

Surgery of tympanosclerosis

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Abstract

Eighty-five cases with tympanosclerosis of the middle ear were treated surgically in this series from 1984 to 1995. Twelve of them were associated with cholesteatoma and had radical surgery performed. An attempt is made here to classify the tympanosclerosis on a patho-physiological basis. A planned two-stage procedure was performed in 36 cases after an interval of 18 months. The majority of patients had stapes surgery carried out by a two-stage procedure. One of the patients who did not have stapes surgery developed anacusis after 18 months post-operatively and in another patient recurrence was observed. Post-operative hearing improvement was found to be satisfactory in the short-term follow-up period of two to five years. The hearing improvement was analysed by using Chi-square value (χ^2) and also plotted in the Glasgow Benefit Plot.

Key words: Ear, middle; Sclerosis; Surgery, operative; Hearing disorders

Introduction

Tympanosclerosis is the 'Cinderella of the middle ear disease' (Gibb, 1976), because it is a forgotten and neglected entity in the practice of otology. It literally means hardening of the middle ear (Tympanum – middle ear; sclero – hardening; and osis – condition). Literature review shows only a handful of papers published on this topic in the last half of this century. This probably is due to the fact that the condition is resistant to surgical treatment as far as the functional outcome is concerned. Also it is possible that the incidence of this sequelae of chronic ear infection is much less in the Western world due to early surgical treatment. But this condition is still prevalent in this part of the world and most of the patients are of a younger age group and commonly have bilateral conductive hearing loss. Hearing aids are very often refused by these patients.

This study has made an attempt to classify the tympanosclerotic involvement of the middle-ear cavity on a patho-physiological basis. It also provides guidelines as to which of the patients will need two-stage surgery. This decision is, of course, often very difficult to make before exploring the middle ear. Its association with cholesteatoma indicates a more serious condition which is also described here separately and is not included in the classification. It is an unsafe type of chronic middle-ear disease, the treatment of which is always radical. An observation was made about the type and degree of hearing loss in different types. Two-stage surgical procedures were recommended for the treatment of this con-

dition in order to obtain a usable post-operative hearing level.

Tympanosclerosis is a clinicopathological entity of a unique nature. It is described as an irreversible end product of inflammatory disease characterized by anatomical distortion resulting in functional impairment, that is, hearing loss mainly of the conductive type. The most common pathological changes are hyalinization, necrosis, calcification, fibrosis and ossification. Electron microscope studies revealed calcific deposits in the mitochondria and lysosomes of fibrocytes as well as degeneration of collagen fibrils in tympanosclerosis (Tos and Pederson, 1974). The diagnosis, though possible clinically, may only be made at surgery. In subclinical cases, the diagnosis is made only by histological examination. The hearing loss is related to the degree and extent of tympanosclerotic involvement of the ossicular chain.

Patients and method

Table I shows the distribution of patients according to age, sex and nationality. The mean age was 28 years and there were no children in this group. All patients presented with a history of hearing loss and clinical examination showed dry inactive ear with perforation of the ear drum and presence of tympanosclerotic plaques (Figure 1). A routine pre-operative evaluation included computed tomography (CT) scan of temporal bone in selected cases. The CT scan did not provide much information except to show soft tissue in the different parts of the middle ear cavity which could be cholesteatoma, or

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TABLE I
DISTRIBUTION OF PATIENTS

| Total number of patients: 85 | | | |
|------------------------------|----|-----------|----|
| Male | 31 | Female | 22 |
| Saudi | 38 | Non-Saudi | 15 |
| Age: 18 to 55 years | | | |

granulation tissue or tympanosclerosis. A pre-operative audiogram was performed one to 14 days before the day of surgery. Post-operative hospital stay was between five and seven days. Seventy-five patients had the soft variety, and 10 had the hard type of tympanosclerotic plaques (according to classification by Gibb, 1976).

Surgical technique Two stage procedures were carried out in 36 cases, 12 cases of type 1 category had exenteration of tympanosclerotic plaques and reconstruction done in the Stage I procedure and 25 had Stage I done and waited for Stage II. Twelve patients had modified mastoidectomy (open technique) performed due to the presence of a cholesteatoma in tympanosclerotic ears.

Stage I. All had their mastoid explored. In 52 patients tympanosclerotic plaques were found in the mastoid antrum, aditus, promontory, fallopian canal and around the round window. But these plaques were not interfering with sound transmission and therefore not mentioned in classification. Although total exenteration of tympanosclerotic plaques from the middle ear cavity and mastoid antrum are unlikely, it is possible and performed here in these cases, but was found to be time-consuming, laborious and painstaking. The tympanic membrane perforation was closed by autologous temporalis fascia. None of these cases had their

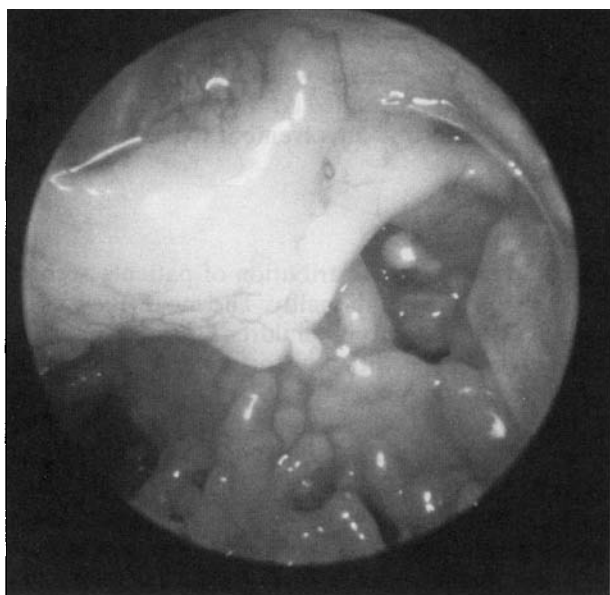


FIG. 1

Tympanosclerotic plaques involving the perforated ear drum, malleus and extending in to the tympanum.

TABLE II
CLASSIFICATION OF 85 CASES

| Classification | No. 85 | AC |
|------------------------------------|--------|-------------------|
| Type 1 | n 17 | Conductive >30 dB |
| Type 2 | n 27 | Conductive >40 dB |
| Type 3 | n 29 | Conductive >60 dB |
| Tympanosclerosis and cholesteatoma | n 12 | |

drum and ossicular chain reconstructed at the same time. Table II shows the classification of these 85 cases. In all type 1 cases the ossicular chain became mobile after removal of tympanosclerotic plaques and did not need Stage II procedure because of their hearing improvement after the surgery.

Stage II. After an interval of one year to 18 months, all type 2 and 3 patients were subjected to Stage II procedure when ossicular reconstruction was carried out behind an intact tympanic membrane (Table III).

In type 2, the incus was removed, refashioned and repositioned between the malleus and the head of the stapes in six cases, because the incudeo-stapedial joint was disrupted and the incus was found to be viable after excision of tympanosclerotic plaques. In the other seven cases, the incus was demineralized and therefore could not be re-used. A homologous incus was refashioned and transposed between the malleus and the head of the stapes. In three cases where stapes superstructure were not viable, a ceravital prosthesis was placed between the handle of the malleus and the footplate. In one of these cases the tympanosclerosis recurred so no reconstruction was attempted in Stage II procedure.

In type 3, 12 patients had their stapes removed and the tympanosclerotic deposits were cleaned from the footplate. A fenestration was then created in the footplate and a large vein graft was used to cover this opening. A Teflon fluoroplastic prosthesis was then placed between the incus and the vein graft. In seven cases a ceravital prosthesis was used between the malleus and the footplate because the incus was not viable and was discarded.

Post-operative period One patient from type 3 had a partial facial weakness on the 10th post-operative day and was treated with steroids immediately. This weakness resolved completely within two to three months. Dizziness but not vertigo was complained of by 10 patients for type 2 and 3 cases in the immediate post-operative period lasting for three to five days but none had nystagmus. They were discharged home on the seventh post-operative day.

TABLE III
STAGE II PROCEDURE (n = 36) OSSICULAR RECONSTRUCTION

| | Type 1 n 17 | Type 2 n 19 |
|---|----------------|----------------|
| Autologous incus transposition | 6 | - |
| Homologous incus transposition | 7 | - |
| Teflon Fluoroplastic incus to footplate | - | 12 |
| Ceramic prosthesis malleus to footplate | 3 | 6 |

**Post-Operative Hearing Improvement;
AC threshold in dB (n 46)**

| | Type 1 (n12) | | Type 2 (n16) | | Type 3 (n18) | |
|---|----------------|----|---------------|----|----------------|----|
| | AC | BC | AC | BC | AC | BC |
| Pre-Op | 50 | 10 | 60 | 15 | 70 | 20 |
| Post-Op | 18 | 10 | 22 | 15 | 30 | 25 |
| Gain | 32 | | 38 | | 45 | |
| Chi-Square Value (X²) | 0.1 > p > 0.05 | | 0.5 > p > 0.1 | | 0.1 > p > 0.05 | |

* Mean average of 0.5, 1 and 2 khz
** Mean average of patients

FIG. 2

Post-operative hearing improvement in AC threshold of n46 who had reconstructive surgery done (mean average of 0.5, 1 and 2 kHz).

Follow-up Post-operative follow-up was two weeks (first follow-up), four weeks (second follow-up), six weeks (third follow-up). Then every three months for two years and six months onwards. The total follow-up period was two to five years. This is a short-term follow-up.

Clinical classification

In this paper, tympanosclerotic involvement of the middle ear is classified into three different types depending on the extent of the disease and impairment of hearing (Table II). This is based on the intra-operative findings of the tympanosclerotic involvement of the ossicular chain, tympanic membrane and middle-ear cavity.

Type 1 Tympanosclerotic involvement of the tympanic membrane and malleus and anterior-superior part of the tympanic cavity.

Type 2 Tympanosclerotic involvement of incus and the incudeo-stapedial joint.

Type 3 Tympanosclerotic involvement of the stapes superstructure and the oval window.

Myringosclerosis, which is tympanosclerotic deposits in the ear drum only, is not included here in

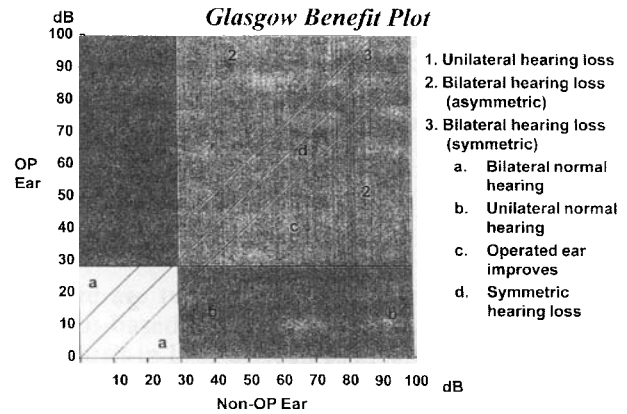


FIG. 3

Glasgow Benefit Plot.

this paper. All 12 patients who had cholesteatoma belonged to type 3 category.

Results

Two stage surgical procedures seem to have produced better results in these cases. Stapes surgery or ossicular reconstruction behind an intact ear drum appears to be useful and safe. Vein graft sealing the oval window opening after partial removal of footplate and the small fenestration technique protects the cochlea. Interestingly enough, all these ears were dry and nonactive except one with cholesteatoma which was infected at the time of surgery. Only one patient from type 3 developed anacusis 18 months after surgery where stapedectomy was not performed and another one from type 2 showed recurrence, where no repair work was carried out. Thirty-five out of 85 cases with tympanosclerosis type 2 and 3 had stapedectomy done. Thirty-four patients showed usable post-operative hearing in the short-term follow-up period of three to five years. Twelve patients from type 1 variety showed a considerable degree of hearing improvement after Stage I surgery.

The post-operative hearing improvement in these patients are shown by both a conventional method (Figure 2) and by the Glasgow Benefit Plot as described by Browning and Gatehouse in 1991

TABLE IV
GLASGOW BENEFIT PLOT (n = 46)
NUMBER OF PATIENTS IN EACH PRE-OPERATIVE IMPAIRMENT GROUP AS THEY FALL INTO EACH POST-OPERATIVE CATEGORY

| TS Type | Pre-op group | No. of patients | Post-op categories | | | |
|-----------------------------|--------------|-----------------|--------------------|----|----|----|
| | | | a | b | c | d |
| 1 n 12 | 1 | 5 | 5 | na | na | na |
| | 2 | 4 | na | 4 | na | na |
| | 3 | 3 | na | 3 | na | na |
| 2 n 17 (1 recurrence) | 1 | 6 | 6 | na | na | na |
| | 2 | 9 | na | 8 | na | na |
| | 3 | 2 | na | 1 | 1 | na |
| 3 n 19 (1 dead ear) | 1 | 4 | 4 | na | na | na |
| | 2 | 8 | na | 5 | na | 2 |
| | 3 | 7 | na | 4 | 3 | na |

na = not applicable to that specific group.

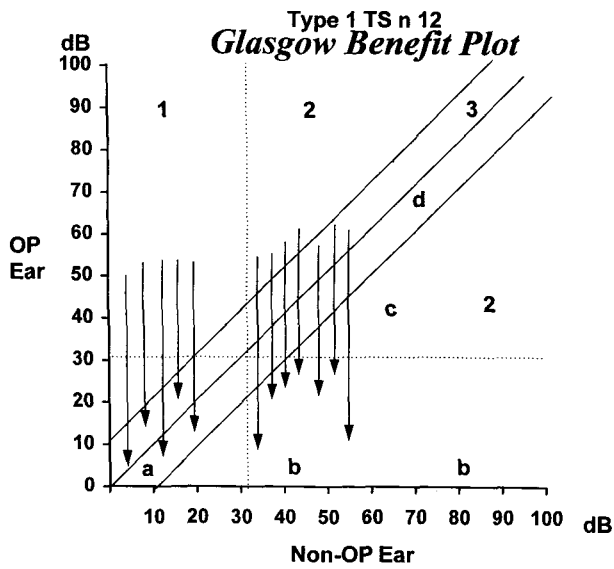


FIG. 4

Post-operative hearing improvement of type 1 patients as shown in the Glasgow Benefit Plot.

(Figures 3 and Table IV). In type 1 patients the air bone gap closure was good i.e. within 10–20 dB (Figure 4) and remained so for a long time. In type 2 patients who had incus transposition performed, either autologous or homologous, the closure of the air bone gap was found to be better (Figure 5) than with those who had ceramic prosthesis placed between the malleus and footplate.

Twelve patients from type 3 group who had a Teflon prosthesis inserted between the incus and the vein graft covering the oval window opening showed excellent results immediately after surgery (Figure 6) but air conduction threshold deterioration was observed after a period of three to five years of about 10 to 15 dB. The seven patients from this group who had a ceramic prosthesis inserted between the malleus handle and the footplate

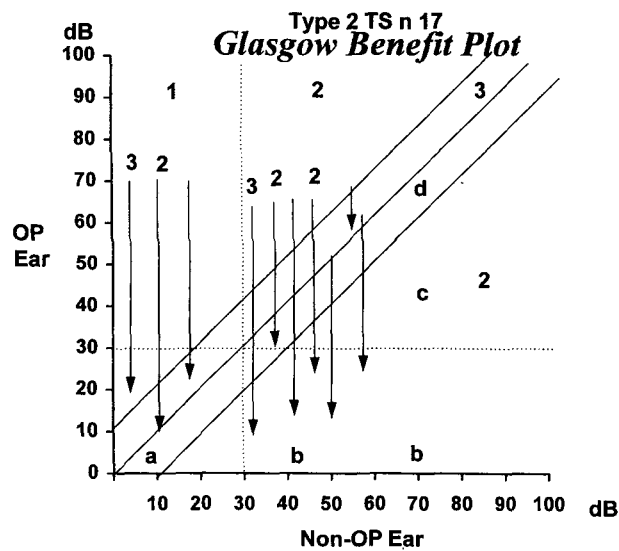


FIG. 5

Post-operative hearing improvement of type 2 patients as shown in the Glasgow Benefit Plot.

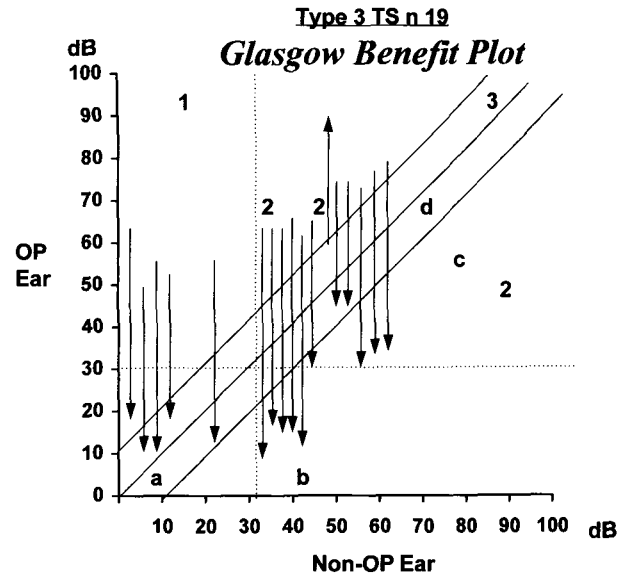


FIG. 6

Post-operative hearing improvement of type 3 patients as shown in the Glasgow Benefit Plot.

without creating an opening in the oval window achieved air bone closure of 25 to 35 dB.

The 12 patients with cholesteatoma and tympanosclerosis who had open technique tympanomastoidectomy done were followed-up regularly. They all had bilateral disease and the non-cholesteatoma ear had tympanoplasty type one done in eight patients with good post-operative hearing. The other four are still attending the clinic for routine follow-up.

Complications

Anacusis One of the patients from type 3 group who did not have stapedectomy done and where a ceramic prosthesis was used developed total hearing loss 18 months after the Stage II procedure. The hearing loss was slow, progressive and complete in one and a half year.

Recurrence Tympanosclerotic plaques recurred in one patient from type 2 variety. This was a young female patient where recurrence was first observed during the Stage II procedure after tympanotomy. The plaques were removed at that time without any ossicular reconstruction. One year later during revision tympanotomy for ossicular reconstruction, a similar recurrence was observed and therefore, no further procedure was done.

Facial palsy Transient facial palsy was observed in one case from type 3 variety after Stage II procedure who improved completely after steroid therapy. Again facial nerve was not exposed in this case.

Retraction of tympanic membrane This was observed in nine cases belonging to type 2 and 3 variety. The retraction was posteriorly but self cleansing. They are under strict observation. Surprisingly, there was no worsening of hearing in these cases, nor any symptoms related to balance.

Discussion

The first description of tympanosclerosis (TS) was made by Cassebohm in 1734 as cited by Gibb, 1976. Since then tympanosclerosis has been mentioned by many authors, Von Troltsch, 1873 as cited by Politzer's book in 1883, Walb, 1893, Zollner, 1956, House and Sheehy, 1960, Gibb, 1976 and Gormley, 1987. It seems that there was a gap of almost half a century when TS was forgotten totally in the world literature.

Tympanosclerosis is a white, firm mass in the middle ear lying between the periosteum and mucous membrane destroying neither but when occurring close to the ossicular chain it causes fixation thereby resulting in conductive hearing loss. It is not a specific pathological entity but an irreversible end-product of the inflammatory disease. Pathologists also agree with this and yet otologists notice disappointing results with refixation after mobilization and recurrence after removal. The questions asked by Zollner (1956) 'Is tympanosclerosis a specific condition and a consequence of special inflammation?' and House and Sheehy (1960) 'Is it a non-specific condition representing the terminal stage of several types of inflammation?', still remains to be answered.

Tympanosclerosis can occur anywhere in the middle ear and mastoid antrum but most commonly in the eardrum, promontory, oval window area, ossicular chain and facial nerve canal. It is not equally distributed throughout the tympanum. It destroys the ossicular chain by the process of strangulation effect on the blood supply and not by direct destructive effect. It is more common where mucosal secretory glands are scanty and ciliary activity is low in the tympanum.

Aetiology

Its aetiology is unknown and therefore, several hypotheses exist:

- (1) Precursor: severe acute otitis media either bacterial or viral (Gibb, 1976). Its association with cholesteatoma leads one to question whether non-infected cholesteatoma can transform into tympanosclerosis in a dry inactive ear.
- (2) Gland population in the middle ear: Sadé (1967) described tympanosclerosis as present in that part of middle ear where gland population is highest and the mucosa is ciliated. Whereas Tos and Pederson (1974) reported tympanic mucosa exhibits less secretory activity in tympanosclerosis than in any other diseases. The mean gland density is low and also existing glands were devoid of secretory activity.
- (3) Stagnation of secretion into the mucosal folds: but no such thing occurs in the tympanic membrane (Smyth, 1972).
- (4) Oxytalan fibres are found in tympanosclerosis plaques; they are intermediate elements of fibrogenesis and as such indicate an abnormal ground substance (Gibb, 1976).

- (5) Immunological basis, Schiff and Yoo (1985); Permeable middle ear mucosa allowing the lamina propria to be infiltrated with immunologically competent elements. Infection, inflammation and trauma will cause this permeability.

Classification

There are two types of classification of tympanosclerosis based on the morphological appearance:

(A) Harris in 1961 divided tympanosclerosis into two categories according to its behaviour:

- (1) Non-invasive and superficial called sclerosing mucositis. They tend not to recur and the periosteum remains intact.
- (2) Osteoclastic mucoperiosis. It is invasive and has a tendency to destroy the bone.

(B) Gibb in 1976 divided tympanosclerosis into two types according to its texture,

- (1) Soft, creamy, rubbery and cartilaginous texture and peels off like an onion.
- (2) Hard, white, firmly adherent and fractures during attempt to remove.

Irrespective of this classification histologically both of them appear same.

Tympanosclerosis can mimic otosclerosis when the ear drum is intact and can co-exist with cholesteatoma. Tympanosclerosis causes conductive deafness when present on the ear drum by interfering with its mobility and by fixing the handle of the malleus. In case of ossicular chain it produces a splinting effect and reduces the mobility of stapes and footplate. Besides causing upper ossicular fixation, it may also erode the long process of the incus, stapedia arch and malleus handle. The hard, infiltrating type of tympanosclerosis may cause sensorineural hearing loss also. Six of the 12 patients who had both tympanosclerosis and cholesteatoma produced a pure tone audiogram with a moderate degree of the mixed type of hearing loss.

Surgery to TS ears has not gained much attention because it was found to be the least amenable middle-ear disease for surgical treatment. Very often it was found to be an annoying complication in otherwise straightforward chronic ear surgery, particularly oval window niche tympanosclerosis. Stapedectomy in such situations is avoided by many and performed by few. So the general attitude is to avoid dealing with ossicular fixation due to tympanosclerosis by surgery for the following reasons. Hydraulic cochlear damage, 'Picking off a bit there and levering off a bit here' causes unphysiological movement of the footplate. This hydraulic effect goes around the cochlear duct. The cochlear damage is higher in mobilization and crurectomy procedures as quoted by Smyth (1980) than with a two stage procedure. Difficulty in removing tympanosclerosis plaques from around the footplate may also cause microfractures and perilymph leakage particularly in the margin of the oval window. In crurectomy, footplate fractures or dislocation may accidentally occur, ultimately leading to a perilymph fistula.

Inadvertent stapedectomy may result in rupturing the annular ligament.

A planned stapedectomy will probably avoid these difficulties because the mobilization procedure is found to have a higher incidence of re-fixation. Two stage surgery is the recommended choice of treatment in tympanosclerosis. Stage I involves eradication of disease and repair of the ear drum defect with an intact canal wall mastoidectomy. In Stage II, ossicular chain reconstruction and stapes surgery is performed behind an intact ear drum. The interval period between these two stages is one year to 18 months. The oval window opening must be closed by vein graft in order to prevent fistula formation and sensorineural hearing loss. Smyth (1972) performed two stage surgery and stapedectomy in 18 cases of tympanosclerosis with 85 per cent success, i.e. ABG closure within 10 dB and no incidence of cochlear loss. Gibb (1976) reported 138 cases of tympanosclerosis ears but stapes surgery was performed on 17 cases with 65 per cent success and one dead ear after a few years. Von Hacke (1985) performed stapedectomy in four cases of tympanosclerosis ears, two with satisfactory result, one dead ear and one with persistent hearing loss. In this series of 85 tympanosclerosis ears, stapes surgery was performed in 34 ears.

Most seriously affected patients in this series are those who belong to type 3 variety and the 12 patients with cholesteatoma. In these cases the oval window was involved extensively and the intra-operative picture was that of a small snowball replacing the whole stapes and lying on the footplate. During the surgical removal the tympanosclerosis plaques came out like onion layers. This is the soft creamy type as described by Gibb (1979) but sometimes the hard infiltrating type was also encountered on the footplate, the removal of which was painfully slow but successful. It was appropriate to perform surgery in tympanosclerosis ears because the patients were of a younger age group and the disease was bilateral. Most of the patients, particularly the females, refused a hearing aid and opted for surgery. Stage II surgery should always be preferred because ossicular reconstruction often involves creating an opening in the oval window and this must be done behind an intact ear drum. Last, but not the least, the surgery will render the ear dry inactive in those cases which might have to use a hearing aid after surgery in cases of failure to improve the hearing.

The Belfast rule of thumb was applied pre-operatively especially prior to the Stage II surgery to reduce the patients' auditory disability (Smyth and Patterson, 1985). They reported that significant patients' benefit is achieved when the post-operative average over speech frequencies is <30 dB or the intra-aural difference reduced to <15 dB, which corresponds to the cross-attenuation of the skull. The hearing threshold of the non-operated ear needs always to be considered because hearing is a bilateral sense and the central auditory system receives information from both the cochleae. The

Glasgow Benefit plot devised by Browning and Gatehouse (1991) is a useful way of reporting the results after any type of reconstructive surgery of the middle ear (Figure 3 and Table IV). Significant hearing improvement is achieved when the AC level moves from area 1 to a, 2 to b and 3 to b (Table IV). The benefit will be marginal or limited when the AC level moves from area 2 to c and 3 to c. No significant improvement will be achieved when the AC level moves from area 2 to d.

The association between tympanosclerosis and cholesteatoma is not uncommon. Harris (1961), Gristwood and Venerables (1982) and Friedmann (1974) reported a 10–13.5 per cent incidence of cholesteatoma in chronic ears with tympanosclerosis. Gibb (1976) mentioned this finding is accidental and Plester (1971) reported only one per cent of such association. But House and Sheehy (1962) and Gormley (1987) reported a much higher incidence of cholesteatoma in tympanosclerosis ears i.e. 28–43 per cent. The surgical treatment of these 12 cases was determined to be radical due to the presence of cholesteatoma. Tympanosclerosis was found intra-operatively after removal of the cholesteatoma from the middle ear. These lesions were always found to be lying deep to the cholesteatoma and were burrowing or infiltrating in type particularly in the vicinity of the oval window and the round window. The question one can ask at this stage 'is this a precursor of tympanosclerosis?', because all but one of the ears in these patients were dry and infection free. One can also possibly argue here to be more conservative in the surgical treatment i.e. to do a closed technique tympanoplasty. But with such an extensive disease this risk is probably not worth taking.

Conclusion

The astonishing amnesia about tympanosclerosis amongst otologists seems to be disappearing probably as a result of the introduction of micro ear surgery. A new classification of this disease is described in this paper depending on the extent of the disease involving the ossicular chain based on the findings during Stage I procedure. A concise and expeditious stapedectomy followed by closure of oval window opening by vein graft or connective tissue must be performed in those ears where the ear drum is healed by natural or surgical means (Smyth, 1972). Most of the authors agree that the two stage procedure gives the best possible result.

The main objective of surgery is to give the patients an intact ear drum. Since most of the cases were of a younger age group with bilateral disease, it was appropriate to reconstruct the middle ear in order to improve the hearing level.

This should be done after detailed patient counselling with explanation of the pros and cons of the surgical outcome. The surgeon has to carefully balance the benefit and risk involving the patients. The experience and the competence of the surgeon is an important aspect to consider in treating these patients.

Its association with cholesteatoma needs further investigation and research. This dual pathology in the middle ear is a matter of concern. The tympanosclerosis in such a situation is often infiltrative in type indicating advanced disease. Moreover, the location of the tympanosclerosis plaques in anatomically risky areas i.e. oval window and facial nerve, makes the surgeon's life more uncomfortable. Sometimes the facial nerve may remain exposed due to the osteolytic process of the cholesteatoma which increases the risk further. Micro-fractures of the thinned footplate may also occur while removing the tympanosclerosis plaques resulting in perilymph leakage which then needs to be covered by a vein or connective tissue graft. The common features in all these cases were a past history of otitis media, dry inactive ears and large perforations located in the pars tensa. None of these 48 cases had marginal perforation or attic defect. The surgery has the advantage of less chance of refixation and less risk to the cochlea provided the oval window opening is closed by vein graft. Very little is known about its aetiology and pathology. The final answers will probably only come from extensive combined research by otologists and pathologists.

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