CORRESPONDENCE

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DAMAGED EAR CARTILAGE AND TYPE OF LEPROSY

A question was submitted to a number of experienced clinical leprologists essentially as follows:

When, in a patient in whom the disease has cleared up, the edges of the cartilages of the external ear are found to be more or less affected ("nibbled," or "rat-eaten"), what form of the disease would you say had been responsible for that condition? In other words, what would be your retrospective type diagnosis with respect to the condition?

In connection with this question it was pointed out that the condition referred to is not common, for ordinarily in a case in which there had been marked lepromatous infiltration the lobes and helices are left lax and the cartilage undamaged after subsidence, and that the condition probably results only from severe reactions with ulceration. It was also mentioned that destruction of the cartilage of the nose in tuberculoid leprosy was not known, so by implication the question meant whether damage to the ear cartilage may occur in that type.

The replies, of interest if for no other reason than because of the differences of experience of the writers and the diversity of their views, are here arranged more or less in regional groups, beginning with the Philippines where the question arose. This symposium is accompanied by a plate showing illustrative examples.

From Dr. C. B. Lara, Culion, Philippines: These past few years the active process of "nibbling" of the ears has not been seen, perhaps because effective anti-leprosy treatment has reduced the predisposition. One is therefore obliged to resort to questioning patients who show such changes, and consulting their previous records. This has been done for nine from such cases among the number here.

From this investigation I gather that the condition is usually bilateral although not necessarily symmetrical, but that sometimes it is unilateral. Practically always it occurs in the course of, or a result of, repeated subacute reactions involving old lesions, with or without associated features of the erythema nodosum-type of lepra reaction; but exceptionally there may be no associated general reaction. Tissue breakdown in the reacting lesions of the ears and consequent ulceration together give the resultant nibbled appearance. Some cases also have sinking of the bridge of the nose, while others do not. Since in all of the cases the condition had been associated with ulceration, the role of secondary pyogenic pathogens must be considered, in addition to the peculiar state of allergy and type of disease. Another factor may be the state of the blood circulation in the tissue affected.

Cases of typical lepromatous leprosy, with history of repeated reactions of the erythema nodosum type but without ulceration, do not show such damage of the ears. On the other hand, most if not all of the cases I have investigated have mani-

fested strongly suggestive or definite features of tuberculoid leprosy of reactive nature. In fact, one of these cases, under observation since the onset of leprosy, was found to be tuberculoid on biopsy, gave a 2+ lepromin reaction, and was essentially bacteriologically negative some three years before the process of nibbling took place. In short, my impression is that nibbling of the ears occurs mostly in patients subject to repeated ulcerative reactions who are suffering from the borderline (or transitional, tuberculoid-to-lepromatous) form of leprosy.

From Dr. José G. Tolentino, Cebu, Philippines: The question raised is an interesting one, which nobody has thought of investigating before. The condition is not common, and instead of giving an opinion based on past experience I have gathered ten cases of nibbling of the ears here, in the Eversley Childs Sanitarium, and I believe my findings are worth passing on.

All these ten cases on admission were bacteriologically positive and hence were classified as lepromatous: 1 as L1, 3 as L2, and 6 as L3. The dates of admission were: 2 in 1931, 2 in 1934, 1 in 1939, 1 in 1949, 2 in 1950, 1 in 1951, and 1 in 1954. One of the 1931 cases was paroled in 1939 and readmitted in 1949; another one has been bacteriologically negative once but is positive again; a third one is now on the negative list awaiting parole.

On admission their lesions were described variously as: (1) nodular infiltration in patches, with numerous flat rice-grain-sized nodules; (2) thin, hyperemic, diffuse infiltrations with tiny nodules; (3) flushing of the skin, with moderate reddish patches of infiltration; (4) reddish-brown areas and moderately thick leprotic infiltrations; (5) advanced infiltrations with elevated papulomacular areas and soft nodules; (6) patches of red infiltration; (7) chocolate-colored papules and macules with pinkish borders; (8) brownish patches and marked infiltrations; (9) pinkish macules, reddish patches, and reddish elevated areas; and (10) pinkish papules and thick red macules.

All of these cases have had lepra reactions with ulcerative lesions described as plaque-like. These reactions produced nibbling of the ears (in one case also of the nostrils, giving a gangosa-like appearance), and multiple stellate, keloid-like scars on various parts of the body. In these reactions the ears were involved in all 10 cases, the face in 8, the extremities in 9, the buttocks in 4, and the trunk in 3.

At present one or both ears present the "nibbled" appearance. There is alopecia of the eyebrows in 9 cases, sunken nose in 4, atrophies in 4, contractures in 4, absorption of digits in 3, blindness in one eye in 2, perforated septum in 1, and plantar ulcer in 1. There are no enlarged, elongated earlobes. All have recently been tested with lepromin, 7 giving negative reactions, 2 doubtful, and 1 weakly positive.

With our present understanding of borderline leprosy, the classification of these cases as lepromatous may be seriously questioned. The patchy character of the skin lesions many of which were described as reddish, the ulcerative nature of the reactional lesions described as plaque-like, and the absence of enlarged or elongated earlobes, are all features not characteristic of typical lepromatous leprosy. The ulcerative reactional plaque-like lesions seem very similar to the ulcerative reaction described by Ryrie in tuberculoid lesions, and are not found in the lepromatous form.

The skin lesions in lepromatous lepra reaction sometimes have the form of bullous erythema nodosum leprosum and leave temporary scars that eventually disappear, but they do not produce the permanent, keloid-like scars of the ulcerative plaques found in these cases. My suspicion that these cases were borderline is based solely on clinical morphology of the lesions and their behavior.

From Dr. José N. Rodriguez, Manila, Philippines: I have observed the "rat-eaten" effect of the edges of the cartilages of the external ear in lepromatous leprosy following acute lepra reaction, ending with bullous and finally pustular lesions. In exceptionally severe cases with such lesions of the ears, the other edges of the cartilage may be destroyed.

I have not seen a similar lesion in the tuberculoid type. However, it seems possible that in severe, ulcerating cases of reactional conditions in tuberculoid cases this effect may result if lesions involve the ear. In such cases I would expect to find marked enlargement of the auricular nerve on the corresponding side.

From Dr. Yoshinobu Hayashi, Tokyo, Japan: In advanced lepromatous leprosy the external ear may frequently be affected by marked leprous infiltration, as other parts of the body are affected by nodules and infiltrations, and there may occur ulceration to the extent of exposing the underlying cartilages. When it happens that such cases clear up completely, we can find thereafter deformity or reduction of the cartilages of the external ear. Thus we recognize that reduction of the external ear may occasionally remain in the lepromatous type of leprosy. Moreover, it may happen that the cartilage of the nasal septum or of the epiglottis is affected in lepromatous leprosy. We have no such experience in the tuberculoid type.

I therefore think that the type of the disease in cases with the condition in question would be lepromatous. In such cases, presumably, there will be some other condition also indicating the diagnosis of the lepromatous type.

From Dr. Tokuzo Yokota, Okayama, Japan: Replying to the question addressed to Dr. Mitsuda, I may say that, in general, the invasion of leprosy bacilli into cartilage is rare and slight, and ulceration of the ear occurs only in an occasional case. It may result from injury, frostbite, or other things.

The meaning of "reaction" in the question is not understood. If it means erythema nodosum leprosum, the condition referred to may occur. In that case the type of disease would be lepromatous. In the reactional phases of tuberculoid and neural leprosy, for instance *akuter Schub*, the condition does not occur.

From Dr. Taiji Nojima, Takamatsu, Japan: [This reply is a two-part contribution.] 1. The question is very difficult, and I could not give a precise reply without seeing the patient. Many years ago I sometimes heard about patients with the "rat-eaten" condition of the ears, but not recently. [There follows speculation about what bacteriological examinations and lepromin tests might show at the time the actual lesions existed.] There will be in the near future a conference of the leprologists of Western Japan at which classification is to be discussed, and I will bring this question up there.

2. The opinion of the meeting, which was held in the latter part of September, was that the question is very interesting, but: (1) It would be very difficult to decide on the diagnosis without seeing the patient. (2) The condition may have been lepromatous, but if so some lepromatous symptoms should be found elsewhere.

From Dr. Dharmendra, Calcutta, India: Regarding the question of retrospective type diagnosis in a patient in whom the disease has cleared up and the edges of ear cartilages are found to be "nibbled," or "rat-eaten," our experience here is in agreement with the views indicated in the letter of inquiry about the affection of the cartilages of the nasal septum and the external ears in the tuberculoid and lepromatous forms of the disease.

In lepromatous cases the ear lobule sometimes shows shrinking, instead of being pendulous, and after repeated ulcerations followed by healing and cicatrization the margin may show the appearance described. Because of the hard cicatricial tissue it may give the impression that the cartilage has been involved, but only rarely is that structure actually affected. In the occasional case in which the cartilage has been damaged, my retrospective type diagnosis would be lepromatous.

From Dr. E. Muir, London, England: I cannot recollect having seen the "nibbled" condition of the cartilages of the external ears. It is only the soft tissues of the ears that, in my experience, are affected in lepromatous cases.

I have never seen the septum of the nose affected except in lepromatous cases. Apart from this, I am unable to contribute to the symposium, but if other workers have seen the condition asked about I think it might be a very interesting matter to ventilate.

From Dr. R. G. Cochrane, London, England: I believe that the condition referred to is rare. I recall having seen one case in which both ears were almost completely destroyed, and—this being the important part of the question asked—the cartilaginous part of the ear was eaten away. That case, seen at Chingleput some 20 years ago, was a borderline one with a severe reaction, with ulcers on the face and elsewhere. If I recollect correctly the right side of the face and the right ear were more severely affected than the left; that is, the condition was asymmetrical.

While this type of deformity is unusual, I believe it occurs in the borderline or dimorphous case. I think that an inquiry about the destructive processes which take place in such cases and the deformities and crippling damage which result would open up a subject of great interest. It is my opinion that the borderline group, particularly the ulcerating reactive form, is the most crippling of all the forms of leprosy.

From Dr. T. F. Davey, Uzuakoli, Nigeria: The "nibbled" ear condition exists in Nigeria, but it is decidedly rare. I have encountered it (apart from an occasional patient who had produced it himself in misguided attempts at self-treatment) only in lepromatous cases in which the infection was advanced when they first sought treatment.

Even in this group involvement of the ear cartilage is exceptional, and when seen it has been regarded as secondary to ulceration occurring during severe lepra reaction, especially in debilitated patients. Superficial ulceration of nodules on the ear is not uncommon, but with normal care such ulcers heal with a minimum of scarring. Where such patients give a history of similar incidents at home in conditions of neglect and malnutrition, it is not surprising when they exhibit the stigmata of past severe ulceration.

From Dr. J. Ross Innes, Busia, Uganda: Reduction of the edges of the cartilages of the external ears as a result of erosion occurring during reaction will, in my opinion, always be due to antecedent reactional conditions in tuberculoid leprosy, and I would have no tendency to attribute it to lepromatous leprosy.

There is a possibility of confusion in African cases about such "nibbling" of the cartilage of the external ear in that yaws is prevalent, both untreated and partially treated, and we should keep a sharp eye out for the signs of yaws in such cases as a complicating or purely causal factor.

From Dr. R. Chaussinand, Paris, France: I believe that the partial destruction of the cartilage of the ears does not possibly enable one to make a retrospective diagnosis as regards the form of the disease. Generally, it involves cases with ulcerating lepromatous leprosy, but I have observed such lesions in Indo-China in patients unquestionably of tuberculoid type but of the lazarine variety.

From Dr. Felix Contreras, Madrid, Spain: Although I have not made any particular study of the matter, which will be done in the future, I believe that damage of the ear cartilage is not caused by specific granulomatous invasion. It usually occurs as a feature of trophoneurotic leprosy, and less frequently it is caused by associated pyogenic infections.

In a rapid survey of the patients now in Fontilles, I have found several healed ("burnt out") cases with loss of substance of the ear cartilage. Almost all of them have some other stigmata of trophic character, such as deformity of nose due to destruction of the cartilage, ocular changes or deformities of the same character, alopecia of the eyebrows, mutilations, etc.

I believe that the great majority of these patients were lepromatous, in its neural variety. However, I also believe it possible that neural tuberculoid leprosy can produce the same stigmata, although of less severity. Most of the patients in Fontilles are lepromatous, and we have very few of the tuberculoid type.

The genuine lepromatous cases, the cutaneous kind in which infiltrative lesions predominate, usually do not have any of the stigmata mentioned. Many patients of this variety have lepromatous infiltrations of the ears, which are often markedly enlarged, and many have repeated lepra reactions; yet when the infection subsides there usually remains only atrophy of the skin without any affection of the cartilage. It is not to be denied that such a condition may sometimes occur as a result of secondary infection.

For the present, in answer to the question, I believe that in a case that has been cleared of active manifestations the retrospective type diagnosis should not be based on any single feature, but on the totality of the stigmata and cicatrices. I think that it would be very unusual to find only the nibbled or "rat-gnawed" ear cartilage, or any other single stigma of whatever location. Almost always we find various things which help in making the retrospective diagnosis, and when changes of trophic character predominate, especially affections of the cartilage and bones, I believe we can be sure that the patients were of the neural subvariety, and that the majority of them were of the lepromatous type (neural lepromatous leprosy).

From Dr. Rolla R. Wolcott, Carville, Louisiana: The question is an interesting one. I have seen the described type of ear in a few patients here and in other leprosaria. Thin, atrophic skin is stretched rather tightly over cartilage which has been partially eroded, resulting in irregular serrations as shown in the accompanying pictures [see plate, Figs. 7 and 8]. The condition has been limited to patients of the older age group, or at least those who have had leprosy for many years. The clinical histories and the amount of scarring of the skin indicate that the patients had undergone severe reactions and extensive ulceration of the skin. All of them have lepromatous leprosy, which is generally quiescent from the clinical point of view but not necessarily negative on bacteriological examination.

I have not seen any of these moth-eaten ears develop here in the last ten years. They seem to be associated with severe reactions of the presulfone era. I would hazard a guess that the reactions were characterized by ulceration and necrosis of lepromatous tissue, and that they were not of the erythema nodosum variety. It is my opinion that the erosion of ear cartilage is caused by the ulceration and secondary infection rather than by leprosy involvement *per se*.

I have seen infiltration of an ear (usually unilateral) during tuberculoid reactions, but I do not believe that tuberculoid leprosy has ever caused the "rat-eaten" erosion of cartilage.

It may be of interest to note that in the section on the ear of Klingmüller's book (p. 439) there is no mention of the "rat-eaten" condition. About affection of cartilage in general, he regarded it as debatable whether that structure is involved *per se* or whether the infection comes from the perichondrium or adjacent structures. He noted that Neisser had pointed out that cartilage can be affected by leprosy since he had demonstrated bacillary globi near the nuclei of cartilage cells, but that Babes was of the opinion that cartilages (e. g., of the nose, ear, eyelid, larynx) are seldom involved even though cartilage cells have shown large clumps of bacilli. Other views cited are in essential agreement. For instance, Sugai believed that the regressive changes seen in cartilage may be secondary to basic processes in the perichondrium.

From Dr. E. K. Chung-Hoon, Honolulu, Hawaii: My reply to the question concerning affections of the external ear in leprosy has been delayed because I decided to take some photographs to illustrate my impression of the condition described [see plate, Figs. 9 and 10].

The question is: the retrospective type diagnosis of a patient in whom the disease has cleared up and the edges of the cartilages of the external ears are found to have been more or less reduced. My answer is: lepromatous leprosy.

I do not recall having seen this kind of ear deformity in tuberculoid leprosy. The patients of whom photographs are supplied were all heavy, advanced lepromatous cases of many years duration, who had had ulcerations of the lepromatous of the ears and of the face and extremities for varying periods of time. The ultimate reduction in the size of the external ears and the nibbled appearance of the remaining cartilages are the residuals that the patients now exhibit. My impression is that this deformity is a trophic one resulting from interference with the blood supply to the cartilage by the lepromatous infiltration and secondary infection.

From Dr. V. Pardo-Castelló, Havana, Cuba: The inquiry about cases of leprosy in which the edges of the cartilages appear nibbled or serrated is very pertinent. I agree that the point is a minor one, but believe that it deserves to be cleared up.

The cartilages of the ears, as well as of the nose, do not have much irrigation. They are composed of a very thin layer of fibro-cartilage, a very lax connective tissue covering, and, finally, very thin skin. Any injury or inflammation easily breaks the cartilages or establishes a chronic inflammation which results in the well-known "prize-fighter's ear" (cauliflower ear). As long as there is no infection the cartilage usually suffers no necrosis, but any infection often results in perforation of the concha; and if it is on the edge of the ear, small areas of necrosis would result in the serrated or nibbled aspect referred to.

I should say that this condition in a case of leprosy occurs in the lepromatous type in which small nodules on the edge of the ear undergo necrosis, either spontaneously or through trauma. I do not believe that such a condition could occur in cases of tuberculoid leprosy.

From Dr. Jacinto Convit, Caracas, Venezuela: We have observed the "nibbled" condition of the ear only in lepromatous cases, as cicatricial lesions remaining after secondary pyogenic infection of ulcerations incident to lepra reaction. I agree that the cases which show the condition are not numerous, but I cannot say that they are rare.

As regards the retrospective diagnosis in cleared-up cases, I believe that as a stigma the condition has diagnostic value, if the possibility of other affections that might produce similar conditions can be eliminated. For instance, dermal leishmaniasis americana, which occurs here in Venezuela in many leprosy foci, may localize in the external ear and produce sequelae of similar appearance.

From Dr. Lauro de Souza Lima, São Paulo, Brazil: I have not observed the condition of the ears described, perhaps because I have not given it due attention. It is really a matter of minor importance, but everything in leprosy is interesting.

From Dr. G. Basombrio, Buenos Aires, Argentina: I have never observed the "nibbled" ear condition in improved or cured cases. I have, however, seen destruction of the edge of the ear cartilage by erythematous lupus and such a condition could be confusing if the patient also had leprosy, or were to develop it later. [This reply was accompanied by photographs showing the two asymmetrically affected ears of a patient with ulceration lupus erythematosus.]

From Dr. Salomon Schujman, Rosario, Argentina: I have seen a few cases with partial destruction of the cartilage of the ear, and remember that they looked as if bitten or gnawed by rats. This has been only in lepromatous cases of long duration and who, during the course of the disease, had suffered frequent, intense and prolonged lepra reactions.

From Dr. Héctor Fiol, Buenos Aires, Argentina: Destructive lesions of the borders of the ears are extremely rare, and occur only in severe reactional conditions with ulcerative or suppurative lesions. There is a question whether the destruction of the cartilage is due to the leprous infiltration itself, or to secondary infection, or to some other, concomitant affection. In the progressive phase of the disease the question is easily answered, but that is not so when one is confronted only by residual sequelae.

In this connection I may mention a nonleprous patient now under my care who has urate tophi in different stages of evolution, some of which are abscessed while others are cicatrized, and in the process the edges of both ears have been "nibbled." (Abscessed tophi are also present elsewhere.) Now, if this patient had leprosy also, with infiltrations of the ears in which such urate deposits had formed, one not aware of the facts might easily be led to a wrong diagnosis in a routine examination.

To return to the question asked, I believe that, given that the destructive infiltration in question was leprous, the retrospective diagnosis of the type based solely on that condition is not possible; other clarifying factors must be taken into account. In my opinion the ulcerations may occur in violent reactional conditions, either tuberculoid (borderline?) or lepromatous, although they rarely involve the cartilage and I have never seen the condition in tuberculoid cases.

DESCRIPTION OF PLATE

PLATE 11

FIG. 1. Extensive "nibbling" or serration of the cartilage, of moderate degree, over almost the entire helix, with loss of soft tissue. The nasal cartilage in this case is intact. Case R. E., Culion, duration of disease 20 years. (This was the case which led to the present inquiry. See also Fig. 5.)

FIG. 2. More marked but less extensive loss of cartilage substance in a frankly lepromatous case, with sunken nose, almost complete loss of eyebrows, and numerous cutaneo-subcutaneous nodules of the forearms and hands, with deformities of the fingers. Indicative of a previous phase of the disease, however, there are extensive parchment-scarred areas on the buttocks and lower back that can be ascribed only to ulcerating tuberculoid lesions that had existed long before. Case F. M., Culion, duration of disease 30 years (with a 6-year parole interval).

FIG. 3. Fairly marked but irregular damage of the ears in an old, clinically inactive lepromatous case, with concurrent loss of the nasal cartilage. Case B. A., Culion, duration of disease 26 years.

FIG. 4. Marked destruction of the ear, also in an old, clinically inactive case. Nasal cartilage intact. Case F. E., Culion, duration of the disease 26 years. (See also Fig. 6.)

FIG. 5. Scars of previous ulcerative reaction lesions on the antero-lateral surface of the left arm, and on shoulder, Case R. E. (Fig. 1). These scars, there and elsewhere, constitute another of the sequelae which, besides the ear lesions, would be taken into consideration in making a definitive type diagnosis of the case as a whole.

FIG. 6. Scars on the latero-posterior aspect of the arm, Case F. E. (Fig. 4).

FIG. 7. Marked destruction of the ears, and asymmetrical, gangosa-like damage to the nose, in a case of lepromatous leprosy. Case No. 1220, Carville. (Photograph supplied by Dr. R. R. Wolcott.)

FIG. 8. Moderate degree of affection of the ear, comparable to but less extensive than that in Fig. 1, in a case of diffuse lepromatous leprosy with the Lucio phenomenon. Case No. 2289, Carville. (Photograph supplied by Dr. R. R. Wolcott.)

FIG. 9. Fairly marked damage to the ear, and loss of nasal septum, in a case of retrogressed lepromatous leprosy. Case S. S., Honolulu. (Reproduced from a Kodachrome film supplied by Dr. E. K. Chung-Hoon.)

FIG. 10. Irregular loss of cartilage substance in a case of retrogressed lepromatous leprosy; close-up view. Case M. C., Honolulu. (Reproduced from a Kodachrome film supplied by Dr. E. K. Chung-Hoon.)

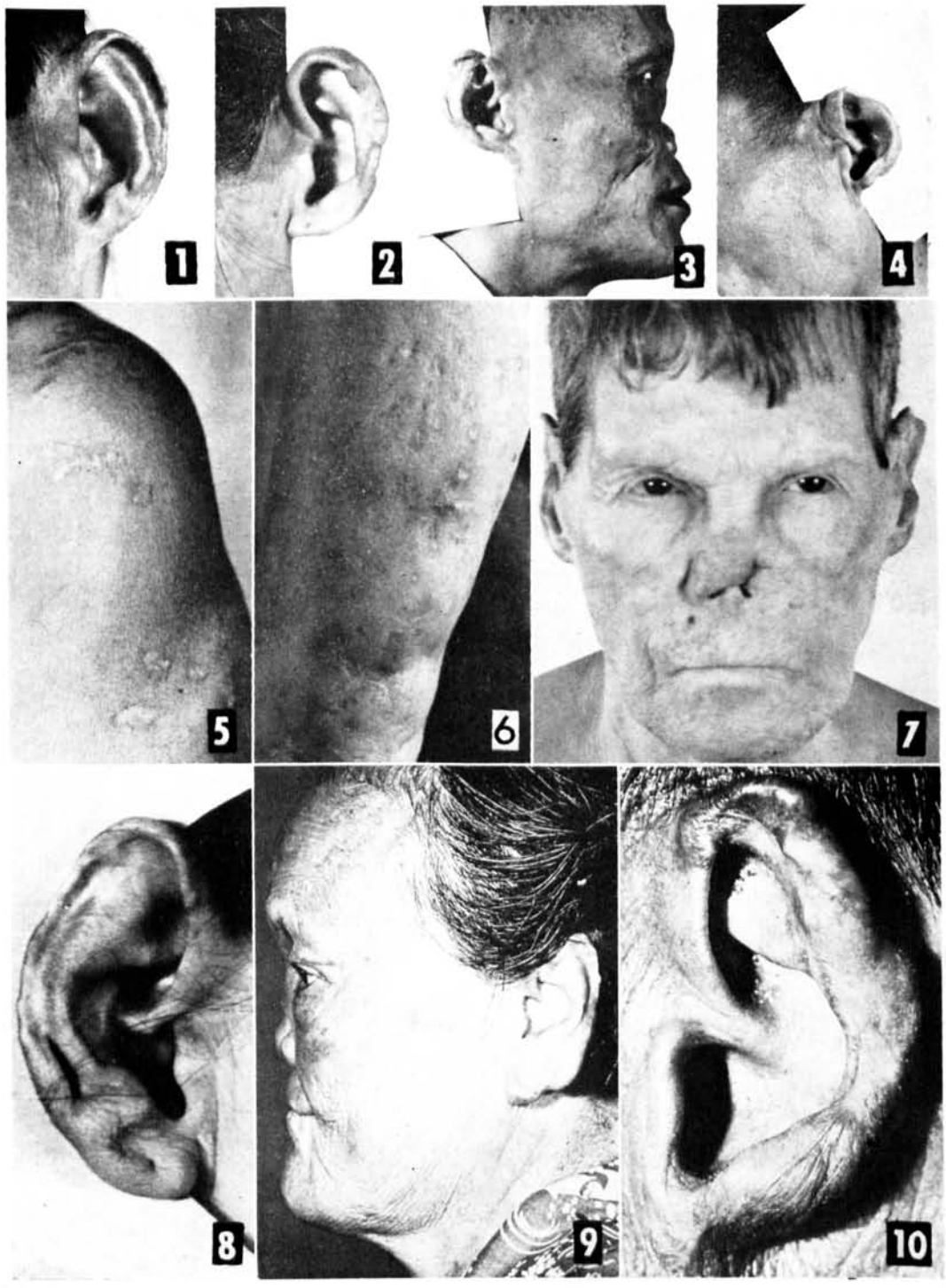

PLATE 11