The Journal of
Laryngology and Otology
(*Founded in* 1887 *by* MORELL MACKENZIE *and* NORRIS WOLFENDEN)

January 1928

OTOSCLEROSIS.*

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Introduction.—The selection of "otosclerosis" as a subject for discussion is one which needs no justification. It is a common disease, which in various ways causes much distress to the patient, and which may often involve serious financial loss. It is truly the *bête noire* of the otologist. We meet with these cases every day in practice, and have to admit that we can do little more than make a diagnosis and give the patient an indefinite idea of prognosis. So far as I know we cannot materially affect any of the symptoms or stay the disease in the least. There is scarcely a feature, pathological or clinical, that does not form a basis for discussion, so indefinite is our knowledge, nor is there any part of otology that produces such a feeling of helplessness as does this subject, upon which I feel, for this reason, particularly diffident in expressing any views. The ultimate aim of all discussion on this matter must naturally be the discovery of the fundamental cause of the disease and the means for its prevention or correction when established. This rather suggests a review of, and a discussion on, the obvious microscopic changes in the labyrinth.

A great deal of work has been done on otosclerosis both from the clinical and pathological aspect, but this has usually ended in little more than speculations on the causes of the

* Paper read at the Section of Laryngology and Otology, British Medical Association, Edinburgh, July 1927. By the courtesy of the Editor of the *British Medical Journal*. Discussion on p. 65.

https://doi.org/10.1017/S0022215100156282 Published online by Cambridge University Press

changes in the bony labyrinth or in expressions of some theory of paracusis. When I was asked to take part in this discussion I decided to take full liberty with the breadth of the subject, and, breaking away from the orthodox lines, to endeavour to stimulate the imagination by looking at the facts from another standpoint, and so perhaps evoke a new line of thought from you in your criticism and speculations. It is in this hope that I propose to deal with some features of otosclerosis and other pathological conditions that produce the otosclerosis syndrome. In this paper I am doubtless only raising for discussion a theory which has been in the minds of many of us, but which has not yet been sufficiently tested to be considered as meriting general acceptance. The term "otosclerosis" is an unfortunate one, which has given rise to much confusion, at least in the minds of students and of patients, and which is not really descriptive of the pathological or clinical phenomena; but perhaps it will be as well to retain it until we are nearer to the true pathology. In my opinion there is a clinical and pathological entity which we may call otosclerosis, but it seems that the causative pathological process may be capable of producing a variety of symptoms and signs of extremely divergent character; it also seems possible that other pathological processes may produce the same symptoms and signs with a similar divergence of character. If this be true, the next question that naturally follows is how the same pathological condition can produce a variety of symptoms and signs, and further, how different pathological conditions can produce similar syndromes.

The Otosclerosis Syndrome.—We need not for our present purpose consider in detail the typical syndrome of otosclerosis, but I should like to draw your attention to certain features relative to this matter. In 1923, I made reference to some of these points in a paper entitled "Osteitis deformans and otosclerosis," in which I quoted cases where I had been able to watch progress from the earliest stage, and I made a statement to the effect that these cases had started with a loss of bone conduction. Since reading that paper I have seen other cases that confirm the view that this is a character usually observed in the earliest stages. It is not unusual for this type, beginning in this way, to be regarded as cases of progressive internal-ear deafness with a good high tone limit, and to progress as such until the later stages, when the low tone

https://doi.org/10.1017/S0022215100156282 Published online by Cambridge University Press

limit gradually rises and a negative Rinné appears with low tones. In some instances, where there is a strong family history of deafness, the deafness may be a pure internal-ear syndrome without any admixture of signs of middle-ear deafness.

The commonest syndrome of otosclerosis is seen in the middle-ear deafness type. There are mixtures of the two extreme types in every degree, and indeed I venture to say that it is nearly always possible to demonstrate some degree of loss of bone conduction, especially of low tones, even in the most characteristic middle-ear deafness type.

The following are examples of the various otosclerosis syndromes. Edelmann forks and the monochord were used in all the examinations.

Case I.—Miss A., aged 32, suffered from deafness in both ears since early childhood, gradually becoming worse; there was no paracusis. Earache was complained of in early childhood, but without ear discharge. Tinnitus of a continual rumbling character had been present for years. Her mother was deaf, also a maternal aunt who had paracusis.

Tympanic membranes were normal. Bone conduction; diminished 50 seconds in both ears. Rinné; positive to all tones in both ears. Low tone limit; ? slightly up, perhaps 20 d.v.; high tone limit; by air conduction 21 cm., by bone conduction 17 cm. in both ears. Conversational voice; R. ear 9 inches, L. ear 3 feet. Whisper; R. ear 2 inches, L. ear 2 feet.

In this case marked internal-ear deafness without middle-ear signs and family history of deafness was quite definite.

Case II.—Mrs L., aged 45, had deafness in both ears, becoming gradually worse for ten years or more. Her husband said there was deafness in the family. Tinnitus of humming character was present for years; no history of earache, discharge, or vertigo. Tympanic membranes showed opacities.

Weber to the right side; bone conduction, diminished in both ears 40 to 50 seconds; Rinné; positive to all tones in both ears. Low tone limit, good in both ears; high tone limit, by air conduction, R. ear 32 cm., L. ear 50 cm., by bone conduction, R. ear 19 cm. L. ear 18 cm. Conversational voice; R. ear 9 feet, L. ear 3 inches. Whisper; R. ear 2 feet, L. ear 2 inches. Paracusis was well marked. Electric aids helped her very much.

Case III.—Mrs W., aged 53, had deafness in the right ear for thirteen years, in the left ear for two years; getting gradually worse. Paracusis was well marked. Tinnitus; on right side, resembled "steam

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escaping"; on left side, whistling was present for many years. Vertigo; an attack recently. She had earache occasionally, but never discharge from the ear. No family history of deafness. Tympanic membranes showed opacities.

Weber; C2 fork not heard on vertex; bone conduction diminished in both ears, 50 seconds. Rinné; positive in both ears to all tones. Low tone limit; raised in both ears to 200 d.v.; high tone limit; by air conduction, 24 cm. in both ears. Conversational voice; R. ear 1 foot, L. ear 9 inches. Whisper; R. ear 1 inch, L. ear 6 inches. Hearing was not improved by inflation. Wassermann reaction was negative.

In this patient marked internal-ear deafness, raised low tone limit, and paracusis were present. I examined her daughter, aged 20. She did not complain of any ear symptoms, and all I could find was a loss of hearing by bone conduction of 7 seconds. The labyrinths of both mother and daughter were photographed by X-rays by Dr Graham Hodgson, who reported that there was evidence of changes in the labyrinth in both patients, such as is seen in common otosclerosis.

The above are examples of the internal-ear deafness syndrome. Others could be given in this group, in which members of the family have ordinary middle-ear types of otosclerosis.

Case IV.—Mr S., aged 33, had deafness in both ears, getting gradually worse for five years. Paracusis was present. Tinnitus was complained of in both ears, off and on, since onset. There was no history of earache, discharge, or vertigo. His mother was deaf. Tympanic membranes were cicatricial.

Weber; right = left; bone conduction; diminished in both ears 15 to 20 seconds; Rinné; negative in both ears to C4. Low tone limit; raised in both ears to between 150 and 200 d.v.; high tone limit; by air conduction 17 cm. in both ears. Conversational voice; R. ear 6 inches, L. ear 6 inches. Whisper; R. ear 1 inch, L. ear 1 inch.

This is an example of mixed internal- and middle-ear types.

Case V.—Miss P., aged 30, had noticed deafness in both ears for nine months; she was getting gradually worse. No paracusis was present. Tinnitus of singing character in left ear. No history of earache, discharge, or vertigo. Father was very deaf. Tympanic membranes showed no change.

Weber; right = left; Rinné; positive to all tones in both ears. Bone conduction; diminished about 5 seconds. Low tone limit; good

https://doi.org/10.1017/S0022215100156282 Published online by Cambridge University Press

in both ears; high tone limit; down in both ears by air conduction to 87 cm., and by bone conduction to 45 cm. Conversational voice; R. ear 24 feet, L. ear 21 feet. Whisper; R. ear 6 inches, L. ear 6 inches. Wassermann reaction was negative.

Case VI.—Mrs C., aged 38, had deafness in both ears, growing gradually worse for three years. No paracusis was present. Tinnitus of buzzing character in right ear. No history of earache, discharge, or vertigo. Mother was deaf. Tympanic membranes showed opacities.

Weber to the left. Bone conduction; diminished in both ears, R. ear 25 seconds, L. ear 20 seconds. Low tone limit; raised in R. ear to 100 d.v., in L. ear to 50 d.v; high tone limit; down in both ears; by air conduction, R. ear 95 cm., L. ear 92 cm.; by bone conduction, R. ear 85 cm., L. ear 72 cm. Conversational voice; R. ear 2 inches, L. ear 12 feet. Whisper; R. ear –, L. ear 9 inches.

The labyrinths in both these cases were photographed, and Dr Graham Hodgson reported changes similar to those of otosclerosis. I have omitted an example of the common middle-ear syndrome, but the cases described may serve to show how the various types merge into one another.

Dr Graham Hodgson has recently taken X-ray photographs of the labyrinth in cases exhibiting the middle-ear deafness syndrome of otosclerosis, as well as in cases of progressive internal-ear deafness, with and without loss of high tones, and he says that the appearances of the bone changes seem to be very much the same in all.

Arguing, therefore, by deduction from clinical evidence, from microscopic sections, and possibly from X-ray examination, I think that there are grounds for believing that the variety of syndromes may be due to the same pathological process, and that the term "otosclerosis" should be used clinically for all types, the different varieties being in all probability determined by some one factor—such, for example, as the point of incidence of the morbid process. Many diseases other than otosclerosis produce similar symptoms and signs with the same extreme divergence of character. Osteitis deformans is a general disease of the bones which attacks the labyrinthine capsule from the outside, and which, so far as I know, does not start within the bony capsule of the labyrinth. Clinically and pathologically osteitis deformans and otosclerosis are different diseases. The deafness found in osteitis deformans varies considerably from a pure internal-ear deafness to an almost pure middle-ear deafness.

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Osteitis Deformans.—In the paper "Osteitis deformans and otosclerosis" referred to above, I gave an account of the aural condition of 9 cases of osteitis deformans. Since this paper was read, in 1923, I have examined 24 new cases of this general bone disease definitely affecting the skull. I propose elsewhere to give a complete account of all these cases of osteitis deformans with ear symptoms and signs, but an abstract of some of the clinical features of these 24 cases may prove of interest. There was a definite family history of deafness in 4 cases; paracusis was described or found by testing in 14 cases; bone conduction was diminished in 20 cases from 5 to 40 seconds; the low tone limit was raised in 20 cases; Rinné's test was negative in both ears to low tones in 18 cases; two of the cases probably had otosclerosis before the onset of osteitis deformans; the diagnosis was confirmed by X-ray examination in 20 cases; the Wassermann reaction has been done on most cases and found to be negative.

It is sufficient here to show that in osteitis deformans the deafness may be very similar to that of otosclerosis. The following are a few examples of deafness in osteitis deformans.

Case I.—Mr H., aged 62, had advanced osteitis deformans in the limbs, and early in the head. Deafness in both ears had come on gradually for three years. No paracusis; tinnitus of a humming character at times in both ears. No history of earache, discharge, or vertigo. Tympanic membranes with opacities. Weber; right = left. Bone conduction; diminished 20 seconds in both ears. Rinné; positive to all tones in both ears. Low tone limit; good in both ears; high tone limit; by air conduction, R. ear 28 cm., L. ear 24 cm.; by bone conduction, R. ear 16 cm., L. ear 16 cm. Conversational voice; R. ear 6 feet, L. ear 4 feet. Whisper; R. ear 3 feet, L. ear 2 feet.

This is an example of the group of cases with the internal-ear deafness syndrome.

Case II.—Mr F., aged 79, had marked osteitis deformans of the skull. Deafness in both ears was noticed about six months before examination, gradually getting worse. No paracusis; tinnitus; "bells" in both ears five months. No history of earache, discharge, or vertigo was obtained. No deafness existed in the family. Wassermann reaction was negative. Tympanic membranes showed opacities. Weber to the left side. Bone conduction; diminished in both ears about 15 seconds. Rinné; negative in both ears to C2 fork. Low tone limit; raised in both ears to about 100 d.v.; high tone limit, by air conduction, R. ear 34 cm., L. ear 65 cm.; by bone

https://doi.org/10.1017/S0022215100156282 Published online by Cambridge University Press

conduction, R. ear 17 cm., L. ear 18 cm. Conversational voice; R. ear 18 feet, L. ear 4 feet. Whisper; R. ear 9 feet, L. ear 3 feet. Hearing much better with electric aid.

Case III.—Mr L., aged 73, had noticed osteitis deformans ten to twenty years ago. Now very marked changes existed in the head. Deafness had been present in both ears twelve to fifteen years, getting gradually worse. Paracusis doubtful. No history of earache, discharge, tinnitus, or vertigo was obtained. He had four sisters, three of them deaf. His wife thinks two sisters also have bone deformities. Tympanic membranes showed opacities.

Weber to the left side. Bone conduction; diminished 10 seconds: Rinné; negative in both ears to C2. Low tone limit; raised to 100 d.v. in both ears; high tone limit; much down in both ears by air conduction. Conversational voice; R. ear 1½ feet, L. ear 2 feet. Whisper; R. ear 6 inches, L. ear 1 foot. Hears much better with electric aid.

Case IV.—Mrs W., aged 65, had very marked cephalic osteitis deformans. Deafness in both ears had been noticed for one year. Paracusis was well marked. Tinnitus of a whistling character. No history of earache or discharge for many years; no deafness in the family. Tympanic membranes were cicatricial.

Weber to the left. Bone conduction; diminished 10 to 15 seconds; Rinné; negative in R. ear to 200 d.v., in L. ear to 256 d.v. Low tone limit; raised in R. ear to 50 d.v., in L. ear to 100 d.v.; high tone limit; by air conduction, R. ear 29 cm., L. ear 30 cm.; by bone conduction, R. ear 16 cm., L. ear 16 cm. Conversational voice; R. ear 1 foot, L. ear 4 inches. Whisper; R. ear 6 inches, L. ear 4 inches.

Case V.—D., aged 60, has very marked deformity. Deafness first noticed in both ears four years ago, was getting gradually worse. No paracusis; no family history of deafness. Tympanic membranes were normal; Weber; fork on vertex not heard. Bone conduction; diminished R. ear 30 to 40 seconds, L. ear 25 seconds. Rinné; negative in R. ear to C2, and in L. ear to C1. Low tone limit; raised in R. ear to 150 d.v., in L. ear to about 40 d.v.; high tone limit; by air conduction, R. ear 25 cm., L. ear 25 cm. Conversational voice; R. ear 1 foot, L. ear 3 feet. Whisper; R. ear 1 foot, L. ear 2½ feet.

Case VI.—Mr B., aged 63, had noticed osteitis deformans for fifteen years, now very advanced. Deafness noticed for three years, was getting gradually worse. Had not observed paracusis. No history of earache, discharge, tinnitus, or vertigo. No deafness in the family. Weber; to the right side. Bone conduction; diminished in both ears; R. 10 seconds, L. 15 seconds. Rinné; negative in R. ear to C2, in

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L. ear to C1. Low tone limit; raised in R. ear to 100 d.v., normal in L.; high tone limit; slightly down by air conduction in R. ear. Conversational voice; R. ear 4 feet, L. ear 24 feet. Whisper; R. ear 2 feet, L. ear 18 feet.

Case VII.—Mr B., aged 59, had very advanced cephalic osteitis deformans; face bones had been affected for three years. Deafness in both ears for five years; paracusis present; tinnitus of buzzing character and like bells. No history of earache, discharge, or vertigo; no deafness in the family. Tympanic membranes were normal. Weber to the right side. Bone conduction; diminished about 5 seconds. Rinné; negative in both ears to C2. Low tone limit (?); high tone limit; by air conduction, R. 34 cm., L. 64 cm.; by bone conduction, R. ear 18 cm., L. ear 18 cm. Conversational voice; R. ear 3 feet, L. ear 1 inch. Whisper; R. ear 6 inches, L. ear –. Inflation did not improve. Hearing much better with electric aid.

Case VIII.—Mr J., aged 67½, was examined in 1922 because of his cephalic osteitis deformans, and I reported his case as follows in 1923, in a paper entitled "Osteitis deformans and otosclerosis."

"Male, aged 58, was not noticed to be deaf. No deafness, earache, or discharge at any time. Tinnitus a year ago of a buzzing sound. No vertigo or headache. Tympanic membranes showed opacities in both. Weber to the right. Rinné; negative to all tones below C3 in both ears. Bone conduction; slightly diminished with Edelmann special forks. High tone limit; slightly down. Low tone limit; good. Conversational voice; R. 24 feet, L. 24 feet. Whisper; R. 21 feet, L. 21 feet (after inflation).

On 28th May 1925, I examined him again, and the following is an abstract of my notes:—

Deafness had been noticed a little over one year; paracusis present; tinnitus as of "steam escaping," for one year. No history of earache, discharge, or vertigo. Tympanic membranes appeared retracted even after Eustachian catheter inflations. Weber; right = left. Bone conduction diminished in both ears 15 to 20 seconds. Rinné; negative to C3 in both ears. Low tone limit; raised to 100 d.v. in both ears; high tone limit; by air conduction, R. ear 19 cm., L. ear 30 cm.; by bone conduction, R. ear 18 cm., L. ear 18 cm. After inflation. Conversational voice; R. ear 9 inches, L. ear 2 feet. Whisper; R. ear 6 inches, L. ear 1 foot.

I think it might be expected that the middle-ear deafness syndrome should be less common in osteitis deformans than in true otosclerosis, as in the latter the point of incidence is so often close to the foramen ovale, whereas in the former the disease, starting from outside the bony capsule, must involve a

https://doi.org/10.1017/S0022215100156282 Published online by Cambridge University Press

considerable part of the capsule before reaching the neighbourhood of the foramen ovale.

It has been my experience, however, that a much larger proportion of cases show signs of obstruction deafness, with a raising of the low tone limit and negative Rinné with low tones. In many patients the loss of bone conduction was a marked feature, even when middle-ear signs were present. I am convinced that we miss many of these cases of osteitis deformans, as it is only when the changes in the skull are very gross that the deformity attracts attention. In one case (Mr L.) I made a diagnosis of internal-ear deafness, and four years later found definite middle-ear otosclerosis syndrome, and the gross changes of osteitis deformans of the skull. In another case (a female) I made a diagnosis of otosclerosis, and after six years found the patient had advanced osteitis deformans of the skull. I have no doubt that osteitis deformans was present in a marked degree at the time of my first examination of these two patients, and in one of them a dentist had noticed something abnormal in the skull shortly after my first examination. In one case, however, I think the patient must have had otosclerosis in early life and developed osteitis deformans in later life. It is possible the two diseases, otosclerosis and osteitis deformans, were concurrent, but I have cut serial sections of the temporal bone of three cases of osteitis deformans and have not found anything of the appearance of otosclerosis changes in any of them. Two of these patients were very deaf, but they were bad witnesses, and clinical examination was of no value. The third was very slightly deaf, and at the time I saw him was too ill to allow of adequate clinical examination.

We know that osteitis deformans may produce a deafness of its own. It is possible, of course, that certain types of deafness occurring in osteitis deformans, say the middle-ear deafness type, may be due to the grafting on of otosclerosis. If this proved to be so, it would then necessarily be a very common complication, but I think it is possible, and indeed probable, that osteitis deformans may in itself be capable of producing all the varieties of syndromes of otosclerosis, as there is a real similarity between these two diseases, in that both cause an osteoporosis and both are capable of producing new bone.

This may be a convenient place to refer to hyperostosis of the external auditory meatus. Some years ago Mr Waggett

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drew my attention to the fact that in these cases he found some degree of internal-ear deafness. In my experience this loss of bone conduction is almost constant in such cases; in many the loss is slight, but with care can always be demonstrated with the Edelmann fork; in some, however, the loss is marked. I mention this condition because it is another example of the association of new bone formation and a labyrinthine form of deafness.

I do not know of any other affection associated with the otosclerosis syndrome. We know of the changes occurring in the middle-ear wall of the labyrinth with chronic suppuration of the middle ear, but I do not think that the symptoms of otosclerosis have been proved to be produced by these changes. In our text-books there still lingers the description of a disease "progressive middle-ear deafness," the symptoms and signs of which are similar to those of otosclerosis. Pathological changes in the labyrinth peculiar to this condition have not been described. I have never been able to recognise this condition clinically, and venture to suggest that the cases on which this terminology was founded were really cases of otosclerosis from the beginning. Perhaps it would be as well to omit this description in our text-books until more definite evidence of its clinical entity has been found.

We may now consider the question as to how this bone disease affects the structure of the auditory apparatus so as to produce the symptoms and signs. This is a highly speculative subject and will afford much matter for discussion.

A great deal of attention has been given by pathologists to the bony changes in the region of the foramen ovale, and microscopically it has been shown how such changes must limit or alter the normal movements of the stapes. The diminished amplitude and force of the impulses transmitted through the stapes suggests a consequent obstruction deafness. This seems to be the explanation of the middle-ear type of otosclerosis syndrome, with a negative Rinné and raised low tone limit. Even if we had not the microscopic evidence of this interference, I think we should have deduced from clinical effects of disease of the middle and external ear the fact that such destruction did exist to cause the middle-ear deafness of otosclerosis.

Similarly, I think we may use other clinical and pathological facts to arrive at the mode of production of the internal-ear type of deafness. For instance, we know that an eighth nerve tumour often indicates its presence in the early stages by

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tinnitus and internal-ear deafness, and subsequently by causing a marked raising of the low tone limit. These symptoms and signs of affection of the eighth nerve by an auditory nerve tumour form a syndrome very similar to the internal-ear deafness type of otosclerosis. I feel that the similarity of these conditions is so marked that it is easy to make a mistake in diagnosis in the early stages, and I can quite well imagine the diagnosis of tumour of the eighth nerve not being verified by operation, if there were no other signs and symptoms to support the diagnosis. The tumour probably causes the auditory effects by pressure, and it is fair to deduce that in otosclerosis the internal-ear deafness syndrome may be produced by pressure on the cochlear nerves by new bone formation, such as occurs frequently at the foramen ovale.

Osteogenesis Imperfecta.—The "otosclerosis syndrome" is common in cases of osteogenesis imperfecta. In osteogenesis imperfecta there is no osteoporosis, and no new bone formation such as is found in osteitis deformans. The bones are of a delicate nature, but are complete and well formed. The following are brief accounts of the deafness of three cases:—

Case I.—Mrs W., aged 45, had typical osteogenesis imperfecta with a history of seventeen fractures before puberty. Deafness began in both ears fifteen years ago. Paracusis was present. She had tinnitus of the nature of "bells" and "steam escaping" for years. No history of earache, discharge, or vertigo was obtained. Her father was very deaf. The tympanic membranes were cicatricial.

Weber; right = left. Bone conduction; slightly diminished. Rinné; negative in both ears to C_3. Low tone limit; (?). High tone limit; by air conduction, 19 cm. in both ears. Conversational voice heard at 1 inch in both ears.

Case II.—H. W., son of the above, aged 14, had nine fractures. No deafness was noticed. Tympanic membranes were normal.

Weber; right = left. Bone conduction; (?) Rinné; negative to C_3 in L. ear and to about 100 in R. ear. Low tone limit; ? good. High tone limit; by air conduction, R. ear 16 cm., L. ear 16 cm. Conversational voice; R. ear 24 feet +, L. ear 24 feet +. Whisper; R. ear 24 feet +, L. ear 24 feet +.

Case III.—E. V., aged 55, had mental condition only fair. Deafness began at the age of 15. Paracusis was well marked. Tinnitus of a roaring character since 18 years of age. Dry postero-inferior perforation of both tympanic membranes.

Weber to left. Bone conduction; (?). Rinné; negative in both ears to C_3. Low tone limit; (?) High tone limit; by air conduction

https://doi.org/10.1017/S0022215100156282 Published online by Cambridge University Press

20 cm. in both ears; by bone conduction, 18 cm. in both ears. Conversational voice; 1 inch in both ears.

The only opportunity I have had of examining a microscopic section of a temporal bone in a case of osteogenesis imperfecta with otosclerosis syndrome occurred recently, when Mr Cleminson showed Professor Nager's specimens. It seemed to me that this was true otosclerosis grafted on the general bone disease. Recently I have cut serial sections of the temporal bone of a case of osteogenesis imperfecta without any ear symptoms, and, as might be expected, there was only the general bone changes without any signs of true otosclerosis.

When we take into account the nature of osteogenesis imperfecta and the proven presence of true otosclerosis in Professor Nager's case, I think we may say that this general bone disease in itself does not cause the syndrome of otosclerosis or of any other type of deafness. The large proportion of cases of osteogenesis imperfecta affected with otosclerosis suggests that the general disease favours the onset of otosclerosis, or that the same fundamental factor is influential in both. Perhaps in some way this intimate association of the two diseases may be an important factor in the elucidation of the main problem.

Commentary.—Out of the above the question naturally arises as to how these pathological conditions may produce the same, and also widely, divergent syndromes. There are certain characteristics in otosclerosis outside the labyrinth, in the Eustachian tube, middle-ear, tympanic membrane, and possibly in the external auditory meatus, which have been regarded by some as part of the disease, but to me they seem to be rather anatomical peculiarities associated with the disease, which is localised in the labyrinth. Perhaps these peculiarities so often referred to by clinicians are relative to an anatomical type or a localised defect that favours the onset of the bone disease of otosclerosis, just as osteogenesis imperfecta apparently does.

I believe the symptoms and signs of otosclerosis must be due to the direct extension of the disease of the bony capsule to the important elements of the auditory apparatus, and that these parts are not primarily affected as part of a general labyrinthine disease, for the following reasons. That osteoporosis may start at one of many points in the bony labyrinth, I think has now been proved microscopically. Clinically, also, I think it has been shown that a variety of symptoms may be produced by the same bone disease, and that the symptoms and

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signs may alter as the disease progresses. These facts together indicate that the syndrome probably depends on the point of incidence of the bone disease and on the direction of extension. In osteitis deformans of the skull the bony capsule of the labyrinth is very resistant to the progress of the disease, and even in advanced cases the labyrinth may stand out sharply defined in an X-ray photograph, as I showed on one occasion at the Royal Society of Medicine, but in most cases of extensive affection of the skull the labyrinth is eventually attacked and deafness is caused. If an otosclerosis syndrome can be caused by this disease, then there is strong evidence that the syndromes are produced by the bone changes attacking the labyrinth from without.

Paracusis.—There are certain clinical differences between otosclerosis deafness and obstruction or true middle-ear deafness that require explanation. I refer especially to paracusis and differences in the Rinné test results. I find it very difficult to suggest a line of discussion on the intricate problem as to how otosclerosis causes paracusis, but the following facts may be worthy of consideration.

The symptom is not always demonstrable in true otosclerosis. It may be present in both the internal- and middle-ear types of this disease. It sometimes diminishes or disappears after having been definitely present. It is absent when the high tone limit by bone conduction is very low. The absence or variation in degree of paracusis does not necessarily mean that the lesion causative of the phenomenon is absent or feeble, as it may possibly be due to the presence or absence of other conditions.

The question as to how the progressive bone disease can affect the labyrinth so as to produce this symptom has always been a subject of much discussion. Usually the discussion centres round the obvious bony changes at the foramen ovale. If, however, the phenomenon is to be explained by interference with the movements of the stapes, then the following clinical facts must be accounted for. The symptom is not necessarily present in cases of slight or great interference with the movements of the stapes by fibrous tissue in the middle ear. It may be a symptom in a case where there are no signs of obstruction deafness (with the exception, perhaps, of the raising of the low tone limit), and also in cases of very severe obstruction deafness type. In the former it is fair to suppose that the movements of the stapes would be restricted but very

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slightly, and in the latter that the stapes would be immobilised It seems that the evidence is against paracusis being caused by changes at the foramen ovale, and possibly the only clinical effects of the changes is the obstruction or middle-ear deafness elements in the syndrome.

If the interference with the stapes is not the cause, then the suggestion is that the probable site of the causative lesion must be in the terminal part of the conducting apparatus between the foramen ovale and the terminal nerves, the membranous labyrinth and the labyrinthine fluid. I have nothing at present to offer for discussion on this subject, and would invite your opinion on the various theories regarding changes in this part.

Vestibular Symptoms.—Some other symptoms and signs have not been considered, as they did not seem important to the object in view, but perhaps the vestibular symptoms should be mentioned. In my experience the vestibular symptoms are present only when the two ears are unequally affected by the otosclerosis. The sudden attacks of vertigo suggest a cause other than the possible bone changes in the neighbourhood of the vestibular nerve-endings, as the effects of the slowly progressing bone disease in these parts would probably be compensated for centrally.

Conclusion.—I have endeavoured to show that the term "otosclerosis" should be so broad as to include a variety of syndromes, and not merely the middle-ear deafness type of the otosclerosis syndrome; that osteitis deformans deafness may simulate that of otosclerosis in all types, but that in certain types the deafness may be due to grafting on of true otosclerosis; that osteogenesis imperfecta is not by itself causative of deafness, but favours the implantation of true otosclerosis; that the clinical peculiarities in the Eustachian tube, middle-ear, tympanic membrane, external auditory meatus, etc., commonly found in otosclerosis, are probably anatomical variations or a local defect, and are not due to otosclerosis—the peculiarities probably favour the onset of otosclerosis; that symptoms and signs of otosclerosis are not due to primary disease in the labyrinth, but are secondary to a capsular bone disease extending to and involving the essential elements.

I hope this unorthodox method of attack will provoke a discussion that may be helpful towards elucidation of the great problems of otosclerosis.

https://doi.org/10.1017/S0022215100156282 Published online by Cambridge University Press