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A CONTRIBUTION TO THE STUDY OF MIDDLE-EAR SUPPURATION WITH SPECIAL REFERENCE TO THE PATHOGENY AND TREATMENT OF CHOLESTEATOMA

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IT is a striking fact that although the word cholesteatoma conveys a very definite idea to the mind of every otologist, nevertheless it would be by no means easy to give a concise definition which would include its various manifestations. Let us begin by considering a few well recognized entities.

1. The primary cholesteatoma. This rare condition, in its perfect form, occurs as a distinctly encapsuled, pultaceous mass. Its outer layers are shiny keratinized skin-like sheets interspersed with cholesterin crystals. This mass lies neatly in a clear-cut cavity in the mastoid bone, or its neighbourhood. The bone is said to be covered with a fibro-vascular membrane, upon which lies a multicellular tissue which has been compared to the rete malpighi of ordinary skin. The tympanic membrane is quite intact.

2. Next, the so-called secondary cholesteatoma. Here there is definite otorrhœa which is in some way believed to be responsible for the condition. In the majority of cases, the perforation is small and lies in the pars flaccida, whilst the pars tensa is practically normal in appearance. This condition is said to be distinguishable from a primary cholesteatoma

which has undergone a secondary infection.* At operation a typical pultaceous mass will be found, although owing to the secondary infection, this is liable to be less perfectly circumscribed. Its centre may be more fluid, and its peripheral laminae less distinguishable. The cellular lining of its bony wall may be much less obvious, or, indeed, not demonstrable.

3. A third condition is apparently recognized which is said not to be cholesteatoma. There is the usual perforation in Shrapnell's membrane, associated with a scanty offensive discharge, but at operation the surgeon does not find the typical encysted, pultaceous mass. Actually a little whitish detritus lies in the aditus, whilst the main mass of the mastoid is acellular or partially sclerosed. It is difficult to see how this really differs from 2, and this distinction has led to various other fallacious hypotheses (see McKenzie see p. 688).

4. In some cases, the perforation occurs in the pars tensa just below the posterior ligament of the malleus, or there may be a large posterior perforation, with the remains of the malleus plastered on to the inner tympanic wall. Polyp formation can be encountered in any of these conditions (see later for " acute cholesteatomogenic polyp "). Such polyps are usually small and soft, dark red and vascular, quite different from the pale, firm fibrous type which I have described elsewhere as the chronic benign polyp.[14]

5. Occasionally in a dry ear, a considerable perforation is found in the pars flaccida, leading up into the attic, which is also dry and lined by a thin shiny adherent membrane. A similar appearance can be seen after any suppurating case has been cured by conservative means.

6. Post-operative mastoid cavities present very similar features to the above. They may be lined by thin shiny adherent pavement epithelium, or they may be occupied by offensive laminated pearly masses—or they may show polyps.

According to most authorities, there can no longer be any doubt that these six groups have one factor in common—namely the presence of epidermic elements, although the difficulties of explaining the presence of these same elements has resulted in a crop of hypotheses which are as ingenious as they are fantastic. This epidermoid theory is said to have received the most convincing confirmation from the pathological and

* See p. 745, Körner's criteria.

histological work of various authorities. We shall find that this is far from true. The following critical analysis of these widely accepted theories will show that not only is there little or no direct evidence of their validity, but that all the secondary difficulties which they entail can be brushed aside by an alternative hypothesis of the most elementary and convincing simplicity :—

To wit, that the six entities are in essence identical and are the natural manifestations of varying degrees of infection or other irritation acting on the pavement epithelium which is the normal lining of certain parts of the middle-ear cleft.* These parts are :—

(1) The promontory. (2) The ossicles. (3) The tympanic membrane. (4) The aditus, antrum and mastoid cells. (5) The petrous cells.

We can, in fact, subdivide middle-ear suppuration into two main types, viz.,

(1) The above " cholesteatomatous " type referable to the pavement epithelium.

(2) The muco-purulent type referable to the ciliated epithelium with its mucous glands lining the Eustachian tube, and the adjacent anterior portion of the middle-ear cleft.

In other words, the postero-superior and the antero-inferior types. Admittedly, this is closely allied to the " metaplasia " theory originally put forward by Wendt, but we shall show later that there are some minor differences.

Let us first consider the epidermoid theories.

The congenital theory naturally arose for two reasons :—

(*a*) Because of those definite, although rare, cases where the above-mentioned circumscribed mass was found almost fortuitously and in the absence of any infection or other external cause.

(*b*) Because of the very close resemblance of these masses to the cerebrospinal epidermoids found occasionally in the brain and elsewhere, and whose origin from primitive epiblast cannot be doubted.

According to Brock (writing in Denker and Kahler's Manual) Virchow gave the seal of his authority to this conception of cholesteatoma as a genuine tumour, and for a time this was

* I shall, in addition, describe later two further clinical entities—namely " Acute perforating epitympanitis " and " Acute enclosed epitympanitis " which must be added to the above six.

considered a sufficient explanation. Later, however, the frequent co-existence of middle-ear suppuration led to many efforts to explain the cholesteatoma from that point of view. The fact being that the cerebrospinal epidermoid is a very rare tumour, whereas cholesteatoma manifestations are appallingly common. (Probably at least one-third of all middle-ear suppurations.) It was obvious that true congenital anomalies could not possibly occur so frequently, and other explanations had to be sought. Unfortunately, however, the mischief was done. The epidermic theory had become an obsession. An otological *idée fixe*. McKenzie[1] made a gallant effort to bolster it up by pointing out that " the neighbourhood of the postero-superior fistula (in the pars flaccida) was a sort of developmental trysting place where various tissues tended to overlap and where accidental cell inclusion could readily occur ". We need merely repeat that in that case, we have in cholesteatoma an anomaly occurring many hundreds of times—more frequently than all other anomalies put together.

Incidentally, it may be noted that the embryology of the ear is so complicated that authors will have no difficulty in finding such anomalies wherever they want them. Thus, Jefferson and Smalley[2] in their article on " Progressive facial palsy produced by intra-temporal epidermoids " are unable to make use of McKenzie's " trysting place " for some of their cases, because these latter had an obviously different site of origin. That gap was easily filled, however, by Teed's[3] work in which a convenient epibranchial placode was implicated as the source of the requisite epiblast. (See p. 747.)

The fact is that not only is the congenital theory quite inadequate to explain the six groups and their great numbers, but, as I shall show, it is quite unnecessary even for the explanations of the rare cases in Group 1.

It is a curious thing that McKenzie himself was well aware of the claims of the metaplasia theory, which he discussed in some detail, only to reject it on the grounds that " it offered too ample an opportunity for cholesteatoma formation ". " In other words, we ought to be able to explain why so many people with long continued suppuration contrive to escape cholesteatoma." The answer to this is that they do not. Cholesteatoma (as defined by my six groups) is appallingly common. The only otorrhœas which are *not*

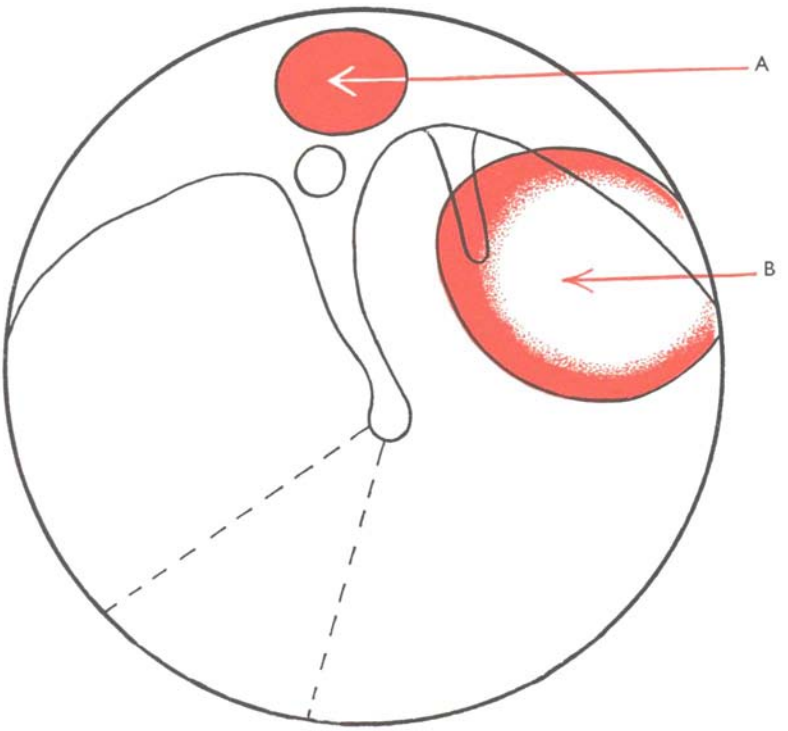

Fig. 1.

A True attic perforation.
B Post superior perforation below floor of attic.

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cholesteatoma are those antero-inferior infections of the ciliated mucous membrane.

As I have said, McKenzie's position was untenable, and long before he spoke, most authorities had abandoned it in favour of some form or other of " immigration theory ".

Bezold started this by drawing attention to the epidermization of fistulae in other parts of the body, and suggested that a like process occurred through the perforation in Shrapnell's membrane. Without delay, Habermann produced the necessary histological evidence. . . . It is interesting to speculate on what this evidence can have been based. One can readily believe that he showed some proliferation and local wandering of epidermis on the margin of the perforation, but could he have shown the continuous extension of a sheet of such epidermis up into the attic and antrum ? The answer is No, because such epidermis has never been demonstrated by anyone. This is, in fact, one of the crucial points.* Apart from the frailty of Habermann's evidence, this immigration theory has other weaknesses, which McKenzie shrewdly discusses ; nevertheless, it has been championed by Continental authorities from Politzer onwards. The true congenital type was admitted to exist, and Körner[4] laid down eight conditions for its recognition (see p. 745), but pride of place was given to this new creation which was classified as a secondary cholesteatoma.

The hunt was now up, and no feat of imagination was neglected in order to drive, or coax, epidermis into the attic.

Bezold was the first to visualize mechanical rather than inflammatory causes. He suggested that blockage of the Eustachian tube would be followed by a sucking in of the tympanic membrane. This was soon crushed by the following arguments :—

(*a*) Generalized collapse of the tympanic membrane such as would follow Eustachian blockage is rarely seen in cholesteatoma. The usual lesion is localized in Shrapnell's membrane or in the postero-superior region. Even so, it is usually a perforation rather than an " insucking " or collapse.†

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* In order not to confuse the issue I have relegated the discussion of this and other histological evidence to page 737.

† It is important to distinguish between these two situations. The posterior superior perforation is situated *below* the posterior ligament of the malleus. . The Shrapnell perforation is above it. Fig. I.

(*b*) A hermetic blockage of the Eustachian tube is rarely if ever demonstrated.

(*c*) Even if a complete collapse of the drum *did* occur, it is not easy to see how it could go on to form a cholesteatoma. A drum " plastered against the inner tympanic wall " is not uncommonly seen, it is true, but it rarely seems to produce a cholesteatoma.

These difficulties were met—at any rate partially—by Wittmaack in his monumental work on the pneumatization of the temporal bone. First he pointed out that the mucous membrane of the attic antrum space could be classified under four main types :—

(*a*) The absolutely normal. A flattish pavement epithelium resting on a fairly vascular fibrous muco-periosteum.

(*b*) The hyperplastic type in which the epithelium is cubical or even cylindrical, and the submucous tissues are much more bulky and loosely built.

(*c*) A fibrous atrophic type.

(*d*) A mixed hyperplastic fibrous type.*

In the hyperplastic cases he showed that the attic might be partially filled with this loose connective tissue. That accessory ligaments and adhesions were demonstrable between ossicles and walls, and above all that it was quite feasible for the attic antrum space to be blocked off in this way from the middle ear proper—the mesotympanum.

It is reasonable then to visualize the absorption of air in the attic, followed by a collapse of its only non-rigid wall—namely Shrapnell's membrane.

Wittmaack's album contains many drawings which support his hypothesis in so far as the hyperplastic submucosa and the localized inward collapse of the pars flaccida are easily recognizable. It must be pointed out, however, that this mechanism does not explain the frequent association of cholesteatoma with posterior superior perforations which are presumably below the level of his adhesions. Furthermore, whilst we can readily accept this *localized* collapse of the pars flaccida under purely mechanical forces, we cannot see how the epidermis can get up into the attic and antrum. To explain this, Wittmaack assumed an intense proliferative

* These, of course, must not be confused with the columnar ciliated epithelium which lines the Eustachian tube and the adjacent parts of the middle ear.

power on the part of the epidermis which enabled it to undermine the devitalized hyperplastic connective tissue. Unfortunately, nowhere in his atlas do we see any evidence of this more extensive burrowing.

The position thus arrived at was that there are three entirely different pathological processes :—

(1) The rare epidermoid tumour (Wahre cholesteatomtumor).

(2) The secondary cholesteatoma (sekundare cholesteatom), due to the invasion of epidermis following a middle-ear suppuration.

(3) The genuine cholesteatoma (genuine cholesteatom), arising by mechanical means on the basis of devitalized tissues.

This concept of tissues in the attic antrum region which actually attract the epidermis, was further modified by Albrecht[8] as follows :—

First, he claims to have demonstrated that the loose myxomatous embryonic tissue which normally fills the middle-ear cleft in embryo, and which should disappear soon after birth, often persists for a varying length of time, and is in fact frequently present in adults. He maintained that such embryonic tissue would be easily undermined by invading epidermis.

Albrecht has also shown that intra-tympanic hæmorrhage is extremely common during birth, and believes that it must have a profound influence, not only on the normal absorption of embryonic tissue, but also on the structure of the mucous membrane and of the mastoid bone itself.

Lange[9] rejects these histological findings, declares they are due to inflammatory processes and contents himself with a belief in the " gentypisch " factor. An innate predisposition to cholesteatoma, because the epidermis has a particularly strong capacity for growing inwards.

Berberich[10, 11] believes that this epidermic overgrowth is evoked by some irritation of the external auditory canal, and mentions eczema in particular. The fact that this is not common in cholesteatoma does not deter him, and he claims to have produced cholesteatoma experimentally by irritating the ear with tar and other substances. His theories and experiments have received little acceptance.

Whatever their differences, these authors agree in ascribing

an epidermic origin to cholesteatoma and postulating an innate predisposition on the part of the individual: instead of a "cholesteatomatosis" according to Voiatchek. Milstein[12] claims to have produced experimental evidence in favour of this variable susceptibility. He exposed the bullae in a series of cats and subjected them to various traumatisms and epidermic implantation, from adjacent skin areas. Similar operations were performed on dogs, and he showed that whilst it was quite easy to produce definite cholesteatomatous conditions in the cats, it was not at all possible to do this with dogs. He compares the varying susceptibility of animals to tar irritation as found by cancer research workers, and believes his work supports the theory of varying predisposition to cholesteatoma.

Despite this widespread obsession with an epidermic origin, the possible derivation of cholesteatoma by metaplasia of the ordinary epithelial lining has never been ignored. Von Tröltsch, in attacking Virchow's tumour theory, went so far as to derive the cholesteatoma from the desquamation products of epithelial cells normally present in the middle-ear cleft.

Wendt was one of the first to suggest that cholesteatoma was caused by a metaplasia of the epithelium following on otitis media.

McKenzie[1] considers this possibility carefully. He admits that clinical and experimental evidence in other regions makes it easy to believe that such metaplasia can occur into the keratin forming cells that give rise to cholesteatoma. He even notes how easily the characteristic sites of cholesteatoma can be explained by the situation of the cubical epithelium in those very sites. He was clearly impressed with the feasibility of this theory, and yet (presumably because of his obsession with the epidermic origin theory) he rejects it on entirely inadequate grounds.

In the discussion which followed McKenzie's paper, the President, A. R. Tweedie, said he

"had never been able to accept the theory that Cholesteatoma had an embryonic origin, or that they were due to an invasion of the skin from the outer meatus. In his opinion, the most tenable theory was that the process was dependent on some past sepsis—often not demonstrable—and that the irritant effect of this on the lining of the antrum and mastoid cells—which, as Neumann had

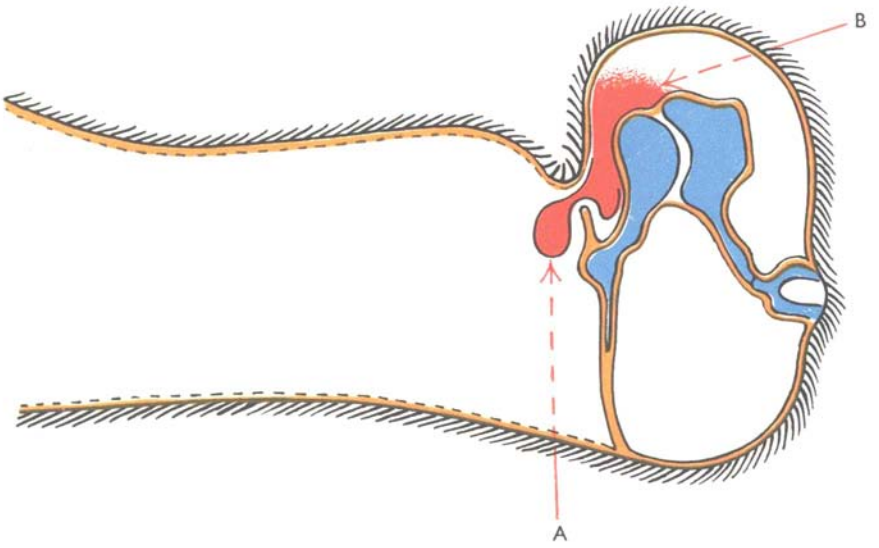

Fig. 2.

Zinc ionisation destroys the surface of the attic polyp as at A but often fails to reach the deeper part at B.

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pointed out, was a peculiar muco-periosteum—led to an accumulation of an abnormal desquamation. These factors, he suggested, would be sufficient to account for such formations, if associated with the one other factor required—a ' bottle neck ' cavity."

This, in fact, is the essence of the writer's thesis.

Yates[13] has complicated the position by accepting the neoplastic theory for primary cholesteatoma, whilst admitting the metaplasia theory for secondary cholesteatoma. He points out that otitis media falls into two main groups :—

(*a*) The leucocytic type such as occurs in the common cold. The micro-organisms are destroyed by leucocytes and there is a tendency to hypertrophy of the mucous glands, with much muco-purulent secretion. This corresponds to the writer's anterior-inferior type.

(*b*) The serous type—occurring characteristically in influenza. This causes such serious damage to the ciliated epithelial cells that they degenerate and are replaced by squamous cells, and a type of epithelium similar to skin. This desquamates off in sheets, which become rolled up in the antrum causing pressure absorption of the adjacent bone.

It will thus be seen that whilst Yates agrees with the writer in ascribing the two main types of otitis media to the two types of epithelium, he seems to have overlooked the fact that these two types of epithelium are normally present—exactly where we want them. Apparently ignoring the normal pavement epithelium of the postero-superior region, he finds it necessary to create some out of ciliated epithelium, and in order to do this, he has had to invoke his two types of infection.

We may summarize our objections to this theory as follows :

(1) To begin with, there is *no need* to postulate this dual response of the epithelium to two hypothetically different types of infection. This merely gives us on the one hand, ciliated epithelium with its muco-purulent response, and on the other hand, squamous epithelium with its cholesteatomatous response. But we already have these two types of epithelium in every normal mastoid in precisely the correct positions for the two types of otitis.

(2) Furthermore, this belief that infection can turn ciliated epithelium into pavement leads us into fresh difficulties. It will be asked immediately how is it that such a metaplasia never occurs in the antero-inferior regions of the middle-ear ?

(3) As a third objection, we might ask how it is that

cholesteatoma is (for all practical purposes) unknown in the paranasal sinuses. In these we have conditions apparently identical with those described above. Identical infections—identical ciliated epithelium lining bony cavities subject to similar mechanical obstructions. Why do we never see cholesteatoma? This amazing discrepancy has indeed led the author to suspect that the pavement epithelium is not a mere modification of the ciliated mucous membrane of the tympano tubo nasopharyngeal cleft, but is indeed profoundly different—possibly being mesoblastic in origin instead of hypoblastic (see also p. 748.)

Despite these faults, Yates' oto-pathology is an encouraging step in the right direction. We are at least thinking once more in terms of metaplasia, rather than "epidermic invasions" (even though he weakens again by conceding the title of "dermatocele").

Let us now return to our own hypothesis. This is based on the belief that the pavement epithelium which normally lines the epi and retrotympanum reacts to infection (and possibly other forms of irritation) by proliferating and throwing off paper-like squames. The exact form that this proliferation takes will vary according to associated circumstances. Suppose it occurs in a mastoidectomy cavity in which polyps and carious bone are still present. The muco-purulent discharge from these will soak up the squames and decompose them into a detritus. If, on the other hand, there is comparatively little discharge, the squames adhere to form sheets, and if there is any obstruction whatsoever, they will gradually roll up into a compact ball, the so-called cholesteatoma perle.

Similar phenomena can be visualized behind the intact tympanic membrane. We know that in acute catarrhal otitis media there is an acute inflammation without spontaneous perforation of the drum. It is easy to understand how such a condition can resolve incompletely, leaving a small focus in the epitympanum. The standard reaction occurs. A gradual exfoliation of squames—pressure erosion of bony boundaries and the formation of a cholesteatoma perle. This is the pathogeny of the so-called primary cholesteatoma.

Nor need we elaborate our hypothesis in order to explain the cases which are reported from time to time in the petrous[2, 16, 17, 18]. An identical process can be visualized just as easily in that region.

Middle-Ear Suppuration and Cholesteatoma

The preliminary stages of the primary cholesteatoma are unlikely to be noted clinically. Nevertheless, the following case, which I have labelled acute (or subacute) enclosed epitympanitis, presents features such as we should expect on theoretical grounds.

CASE I. Captain C. complained of some right otalgia about January 15th, after prolonged exposure on the bridge of his ship. The pain subsided, but the ear remained deaf. He visited aurists in Port Said, Algiers, Marseilles and other ports, and was told that he had an inflammation which would subside in due course. His middle ear was inflated, but when I saw him on February 22nd he was as deaf as ever. I found the pars tensa somewhat lacking in lustre, but it moved well under the pneumatic speculum, and there was no evidence of exudate in the mesotympanum. "*The pars flaccida was generally congested and indeed seemed to be bulging somewhat, sufficiently to obscure the normal protuberance of the short process of the malleus.*" (Incidentally, he suffered from chronic nasal catarrh—his right antrum was dark, and I strongly suspect that a sinus infection was the underlying cause of his ear infection.) Unfortunately, he could not spare the time to have his sinuses investigated, and a week later, despite some intensive diathermy, he sailed away from Liverpool as deaf as ever.

Now I cannot believe that a mere fluid exudate could remain indefinitely, and cause so much trouble as this. It is much more likely that behind that bulging pars flaccida there was a collection of squames and a small acute polyp, obstructing the movements of the incudo-malleolar joint and producing marked deafness.

What is the prognosis in such a case? Clearly it is only too liable to go on to perforation and the classical condition of secondary cholesteatoma. Such a catastrophe may easily be precipitated by injudicious syringing. How often do we see patients with typical scanty malodorous discharge from tiny perforations in the pars flaccida, who declare emphatically that they had no discharge until the ear was syringed? (See Case III, p. 707.) That they merely had consulted their family doctor on account of a deafness of one or two months' standing. Admittedly some of these cases may have had discharge without knowing it, but in other cases this seems very unlikely. One is reduced to the belief that some non-perforating process was present, causing the preliminary deafness, and that the syringing converted it into the classical

of fact, although the onset of epitympanitis is usually so insidious, I have observed two such cases in the early stage of the perforation, and, on closer questioning, each admitted to definite, though slight, preliminary otalgia. Otto Mayer[15] has also given an account of this acute epitympanitis which he has observed in two cases. Because at operation he also found cholesteatomatous squames in the epitympanum, he maintained that the ingrowth of epidermis can occur with great rapidity in the course of acute otitis media—a further example of the lengths to which clinicians may be led by their unhesitating acceptance of the "epidermic immigration" theory.

This conception of a perforation of the pars flaccida occurring as a result of erosion by an epitympanic polyp, has always seemed to me more reasonable than the alternative, namely, that it was caused by pressure of accumulated fluid discharge. In the latter case, we have to assume that the attic is completely blocked off from the mesotympanic space by the folds of Tröltsch, so that the fluids cannot seep down it. Clearly, if fluid could seep down under gravity, we should get the classical bulging of the drum and the ordinary central perforation. Such a water-tight blocking off may possibly occur, as Wittmaack claims, but we really need not postulate it, if we accept the polyp theory. It is easy to visualize such a polyp, imprisoned within the narrow confines of the attic, gradually but inexorably increasing in size, soaking the pars flaccida into a sodden condition and then quietly eroding through.

It should be noted that these conditions, polyps and squames, are both superficial phenomena. Although the bone is gradually eroded by pressure (others say by actual solution), nevertheless caries, necrosis and sequestrum formation are rare and late results, presumably due to vascular deprivation. Consequently in the majority of cases, the correct treatment will be rewarded by complete cure. Treatment consists in laying open the cavity. If this is done adequately, whether by nature or by the surgeon, squames can no longer accumulate and the vicious circle is broken. If, at the same time, any polyps are treated, the whole cavity rapidly acquires the above-mentioned thin, shiny lining, a complete *restitutio ad integrum*.

In certain cases, unfortunately, desquamation continues most obstinately and, unless the patient is kept under close

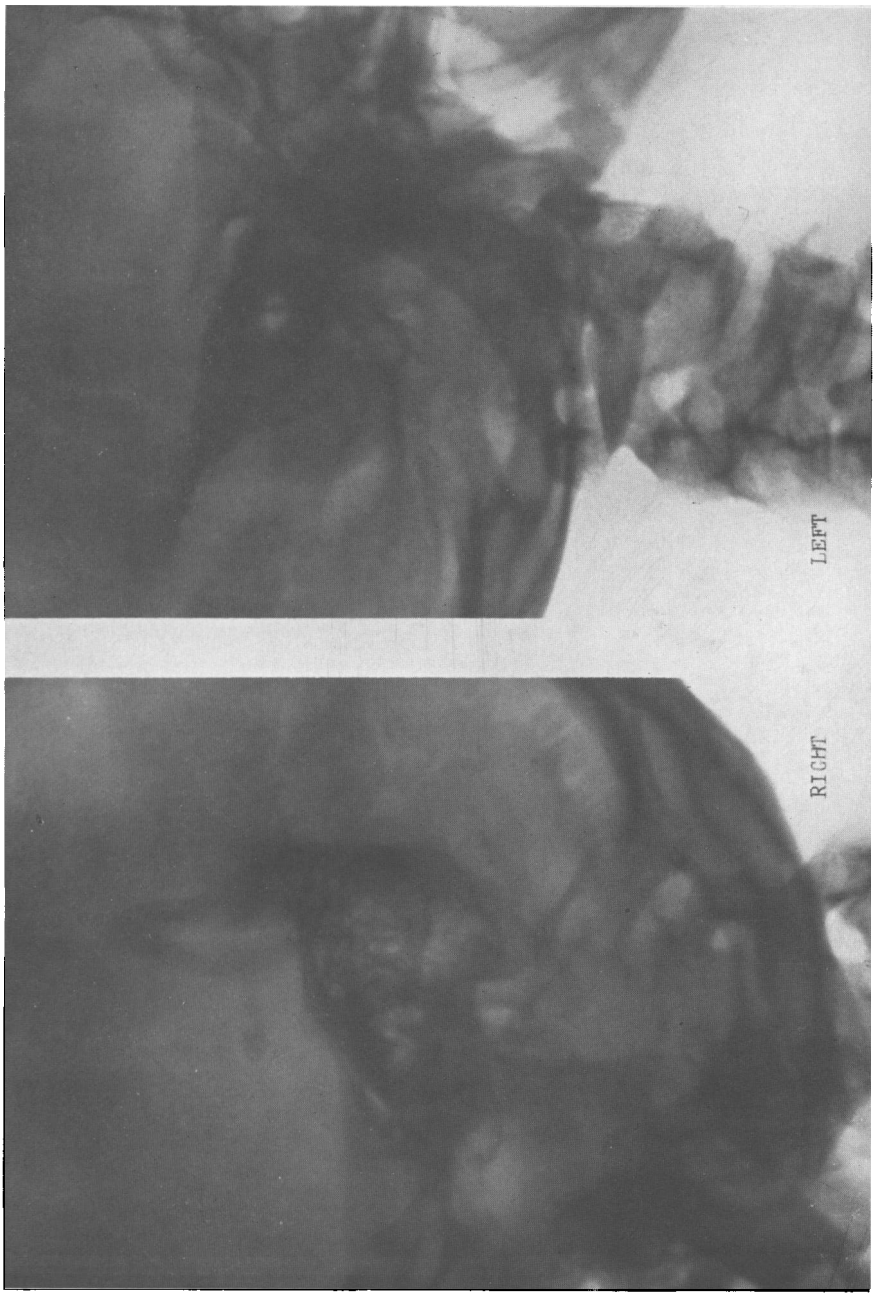

Fig. 3.

Mastoid asymmetry. Right, poorly pneumatized. Left, diploetic.

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observation, the accumulation accelerates under the effect of its own local irritation. Polyps reappear, and the patient returns with a fully developed "cholesteatoma". This relapse is only too likely to occur in regions such as the attic where anatomical and mechanical causes make exfoliation and desiccation difficult. In other cases, treatment may have been terminated too soon. The multicellular layers may not have been completely coaxed back to the normal pavement epithelium condition. Even a quite small patch can be the starting point for such a recurrence, and it goes without saying that a focus of caries will have a similar effect. This inability to cope with the products of its reactions distinguishes the pavement epithelium very clearly from the ciliated mucous membrane. Assuming there is no underlying bone disease, a mesotympanitis can be readily and easily cured (by ionization, or similar classical methods). Relapse only occurs if further infections arrive, either *viâ* the Eustachian tube or the blood stream, and such relapses frequently resolve spontaneously. Not so with lesions involving pavement epithelium. Here, the slightest patch of accumulating squames is only too likely to be the starting point of a vicious circle—more irritation, producing more exfoliation. Of course, our picture based on cases under treatment is unduly pessimistic. These are constantly open to attack from the outer world, and so are particularly prone to relapse. In the case of enclosed foci, as is well known, the absence of secondary infection results in a much slower and more insidious progress.

One further observation will enable us to complete our clinical picture, namely that these inflammations seem peculiarly prone to occur in certain types of mastoids. This phenomenon will be discussed later on (p. 703 *et. seq.*), when it will be shown that the normal, fully pneumatized mastoid rarely succumbs to a cholesteatoma infection. The acellular mastoid, on the other hand, seems peculiarly susceptible, a characteristic which has far reaching implications, both theoretical and practical.

To Sum Up.

(1) The writer believes that the various types of cholesteatoma are identical in origin, and that they arise from the normal inflammatory reactions of the pavement epithelium of the epi, retro and pretympanum, in certain types of mastoids.

(2) These inflammatory reactions are associated with the formation of squames and polyps.

(3) A detailed examination of these reactions enables us to predicate the existence of other clinical conditions such as acute perforating epitympanitis, acute enclosed epitympanitis and so on. Case histories exemplifying these entities are given later.

(4) When we marshal these various entities together and admit their organic unity, we realize that no standard theory gives a convincing explanation for *all of them*. On the other hand, the writer's modification of the metaplasia theory correlates them all in a simple and entirely logical manner.

On the Structure and Development of the Mastoid

Before attempting to deduce anything as to the pathogeny of cholesteatoma from the structure of the associated bone, it is essential that we should be quite clear as to the distribution and development of the various types of mastoid bone.

The natural distribution has been the subject of much investigation, although, unfortunately, ambiguous terminologies make it difficult to utilize some statistics.

Turner and Porter[23] examined a large number of skulls and elicited the following facts. They divided skulls into cellular (which included all degrees of pneumatization) and acellular (which included both ivory and diploetic types). The two classes occurred in the proportions of 80 per cent. to 20 per cent.—a ratio which closely agreed with figures previously published by Cheatle[24] and by Zuckerkandl. These figures referred to Europeans, and it was found that the incidence of acellular mastoids in other races was considerably lower. In Polynesians and Eskimos it was almost zero. Unfortunately for our purpose, they seem to have included under "pneumatized" the "mixed group" which must be regarded as a partial failure of pneumatization, and which we shall find on clinico-pathological grounds is more closely allied to the acellular type.

Leaving aside this source of ambiguity, we can still say that the fully pneumatized mastoid is much more common than the acellular.

Asymmetry was remarkably common. In at least 8 per

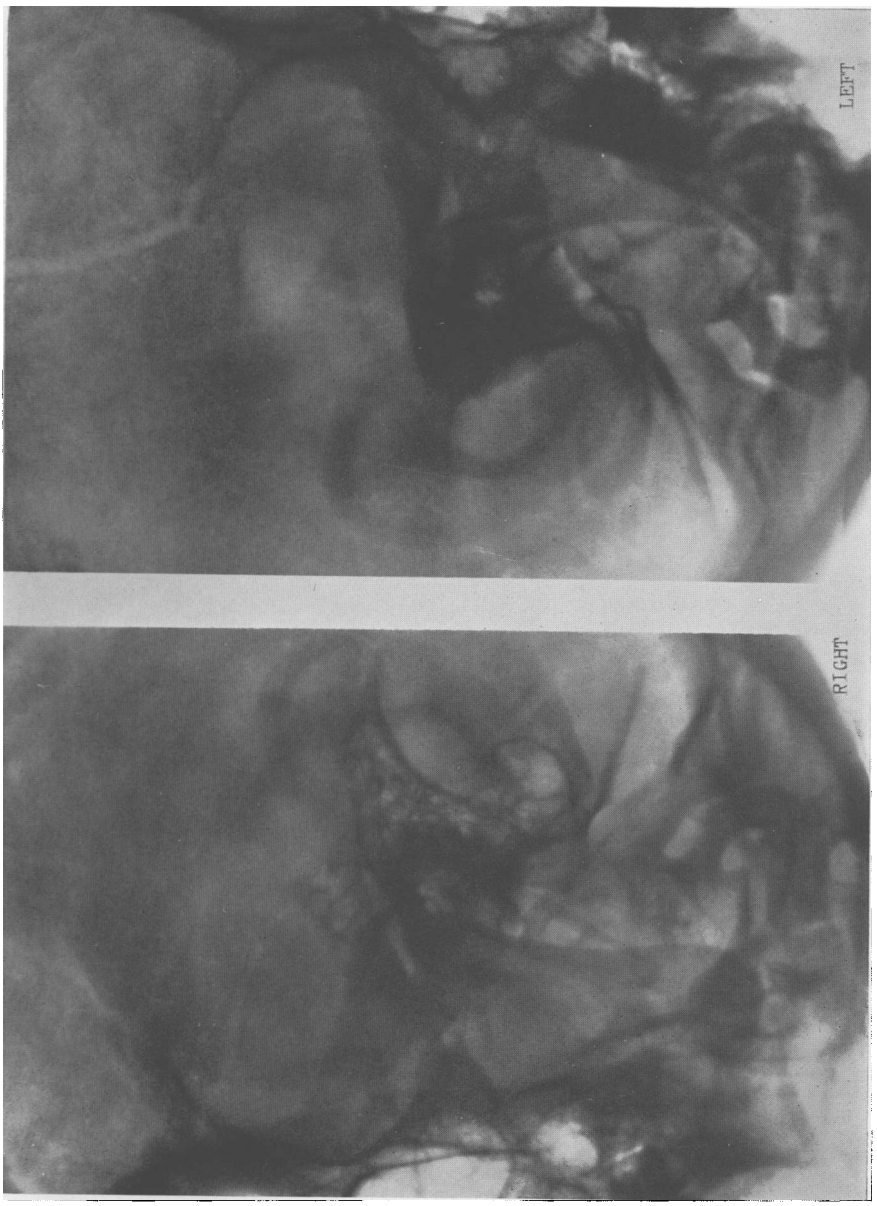

Fig. 4.

Mastoid asymmetry. Right, fully pneumatized. Left, diploetic.

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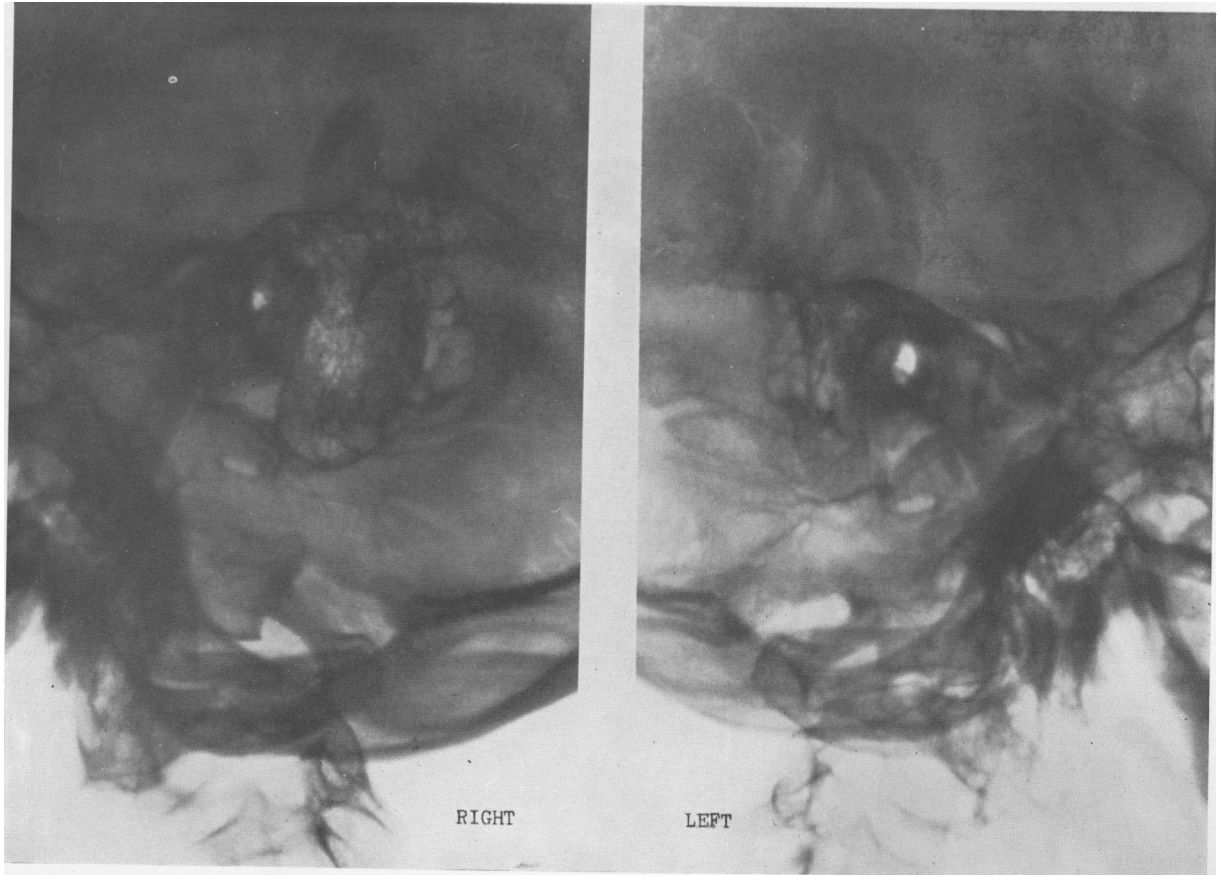

FIG. 5.
Mastoid asymmetry. Right, fully pneumatized. Left, poorly pneumatized, *i.e.* "mixed."

cent. of Europeans one mastoid is acellular, whilst the other is cellular. In a further 7 per cent. there is a marked difference between the degree of pneumatization on the two sides.

The following brief account of the development is extracted from Ruedi's classical work. In the fifth intra-uterine month, ossification begins at various points in the cartilaginous mass of the labyrinth. Somewhat earlier, membranous ossification begins in the tympanic ring and the calvaria. The labyrinth mass is invaded by the tubotympanic pouch which has grown out from the primitive pharynx. The two layers—bone and pouch—are separated widely by a considerable mass of loose embryonic connective tissue. Primary ossification proceeds rapidly, so that by the seventh month a considerable mass of diploetic bone has been laid down. By this time, the inner ear spaces, the ossicles and tympanic membrane have practically reached their adult dimensions. The rapidly expanding brain has to be accommodated, and we find the pyramid mass lengthening. In addition, however, whilst osteogenesis continues in the outer layers, the picture becomes complicated by the most intense osteoclasis in the deeper structures. This osteoclasis not only increases the overall bony dimensions, but also results in a considerable alteration in the structure of the diploetic bone. Lamellae are dissolved away, marrow spaces entered, and their highly vascular and cellular contents replaced by the embryonic connective tissue. In addition to this, a variable degree of contraction occurs in the embryonic connective tissue, so that the central pouch gradually approximates to the bony walls. In the final state it comes to line them and constitutes their muco-periosteum.

It is essential when speaking of pneumatization to realize that it consists of these two processes, namely :—

(1) The active erosion of diploetic bone to form cells of varying size.

(2) The gradual disappearance of the intervening embryonic tissue so that finally the central pouch comes to line the bone.

A true air cell cannot be formed unless these two processes occur.

According to Wittmaack[5, 6] we must regard the mucous membrane of the primitive pouch as the driving force in this metamorphosis (although it rather appears as if he includes the embryonic connective tissue as part of this membrane).

Ruedi[20] is quite emphatic that the mucous membrane plays a purely passive part. He differentiates it sharply from the intervening mesenchyne and points out that histologic evidence (and comparison with osteogenesis in other bones) all points to the bony periosteum as the determining factor. In this he is supported by Eckert Möbius.[21] Krainz has suggested that the actual atmospheric impact is of great importance, but this cannot seriously be entertained.

Let us now consider whether the various types of mastoid can be explained in terms of embryology. First, we recall the standard classification of mastoids into :—

(1) Well pneumatized.
(2) Acellular.
    (*a*) Diploetic.
    (*b*) Compact.
(3) Mixed.

(This latter group is unfortunately rather vague ; it includes all types which do not fall into the first two groups. Its main type is the poorly pneumatized one in which much of the bone remains diploetic.)

As Wittmaack has pointed out, these types are readily explicable from our embryology. If no bone absorption occurs, we are left with a purely diploetic bone. The well pneumatized or the mixed bone results according to the extent to which both processes are consummated. With regard to the ivory type, we are not so clear. Wittmaack suggests that if bone absorption is complete, but the embryonic connective tissue does not absorb, it will undergo a secondary osteogenesis and thus produce a homogeneously compact mastoid.

In evolving an explanation for these variations, Wittmaack pointed out that any given structure in the body owed its particular characteristics to two different classes of influence. Namely, (1) the " Genotypisch " (or idiotypical) factors which are nothing more than the innate characteristics of the individual—his " genes " ; (2) the phanotypical factors which are fortuitous influences from the external world.

Now it would be quite easy to say that the types of mastoid are merely different classes referable to different genes, much as we explain blue eyes and golden hair, were it not for the

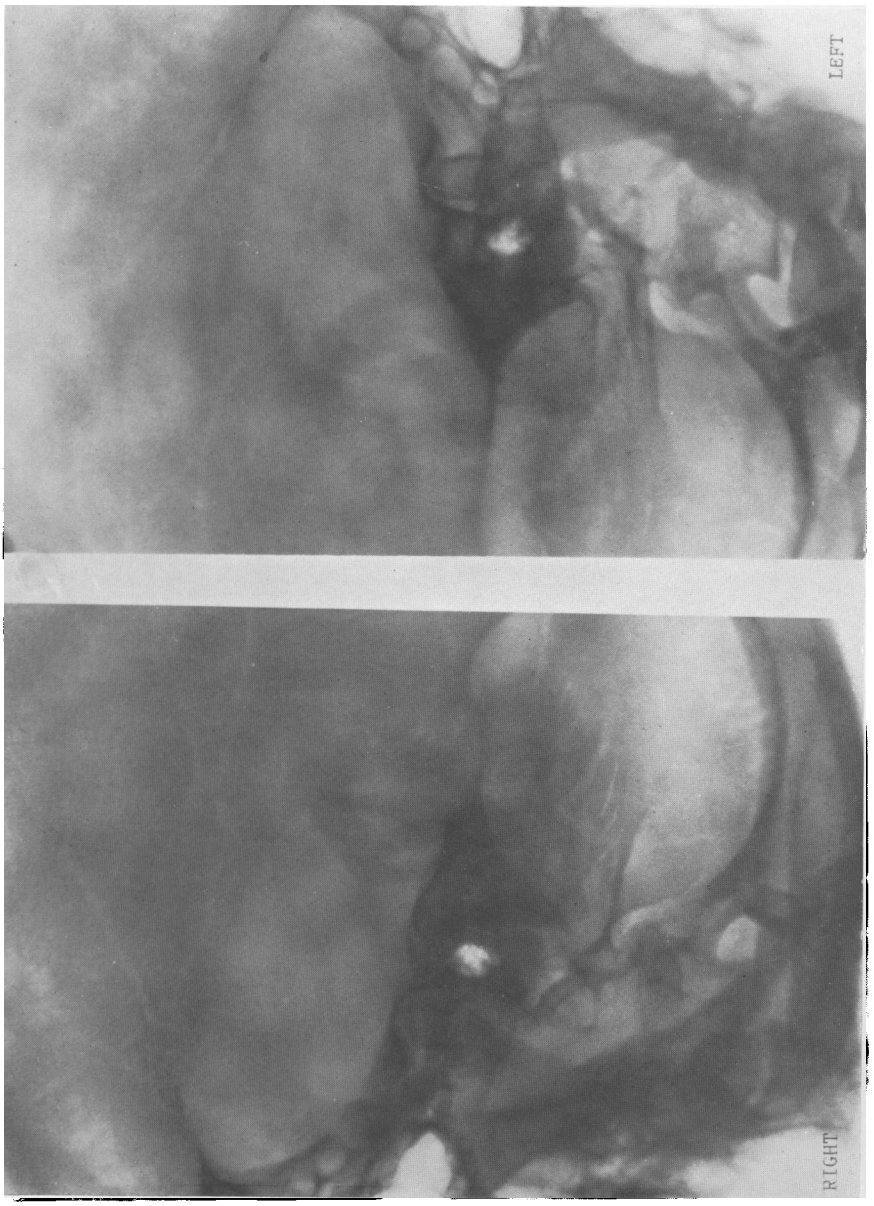

Fig. 6.
Typical small diploetic mastoids on both sides.

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fact that asymmetry of mastoids is extremely common. Something like 10 per cent. of all individuals have a well pneumatized bone on one side and an acellular bone on the other.

Wittmaack contends that the " genotypic " urge is invariably towards complete pneumatization, but that this process can be hindered by " phanotypic " forces. Such an influence was found in infective otitis media neonatorum—a condition which has indeed been shown to be amazingly common. Wittmaack, however, went farther than this, and pointed out that there was a well known and quite different entity—to wit, " aseptic otitis media " also occurring frequently in earliest life, which he described as a foreign body otitis (Fremdkorperotitis). He visualized this as arising during birth or even in utero from the entrance of meconium or amniotic fluid into the tympanum. He linked this up with his work on the structure of the mucous membrane, by saying that the infective otitis media produced the fibrous atrophic mucosa, whilst the " Fremdkorperotitis " produced the hyperplastic mucosa (see p. 690).

Albrecht, Ruedi and others have shown that whilst such otitis media is undoubtedly quite common, yet there seems to be no relationship between its intensity and the degree of pneumatization. Thus it is not unusual to see a badly pneumatized bone with a sterile middle ear, and conversely a grossly purulent exudate may be associated with well defined pneumatization. Albrecht suggested that intratympanic hæmorrhage might be responsible, but Ruedi has shown that these gross variations in pneumatization are demonstrable in the stillborn child, where naturally birth traumata have had no time to operate.

Summing up—we can agree that the general structure of the bone may be to a certain extent determined by the innate characteristics of the individual—the " pneumatizing power " of his tissues (whether periosteum—connective tissue—or mucous membrane). The fact that in many individuals the two mastoids differ widely in structure, suggests that this innate tendency may be modified by fortuitous external factors. So far, however, we have no certain knowledge as to the nature of the " phanotypisch " factors.

The above represents what may be called the Continental view of the problem, and to complete the picture we must note

an alternative hypothesis which is still widely held in England, although almost abandoned elsewhere—namely, that the various types of non-pneumatized mastoid are the end results of continued suppuration acting on a bone which was originally normally pneumatized.

This simple view, at first glance, seems thoroughly reasonable. It would explain, for instance, quite easily, why almost all radical mastoid operations involve apparently sclerosed bones. Unfortunately, it is quite untenable. A quite casual examination of a series of X-rays reveals that the acellular mastoid is a much smaller structure than the pneumatized type. This is particularly obvious in the asymmetrical cases. (Figs. 3, 4, etc.) It is impossible to believe that an inflammatory process could shrink down a large pneumatized bone to its small acellular fellow, otherwise we should surely expect that transient inflammations would only produce minor degrees of sclerosis. An examination of the clinical details shows clearly that the duration and severity of the discharge bear no relation whatsoever to the structure of the mastoid. In point of fact, in most cases the mastoid is completely acellular whether the otorrhœa has been present for a few weeks or for twenty years. It is true that the poorly pneumatized mastoid at first sight seems to provide a link. May we regard it as a fully pneumatized mastoid which under the influence of a comparatively recent inflammation is partly on its way towards complete acellularity? Here again, examination of the relevant photographs shows how untenable this is. It is impossible to see how a pneumatized bone could change over to a poorly pneumatized or acellular type. (See especially Fig. 5.)

The fact is that this chronic inflammation very rarely attacks a well pneumatized mastoid. The writer has not seen it once in a series of over fifty cases.

Unfortunately, radiologists and otologists still occasionally describe the acellular mastoid as "sclerotic" and make no distinction between it and the true post-inflammatory sclerosis. The two conditions are in no way related, and usually an X-ray can distinguish between them quite readily. W. Meyer, for instance,[38] used X-rays to study twenty-eight cases of severe acute otitis which occurred in well pneumatized mastoids, but which did not necessitate operation. He showed that in most cases there was no mastoid sclerosis four months later. In

a few cases there was a limited patchy sclerosis which, however, in no way resembled the true acellular mastoid.

It might be expected that this inflammatory theory could be finally refuted by the demonstration of acellular mastoids in patients with normal drums, normal hearing and with no history of otorrhœa. Such cases undoubtedly exist, and the protagonists of the inflammatory theory are compelled to postulate in such cases a chronic osteitis of the mastoid which has produced advanced bone changes whilst remaining absolutely symptomless.

The practical importance of this was well brought out in a discussion on mastoid asymmetry, *Proc. Roy. Soc.*, 1937, xxx, p. 423. Brayshaw Gilhespy (ibid., p. 425) said, " I am interested in this subject from a medico-legal point of view. Could the fact of an acellular mastoid be produced in a court of law as evidence of previous ear trouble in a case of injury causing deafness? If pneumatization was not present the deafness might have been present in some degree before the accident. It would depend on whether the judge was a disciple of Wittmaack, or belonged to the English school of thought."

In the writer's opinion, the otologists' duty is to report that such a mastoid can in no way be construed as *proof* of preceding ear disease. At most, it may be accepted as evidence of a predisposition towards such disease.

A further point arose at the above meeting in a discussion on a case of Ménière's syndrome, in which the affected ear was acellular, the opposite ear being normally pneumatized. On the strength of this finding, it was advised by one speaker that the affected mastoid be explored because " he thought it highly probable that, in spite of a normal drum head being found on otoscopy, a chronic infection was present in the mastoid ".

Here again, in the writer's opinion, it is quite erroneous to argue that because a mastoid is acellular therefore it must be (or must have been) the site of prolonged inflammation. To explore such a mastoid in the expectation of finding chronic osteitis simply because of the absence of pneumatization would be absolutely unjustifiable.

The problem has been approached from a different angle by Ziegler, who studied the histo-pathological appearances in mastoiditis. He stated quite emphatically that there was no

evidence whatsoever to support the theory that chronic inflammation could produce the true acellular bone.*

Armed with these conceptions and definitions, let us now turn to a problem which concerns us more closely, namely, "DOES THE STRUCTURE OF THE MASTOID THROW ANY LIGHT UPON THE PATHOGENY OF CHOLESTEATOMA?"

Suppose we are able to show that cholesteatoma was exclusively associated with a special type of mastoid, and that the same type of mastoid was the ground basis for the other conditions which I have claimed are essentially identical with cholesteatoma. We should have some presumptive evidence in favour of this identity. Let us divide our question into two as follows:—

(*a*) What sort of mastoid is associated with cholesteatoma?

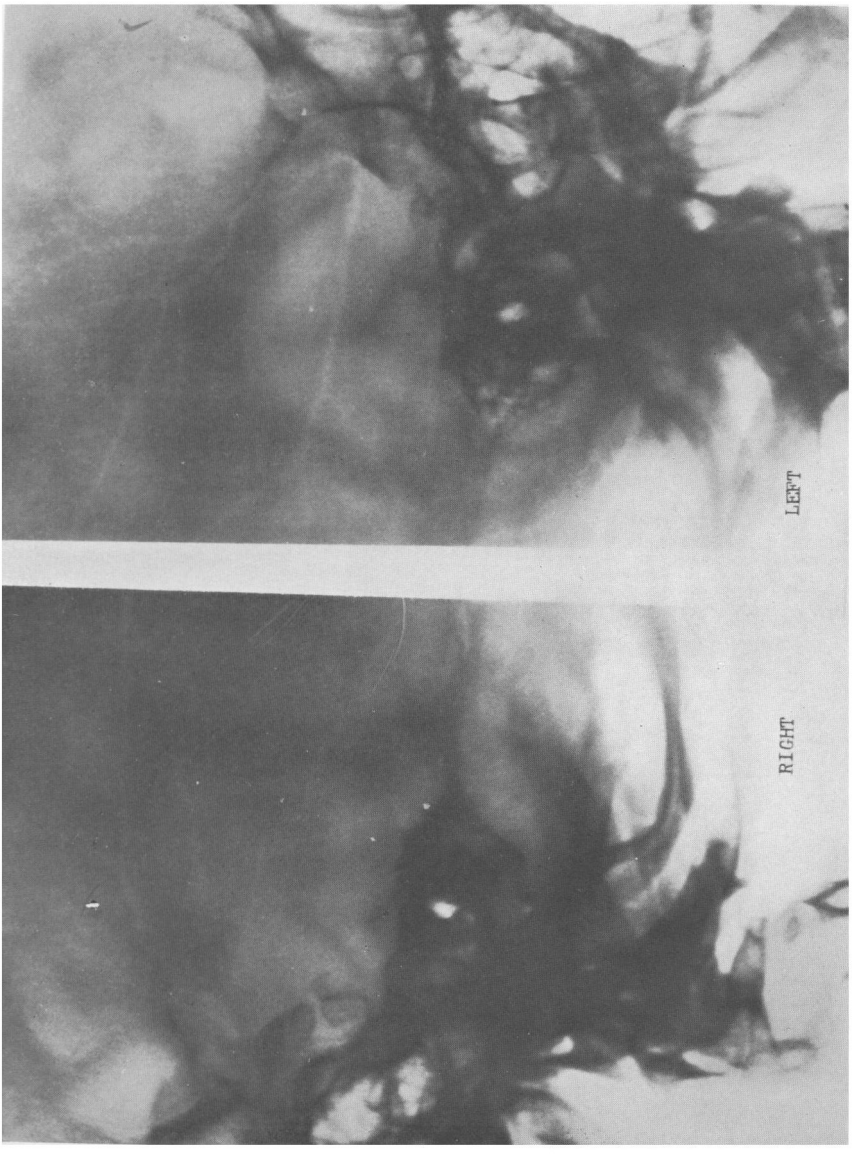
(*b*) What sort of mastoid is associated with cognate conditions such as acute epitympanitis?

Taking question (*a*) first. Wittmaack says, "For many years we have not seen cholesteatoma in a well pneumatized bone. Even if we do not find a completely compact bone, still there is a very marked reduction of air cells. . . . There are only gradual and unimportant differences between such a mastoid and the completely compact type."

This extraordinary weakness of the acellular mastoid does not seem to have received the recognition which its importance warrants. The following analysis of a small series of cases may serve to stress the point.

Out of thirty-one cases of chronic perforating epitympanitis which would merit the title of secondary cholesteatoma, not one occurred in a fully pneumatized mastoid. Four were ivory, two were "mixed" and the remaining twenty-five were diploetic. Even if we classify the "mixed" cases as pneumatized, we still have a proportion of twenty-nine acellular to two cellular, which is strikingly different from the proportion of 20 per cent. to 80 per cent. given for the general population.

* "Will man unter diesen Umstanden uberhaupt die Moglichkeit einheitlicher Sklerosierung des pneumatisierten Warzenfortsatzes durch entzundliche Vorgange offenlassen, so muss man betonen, dass sie sicher ausserst selten ist. Von mir konnte sie nicht beobachtet werden. Dass eine teilweise, gewissermassen herdformige Sklerosierung des pneumatisierten Warzenfortsatzes entsprechend dem Nebeneinander der verschiedenen Entzundungsformen moglich ist und auch gefunden wird, geht aus dem bisher Gesagten ohne weiteres hervor."



Right, mastoid ivory compact. Left, diploetic.

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(Actually, the two " mixed " cases were such as Wittmaack would classify along with the acellular.) Nor does this end the story.

Out of the twenty-nine acellular, twelve were asymmetrical, that is to say, the opposite ear was well pneumatized. The two patients with a diseased " mixed " mastoid had a " mixed " mastoid on the opposite side.

Thus, in thirty-one cases the opposite ear was :—

Pneumatized	in 12	31
Acellular	in 17	
Mixed	in 2	

Every one of the twelve pneumatized bones showed an intact tympanic membrane. It is true one gave a history of a transient otalgia, and several showed atrophic retracted or milky infiltrated drums, but in no case was there any demonstrable perforation or other gross disease.

In striking contrast, out of nineteen acellular mastoids no less than sixteen showed *gross* disease of the drum. In some cases a well established chronic perforating epitympanitis. (It is, of course, well recognized that cholesteatoma is frequently bilateral.) In others there was gross scarring with retraction, especially in the postero-superior quadrant, although frequently the patient could give no history of ear disease.

It is thus beyond question that this disease—chronic perforating epitympanitis—occurs almost exclusively in association with the acellular mastoid.

Let us now compare the findings in " acute perforating epitympanitis ". Here are details of seven cases.

Seven Cases of Acute (or Subacute) Perforating Epitympanitis

Case II. Miss V. T. Left otorrhœa three months insidious onset. Polyp pouting through pars flaccida—destroyed by bi-polar electrolysis—recurred—destroyed again. Ear now dry. N.B.—Right drum also shows a healed post.-superior perforation. No history of disease obtainable. Both mastoids diploetic—no inflammatory sclerosis. (Fig. 6.)

Case III. Miss P. Slight left otalgia two months ago with deafness and tinnitus. Ear syringed by family doctor. Two days

later otorrhœa commenced. On examination typical polyp in attic. Left mastoid diploetic. Right normal cellular. Right drum shows no gross disease.

CASE IV. Mrs. M. Right ear slightly deaf many years. Hearing became definitely worse with tinnitus four weeks ago. No pain. Scanty discharge. Post.-superior perforation and tiny polyp. Ivory mastoid. N.B.—Left ear has been deaf many years. "Never ran." Posterior perforation. Ivory mastoids both sides.

CASE V. Miss I. T. Left ear gave no trouble until ten weeks ago. Painless onset of deafness and tinnitus and scanty discharge. Post.-superior perforation—"mixed" pneumatic diploetic mastoid. Right drum badly cicatrized. No otorrhœa. Mastoid "mixed". (Fig. 7.)

CASE VI. Mrs. W. *Right otalgia (slight) four weeks ago.* Ear gradually began to "feel full". On examination a blackish mass of wax and debris removed, revealing a tiny posterior perforation. One week later ear dry but hearing remains "cloudy". Pars flaccida remains congested. Mastoids diploetic. (Fig. 8.)

CASE VII. Mrs. B. Attended because of "a boil in the ear" and recent deafness. Actually there was a furuncle, but after this was cured a small perforation in the pars flaccida was found to be discharging a little pus. The condition healed under treatment. Diploetic mastoids.

CASE VIIA. Mr. J. B. H. Slight right otalgia with tinnitus and scanty otorrhœa one week. Small post.-superior polyp. Healed under treatment. "Mixed" pneumatic diploetic mastoids. Left drum seems normal.

It is apparent that this condition shows a predilection for the acellular mastoid just as cholesteatoma does. Not one case occurred in a fully pneumatized bone. Four were diploetic, one ivory, and the remaining two were "mixed".

Admittedly this is no proof of the identity of cholesteatoma and acute perforating epitympanitis, nevertheless it seems reasonable to argue that the latter is merely the preliminary stage of the former.

Let us for comparison see how this group can be explained by the other theories. No one seeing a polyp bulging through the pars flaccida could seriously entertain Wittmaack's suggestion of a preliminary indrawing of Shrapnell's membrane. Nor is it possible to see how a congenital condition could

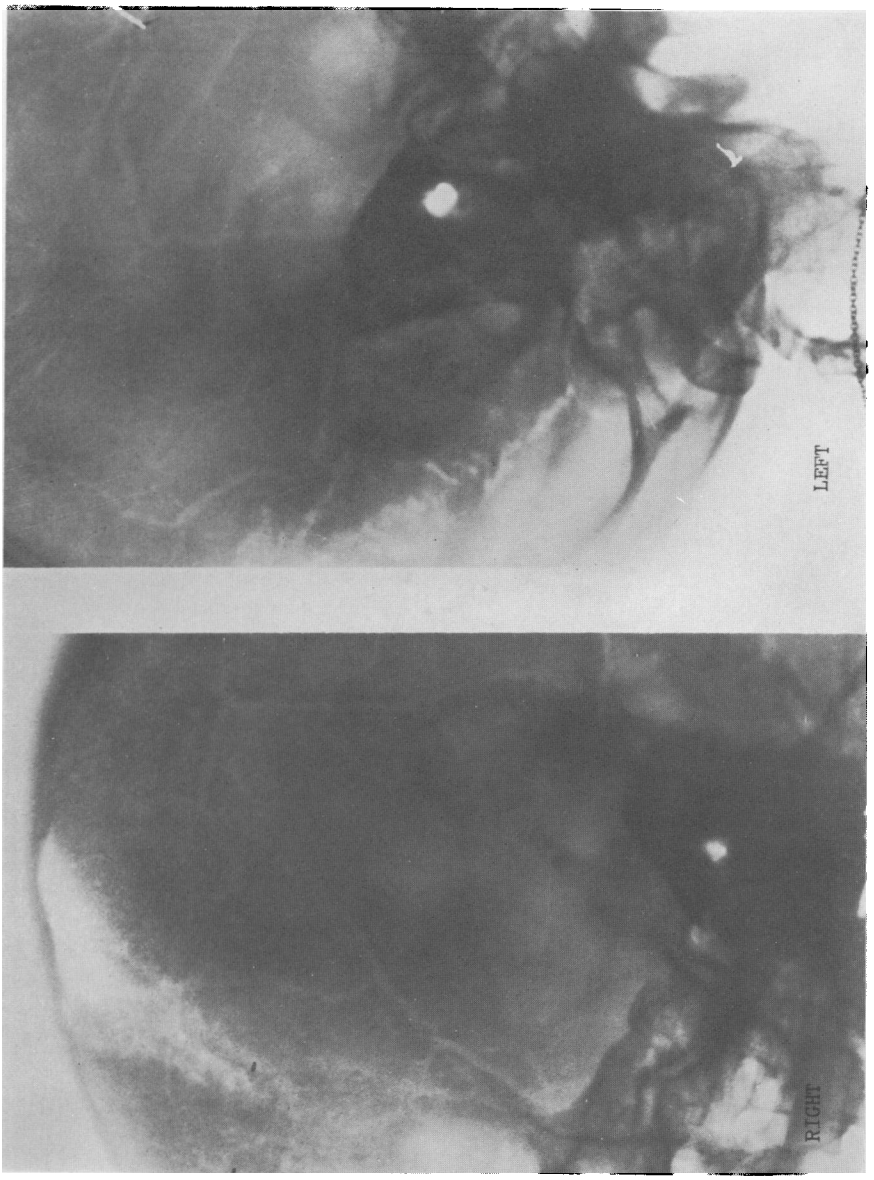

Fig. 8.

Right, mastoid dense inflammatory sclerosis. Left, diploetic.

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produce these manifestations. Certainly Cases VI and VII can only be regarded as transient inflammations. What of the " immigration hypotheses " ? Do these cases supply any confirmatory evidence ? Obviously not. To begin with, Cases VI, VII and VIIA would have to be rejected as non-cholesteatomatous in view of the fact that they healed. And yet we cannot help feeling that they are merely mild attacks of a condition which in more severe cases produces cholesteatoma. As usual, they have acellular mastoids. Cases II, III, IV and V would be admitted as candidates for the title. But Case II has closed over and healed. Must we relegate it along with VI, VII and VIIA ?

The question, " Are they or are they not cholesteatoma ? " reveals the underlying weakness in the whole immigration theory. Nothing but the demonstration of undoubted epidermic tissue in the attic could clinch that point. But I have already shown that such evidence has never been provided. Nor would we be any better off if these cases gave a history of a year or ten years. At no stage is it necessary (or helpful even) to introduce " epidermic immigration " in order to explain the persistent suppuration. The chronic irritation of retained squames and polyps is quite sufficient to explain the indefinite persistence of the condition.

This failure of standard theories is in striking contrast with the modified metaplasia theory, which supplies a framework into which every one of these clinical entities dovetails in the most harmonious manner. The theory, so to speak, prophesies that such conditions must exist, and conversely their discovery confirms the theory.*

(To be continued)

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* It would be instructive to apply a similar argument to other cognate inflammations. Unfortunately they are not seen often enough to justify statistical deductions. In two recent cases of enclosed epitympanitis, the four mastoids were diploetic. In Patterson and Smelley's six cases of chronic enclosed pre-tympanitis, the mastoid was pneumatic in two, acellular in two and not described in two. Unfortunately these groups are not big enough to give us any statistical confirmation of the theory. It is, however, interesting to note that whilst in accordance with the embryonic theory, the petrous cholesteatoma would be quite frequent, the metaplasia theory declares it must be very rare. Clearly if a mastoid is acellular it is very unlikely that the petrous will be pneumatized, and consequently there will be no mucous membrane to act as the originating focus for cholesteatoma.

A. Tumarkin

Der Verfasser glaubt, dass die verschiedenen Typen des Cholesteatoms auf die gleiche Ursache zurückgehen und dass sie in einzelnen Formen des Warzenfortsatzes aus den normalen entzündlichen Reaktionen des Schleimhautnelages im Epi-, Retro- und Prae-Tympanalraum entstehen. Diese entzündlichen Reaktionen gehen mit der Bildung von Schollen und Polypen einher. Die Annahme des Verfassers ist eine modifizierte Metaplasietheorie, die nach seiner Ansicht die verschiedenen Formen in einfacher und logischer Weise zusammenfasst.

. . . .

L'auteur croit que les différentes formes de cholestéatome ont une même origine, et qu'elles dérivent des réctions inflammatoires normales de l'épithélium pavimenteux de la région épi, retro et prétympanique dans certaines formes de mastoïdes. Ces réactions inflammatoires sont associées avec la formation de squames et de polypes. L'auteur propose une modification dans la théorie de la métaplasie qui groupe les diverses entités d'une manière simple et logique.

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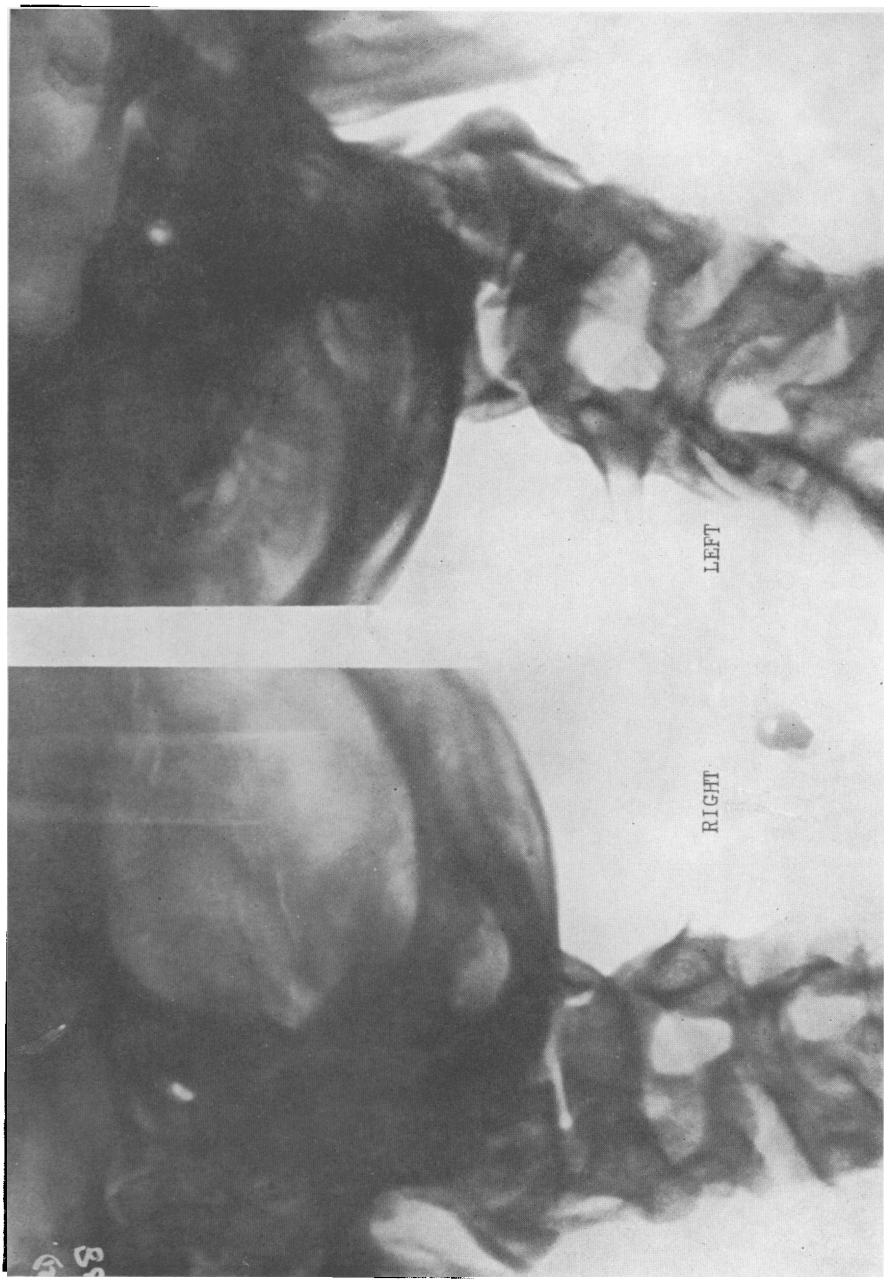

Fig. 9.
Both sides compact ivory.

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