and 4th decades (mean age 26.8 years) with onset above the age of 50 in a single case and no case was observed below the age of 10. This is as shown in figure I. The family history of otosclerosis was present in 19.7% of the cases. The incidence of bilateral disease is present in 84.2% of the cases.

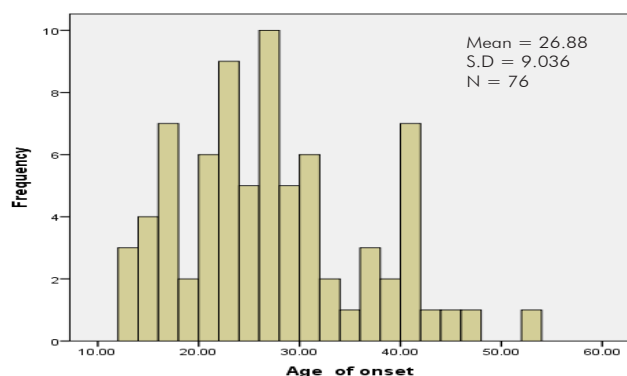


Figure I: Onset of symptoms of otosclerosis (n=76)

Table 1. Demographic Characteristics of Patients with Otosclerosis (n=76)

Variables	Frequency	Percentage(%)
Gender		
Male	32	42.1
Female	44	57.9
Age of onset		
10-19 years	16	21
20-29 years	35	46
30-39 years	14	18.4
40-49 years	10	13.1
50-59 years	1	1.3
Family History		
Yes	15	19.7
No	61	80.3
Laterality		
Unilateral	12	15.8
Bilateral	64	84.2

While reviewing the distribution of the disease according to the ethnicity in our study, we found that, among the first ten ethnic group of Nepal as per the population composition¹⁶, otosclerosis was more prevalent in Brahmins (51.3%) followed by Chhetris (18.4%) as shown in table 2.

Table 2. Distribution of Patients according to Ethnic Group (n=76)

Ethnic groups	Frequency	Percentage (%)
Brahmin	39	51.3
Chhetri	14	18.4
Newar	2	2.6
Tamang	2	2.6
Yadav	5	6.6
Nepalese muslim	3	3.9
Other	10	13.2
Tharu	1	1.3
Total	76	100.0

On evaluating the geographical distribution of the disease, the patients visiting our tertiary centre for the surgery were distributed from various parts of the country. The majority of the patients were from Kathmandu followed by Rupandehi as shown in figure II.

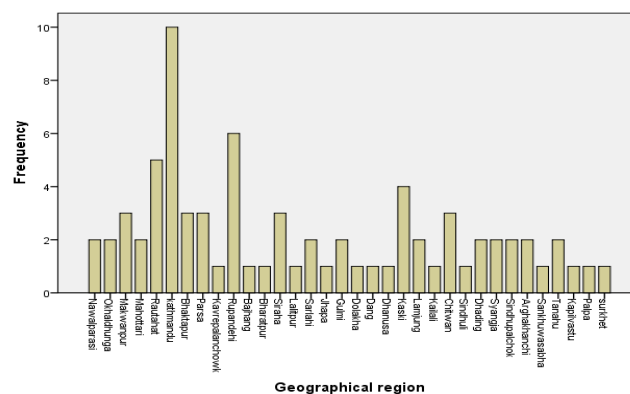


Figure II: Distribution as per geographical region (n=76)

DISCUSSION

Gender and Otosclerosis

The gender distribution in our study with female : male ratio of 1.4 : 1 corresponds with the study done by Neidermeyer et al in 2007 in Switzerland.¹⁷ It is nearly similar to the study done by Soifer et al in 1970 in which females: males ratio was 1.6:1¹⁸. Recent study in England in 2018 by Crompton et al suggested the sex ratio of 1.8:1.

Disease laterality

In our study, the incidence of bilateral disease is seen in 64 subjects (84.2%) which is quite different from the study done by Crompton et al in 2019 in England which is 62% and Migirov et al in 2014 in Germany which is 65.7% respectively.^{13,19} We found 23% of males and 15% of females have unilateral disease. Though incidence of unilateral disease is more in males than females the result is not statistically significant ($p=0.54$). There was no positive family history in 9 out of 12 (75%) subjects with unilateral disease and 12 out of 64 (18.75%) subjects with bilateral disease had positive family history.

Family History and Otosclerosis

Worldwide study of otosclerosis suggest us that it can be either familial or sporadic according to the specific group of population. We studied the profile of patients of otosclerosis comparing familial and sporadic cases. Familial cases were defined as having one or more family members with otosclerosis. In our study, 25% of females have positive family history and 23% of males have positive family history of hearing loss suggestive of otosclerosis. No significant differences were observed between familial cases and sporadic cases in terms of early onset of disease, laterality of disease involvement and specific ethnic group or with geographical region. The result of our study is similar to the study done by Shin et al in 2001 in France among 211 patients in which a family history of otosclerosis was found in 24.2% individuals.²⁰ In similar study done by Crompton et al in 2018 in England, positive family history was found in 41% of the individuals. Among the affected family members, the disease manifested irrespective of the particular gender suggestive of autosomal pattern of inheritance.

Onset of Otosclerosis

From our study it was seen that the onset of subjective hearing loss due to otosclerosis mainly become apparent after the first decade and the incidence become high at 3rd and 4th decade. However, it was rare after age of 50 years and before 10 years of age. The mean age of onset is 26.8 years with standard deviation of 9.03. The mean age of onset of otosclerosis is similar to the study done by Ben et al in 2001 in Tunisia which was 25 years.²¹ prevalence varies from 0.4% to

0.8%. The presence of a genetic factor associated with hormonal, biochemical or environmental factors, probably lead to variable expression of the otosclerosis according to age and sex. Mean age at onset of disease is 25 years (range 16-35 year). The study done by Manche et al in South India in 2016 reported the age of onset (mean $\pm$ standard deviation) as 33.7 ± 14.7 years.¹¹ The actual age at onset of deafness due to otosclerosis is not always easy to determine, mainly because of gradual onset and time gap between the involvement of both ear in cases of bilateral disease.

Ethnicity and Otosclerosis

In our study we found that, among the first ten ethnic group of Nepal as per the population composition¹⁶, otosclerosis is more prevalent in Brahmins (51.3%) followed by Chhetris (18.4%). The reason for higher prevalence in Brahmins and Chhetris may be the genetic factor. However, it may be due to the higher literacy rate among people of Brahmin ethnic group as mentioned by Subedi et al in 2016.²² It may also be due to the higher number of population of Brahmins as compared to other minor ethnicities.

Geographical Region and Otosclerosis

Mostly the patient who underwent surgery in ENT- HNS department of TUTH are from the Kathmandu, Rupandehi and Rautahat districts. Location of the tertiary care centre itself in the Kathmandu district may be the cause why prevalence is seen more in Kathmandu district due to easy accessibility for hospital care facility. In a study done by Subedi et al in 2016, the highest literacy rate (86%) was reported in Kathmandu district.²² This higher literacy rate and the easy accessibility for the hospital care among people residing in Kathmandu district may be the cause for higher prevalence of disease in Kathmandu.

Limitations and recommendation

The main limitation of this study is the retrospective data collection and which is only from single center with small sample size. Multicentric study on larger sample along with multi-generation pedigree chart analysis is highly recommended. The multicentric study would also give the representative picture of the distribution of the disease in the whole country.

CONCLUSION

Otosclerosis is an adult onset disease with nearly equal incidence in females and males in our country. The disease was seen more commonly affecting bilateral ears rather than unilateral and approximately one-fourth with positive family history. Brahmin is the most affected ethnic group.

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