

OTO-SCLEROSIS (IDIOPATHIC DEGENERATIVE DEAFNESS).

DR. ALBERT A. GRAY, Glasgow.

To the otologist who wishes something to cure, oto-sclerosis offers little, to him who wishes something to investigate it embodies the most interesting theme in otology and one second to none in the whole realm of medical science. In this paper it is proposed to review very briefly the more important aspects of the work which has been done on the subject during the last fifteen or twenty years or so.

Age Incidence.—As regards age incidence, there is general agreement that otosclerosis arises most frequently between the ages of twenty and forty, but cases under twenty and even under ten are more frequently recorded than formerly.

Sex.—It is also agreed that the disease is more frequent in women than in men in the proportion of about three to two, although Gray¹ puts it a little higher. The latter writer also points out that the number becomes much more equal in the two sexes after the age of forty.

Heredity.—The subject of heredity in otosclerosis, as in other branches of medical science, is difficult. Körner² and Hammerschlag³ attach very great importance to hereditary influence, and Gray⁴ considers that this factor is much more frequently present than is usually suspected. On the other hand, Randall⁵ estimates hereditary influence much lower as a factor than most observers. The percentage of cases which may be considered as inherited varies greatly, according to different authorities. Thus Siebenmann puts it at 35 per cent, while Bezold estimated it at 82 per cent. It is very doubtful if the percentage can be given in actual figures of any value, since different observers do not agree as to what does and what does not constitute evidence of hereditary influence.

As regards hereditary influence as shown in actual family trees, Gray,⁶ showing its occurrence in varying degrees, Hammerschlag⁷ gives an extremely interesting family tree which illustrates the case in which an uncle, who was a victim of oto-sclerosis married his niece, who was also a victim of the same disease. Seven children were born of this marriage and every one became the subject of oto-sclerosis.

A curious case is described by Paul Bonsour. A woman in whose family history no evidence of oto-sclerosis was found, and who did not suffer from the disease herself, was twice married. Neither her first husband nor her second husband suffered from deafness, but the family histories of neither husband are given. By her first husband she had one son, who became the victim of oto-sclerosis at the age of fourteen. By her second husband she had also one son, who became oto-sclerotic at the age of twelve.

In regard to the subject of hereditary influence in oto-sclerosis, there is no evidence that the Mendelian principles can be applied to this condition. Further investigation may bring about a change of view in respect to this aspect of the question, but at present the difficulties of obtaining facts in support of the application of Mendel's laws to the particular case of this disease, have not yet been overcome.

Etiology.—The question of the etiology of this disease is a very difficult one. There are few affections, local or constitutional, that have not been cited as the fundamental causative agent of oto-sclerosis. Some of the suggestions are almost bewildering. Thus, while the ovary is regarded by some writers as the evil genius of the play, others consider the testicles the offender. Now it is difficult to see how either of these structures can be looked upon as the cause of the disease, since it is very common in both men and women. Perhaps, however, there is some internal secretion which is identical in both organs, the excess or deficiency of which may be the cause of the disease. The thyroid gland, the pituitary gland, the suprarenal capsule have all been looked upon as the causative factor. Now, it must be noted that a large percentage of oto-sclerotics remain in habitual good health, which is hardly compatible with excess or deficiency of the internal secretion of any of these glands. Furthermore, when these glands are obviously diseased, as in myxedema, exophthalmic goiter, Addison's disease, acromegaly, etc., there is not found to be any particular tendency to oto-sclerosis.

Absorption of toxins from various sources have been looked upon by many as the cause of oto-sclerosis. Thus intestinal stasis with consequent absorption of poison through the bowel, appendicitis, diseased tonsils, abscess of the maxillary antrum and sphenoidal and ethmoidal sinuses have all been considered causative factors. And within narrow limits no doubt some claim may be made for these views.

Infection from the middle ear is considered by Fraser^s to be the chief factor. This is somewhat similar to the view of Haberman and there is no doubt that oto-sclerosis does sometimes occur in the course of chronic middle ear disease.

Attempts have been made to associate oto-sclerosis with various diseases of bone as having a common etiological factor, such diseases, for example, as osteo-malacia, rickets, osteo-arthritis. But so far as the present writer is aware there is no evidence that oto-sclerosis occurs with any particular frequency in these diseases.

Factors such as pregnancy, anemia, nervous exhaustion, are well-known and recognized; but, after all is said concerning these, it must be admitted that oto-sclerosis occurs more frequently when they are, individually, absent than when they are present.

Perhaps the present reviewer may be excused for suggesting that when we start to investigate the etiology of oto-sclerosis, we do not set the problem before us in the proper light. Instead of considering the problem in the light of an individual case or of a small group of individual cases, it might be better to view it from a distant point of view. Thus, oto-sclerosis consists of a change in the bony capsule of the labyrinth, bilaterally symmetrical, making its appearance very gradually and occurring in individuals who may be in ill-health from various widely different causes, but very frequently are in the best of health. Other changes in the nerve elements in the ear are frequently present and perhaps always. The other general aspect of the problem is that the majority of individuals never do suffer from oto-sclerosis, no matter how badly or from whatever cause their general health becomes affected, or whether they suffer from some other local disease of the ear, such as suppuration, middle ear catarrh, etc., or not. Now it appears to the writer that, viewed from this point of view, the logical conclusion to be drawn is that the cause of oto-sclerosis, that is the condition without which the disease cannot occur, is to be found in the organ of hearing itself, and, further, that this condition exists in the organ of hearing of certain individuals and in these individuals only. From this point of view, which the present writer believes to be the correct one, oto-sclerosis is idiopathic. That is to say, the disease occurs in people who have an inborn tendency to it, and that this is the only constant factor in the etiology. No doubt disturbances, either locally in the ears or constitutionally in the bodies of these individuals, may precipitate the onset of the disease, but no particular one of these disturbances need be present and in many cases they may all be absent.

The single constant factor in the innate tendency to the disease and the fact that hereditary influence is evident in many cases is strongly in support of this view.

Pathology.—Our knowledge of the pathological anatomy of otosclerosis has made great advances during the last twenty years, but even in this respect otosclerosis still offers many baffling problems. Regarded from the point of view of the pathologist the outstanding feature of the disease is the change which occurs in the bony capsule of the labyrinth. The exact site at which the change takes place is, in the great majority of cases, immediately in front of the oval window, but not uncommonly focuses are also found in other parts of the cochlear portion of the capsule. Very rarely, if ever, is any change found in the bony capsule of the semicircular canals. The condition is bilaterally symmetrical to a striking degree, though Manassi has described a case in which only one ear was affected by the characteristic change in the bone, the opposite ear being the seat of changes in the nerve structure of the cochlea.

Until recently it was supposed that the change lay in absorption of the normal bone, associated with the deposition of new bone, the latter being in excess of that which was absorbed. Furthermore, the new bone is found to be more porous in character than the old, and contains medullary spaces. In 1917, however, the present writer⁹ described two other types of change in the bony capsule. In one of these latter types there is absorption of bone without any deposition of new bone at all. In the third type there is both absorption and deposition of bone, but the absorption takes place more rapidly than the deposition and hence a process of continuous rarefaction takes place until the diseased area shows only a few fine trabeculae of bone, the large enclosed spaces being filled with marrow.

As regards the first, and by far the commonest type of bony change, a great deal of controversy has taken place as to the process by which it occurs. Siebenmann, Brühl, the present writer, and others, maintain that the process is not of the nature of a chronic inflammation, while Panse, Manasse, Fraser,¹⁰ Muir, Bryant and others look upon the process as being inflammatory in character, and consider it to be a form of osteitis. Now, every pathologist admits that chronic inflammation is a condition which it is not easy to define, and the present writer does not desire to express himself very strongly on this matter. It should, however, be noted that a sharp line of demarcation is not what one usually associates with chronic inflammatory activity, but rather with the outline of some

innocent tumorous or with an infarct. Round cell infiltration also is usually associated with chronic inflammation, and the present writer has failed to find such a condition in his own specimens.

The actual process by which the absorption of the normal bone occurs is also under dispute. Manassi¹¹ holds the view that the first change in the bone is not absorption, but deposition of new bone. This deposition of new bone, according to him, causes absorption of the old bone by means of simple pressure. Furthermore, Manassi maintains that the old bone is not absorbed by osteoclasts. Brühl, Siebenmann, the present writer, and others hold the view that the old normal bone is absorbed first, and that the new porous bone is subsequently deposited in its place.

The present writer ventures to think that the old bone is absorbed by osteoclasts just as the process occurs in the ordinary natural phenomena of bone physiology. It is true, as pointed out by Manassi, that frequently one does not find osteoclasts along the line of demarcation. But this is probably merely due to the fact that in the great majority of cases the disease had begun many years before the examination of the temporal bone was made and consequently activity had ceased so far as further changes in the bone were concerned. That this is actually the case is shown by the fact that as time progresses the affinity of the diseased porous bone for stains diminishes until it becomes no greater than that possessed by the old normal bone of the labyrinthine capsule. It must not, therefore, be expected that osteoclasts will be in any noticeable evidence at the line of demarcation in these cases, because absorption is no longer going on. When, however, an early case of oto-sclerosis is examined, it is found, as shown by the present writer,¹² that osteoclasts are abundantly present along the line of demarcation.

Mager is of the opinion that the absorption of the bone is brought about by the proliferation of the connective tissue cells of the marrow.

Leaving the subject of the bony change as found in the great majority of cases of oto-sclerosis, a few words may be said in respect to the two other types described by the present writer.

In one of the cases the patient was in a fairly advanced stage of phthisis pulmonalis. When the temporal bones were examined after death it was found that a change in the bone had occurred in the usual place immediately in front of the oval window. The lesion was, as is usual in oto-sclerosis, bilaterally symmetrical and there was the usual sharp line of demarcation. In the diseased area the

bone had been absorbed to a very large extent, but as new bone had been deposited, a considerable part of the diseased area consisted of little more than a network of blood vessels. It is difficult to explain the occurrence of this type of bony change, unless it be that the phthisis from which the patient suffered, interfered in some way with the deposition of bone in place of that which was absorbed.

The third type, also described by the present writer, was found under peculiar conditions. The patient died at the age of eighty-five. She had been the victim of oto-sclerosis for sixty years. On examination of the temporal bone the usual bony change of oto-sclerosis was found to be present in the usual place in front of the oval window. This had led to fixation of the stapes, and, indeed, the whole of the footplate of the stapes had undergone a similar bony change. There was found, however, another change in the bone in the region behind the oval window. This consisted of an extensive absorption of bone, associated with a very small deposit of new-formed, deeply stained bone. As a consequence of this process, the bone in this area had undergone rarefaction to a remarkable degree. The process here was evidently active at the time of death, as osteoclasts were found in the process of absorbing the old bone, and in some parts even absorbing the newly-deposited, deeply stained bone.

While great advances have been made in regard to the pathological aspect of oto-sclerosis, no great corresponding advances have been made in our knowledge of the disease as viewed from the clinical standpoint.

The causative factor of tinnitus is still in doubt, though several theories have been put forward to explain its occurrence. Neumann considers it to be due to degenerative changes in the nerve-structures and some look upon it as due to changes in the labyrinthine fluids, either chemical or physical. It may perhaps be due to pathological changes in the cortex cerebri.

The explanation of the occurrence of paracusis is still to seek. As is the case in regard to tinnitus, so also with paracusis; there has been much speculation, but little observation of new facts. No doubt this is in large part due to the inherent difficulties of investigation. The present writer ventures to suggest that these difficulties will remain until we are in possession of a much greater knowledge of nerve-physiology than we have at present.

In the matter of diagnosis, Fröschels has introduced a new test. It is based upon the diminished sensitiveness of the meatus and

tympanic membrane which occurs in oto-sclerosis. By means of a small feather attached to a probe, the membrane is greatly stimulated. If the patient does not feel a tickling sensation so much as does a normal individual the probability is that oto-sclerosis is present. The difficulty of the test lies in finding a normal standard; but with experience the investigator can often arrive at a helpful conclusion, for there is no doubt of the fact that in oto-sclerosis the sensitiveness of the structures mentioned is greatly diminished.

There has been but little advance in the treatment of the disease. Otologists are coming more and more to regard attention to the general health as the chief element in treatment. Avoidance of exhaustion, either of the mind or body, plenty of sleep, and the treatment of any constitutional affection (especially anemia in young women) are still the recommendation of those most experienced in the knowledge of oto-sclerosis.

A new method of treatment has recently been recommended by Allfell. It consists of syringing the ear with hot and cold water alternately. The temperature of the cold water should be 20° C. and that of the hot water 50° C. The present writer has tried this method and found it valueless in oto-sclerosis. Incidentally, he has found it useful in some cases of Menière's syndrome, but oto-sclerosis was absent in those cases.

BIBLIOGRAPHY.

1. GRAY: Otosclerosis, H. K., Lewis, 1917.
2. KORNER: *Zeitschr. f. Ohrenh.*, Bd. 50, S. 98.
3. HAMMERSCHLAG: *Monatschr. fr. Ohrenh.*, 1910, S. 762.
4. GRAY: *Op. cit.*
5. RANDALL: *Amer. Jour. of Med. Sci.*, July, 1910.
6. GRAY: *Op. cit.*
7. HAMMERSCHLAG: *Loc. cit.*
8. FRASER: *Jour. of Laryng., Rhinolog., and otolog.*, Jan., 1914, p. 513; also *Proc. Roy. Soc. Med., Otolog. Section*, May, 1916.
9. GRAY: *Op. cit.*
10. FRASER AND MUIR: *Jour. of Laryng., Rhinolog. and Otolog.*, Nov., 1916, p. 477.
11. MANASSI: *Die otitis chronica metaplastica*, J. F. Berfmann, 1912.
12. GRAY: *Op. cit.*, pp. 78 and 91.