

Redundant and Wrinkled Facial Skin in Lepromatous Leprosy A Correlation of Clinical and Histopathological Findings¹

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Redundant and wrinkled facial skin is a common finding in patients with lepromatous leprosy of long standing. The disease has a predilection to become more infiltrated in selected sites of facial skin, namely, in the earlobes, forehead, eyelids, maxillary and nasolabial-mental areas (Fig. 1). When these changes occur in younger patients they are responsible for the appearance of premature aging. Because of the frequency of redundant and wrinkled skin in patients with leprosy, a causal relationship has been presumed; however, the specific role of lepromatous disease in the development of these changes has not been determined nor differentiated from other possible etiological factors. Furthermore, the proneness to recurrence of redundant skin following surgery questions the advisability of routinely recommending corrective procedures for this problem. It is with these thoughts that a histopathologic study of collagen and elastic tissue in facial skin of patients with leprosy was undertaken.

MATERIALS AND METHODS

In the course of rehabilitating patients with leprosy they occasionally request plastic surgical correction of their facial disfigurement due to redundant and wrinkled skin. This motivation usually is on a psychological basis since other patients with similar deformities but who are adjusted to an institutionalized environment do not seek such relief.

During the past ten years a variety of corrective procedures have been done for redundant and wrinkled facial skin on patients at Carville. These include lobuloplasty of the external ear, blepharoplasty for the upper and lower eyelids, and both the conventional, and a modified, nasolabial rhytidectomy. The type and number of procedures done by the plastic surgical staff are listed in Table 1.

Some specimens were dominantly redundant and others dominantly wrinkled; however, both features were present to some degree in all cases. The specimens were stained with hematoxylin-eosin, acid-fast, Masson's trichrome, and Verhoeff's stains.

RESULTS

In most cases with lepromatous disease of long standing, the epidermis is thin and possesses a flat dermal-epidermal junction due to absence of the rete pegs. In the immediate sub-epidermal area, there is a zone which is uniformly free of lepromatous infiltrate and in which area collagen is usually unaffected ("free zone"). Although disease infiltrate mainly involves the upper layer of the dermis, it may extend throughout its entire thickness.

In progressive, active disease, dermal collagenous and elastic fibers are replaced by masses of cellular infiltrate, between which narrow strands of collagen are noted (Fig. 2). One interpretation of such findings is that the dermal fibers are displaced and compressed by the lepromatous infiltrate.

With chronicity of the disease process, foci of cellular infiltrate are noted within a more loose fibrillar matrix which possesses a granular and/or homogeneous substance. With this reduction of cellular infiltrate, collagen and elastic tissue fibers are loosely arranged, fragmented, and even absent. Residual cellular infiltrate becomes sparse

¹Received for publication 21 July 1973.

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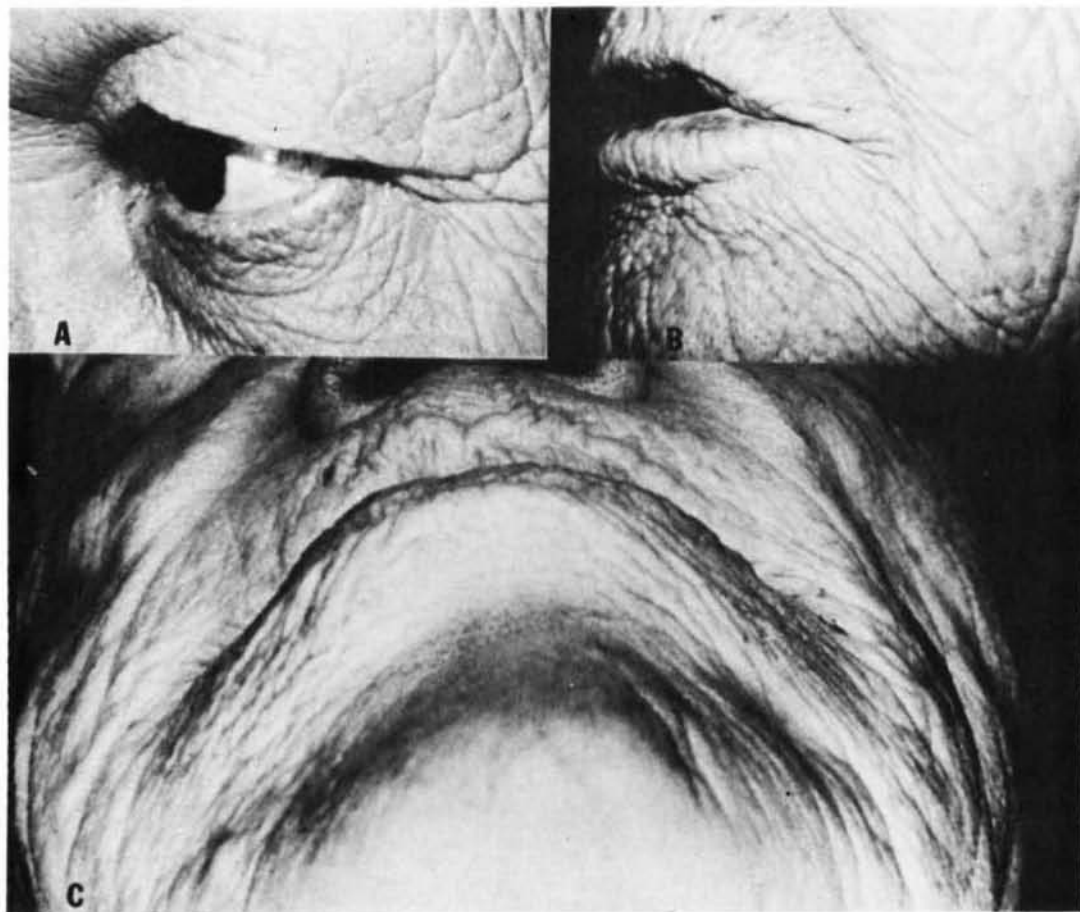


FIG. 1. A. Pendulous upper eyelid.
 B. Generalized wrinkling, facial skin.
 C. Redundant and wrinkled skin, nasolabial region.

TABLE 1. *Plastic surgical procedures of facial skin done at Carville from June 1963 to January 1973.*

Procedure	No. procedures	No. specimens
Bilateral lobuloplasty	31	62
Blepharoplasty:		
Bilateral upper & lower	9	36
Bilateral upper	8	16
Bilateral lower	12	24
One upper lid	4	4
One lower lid	1	1
Rhytidectomy:		
Conventional—right	8	8
left	7	7
Nasolabial	5	5
Total	85	163

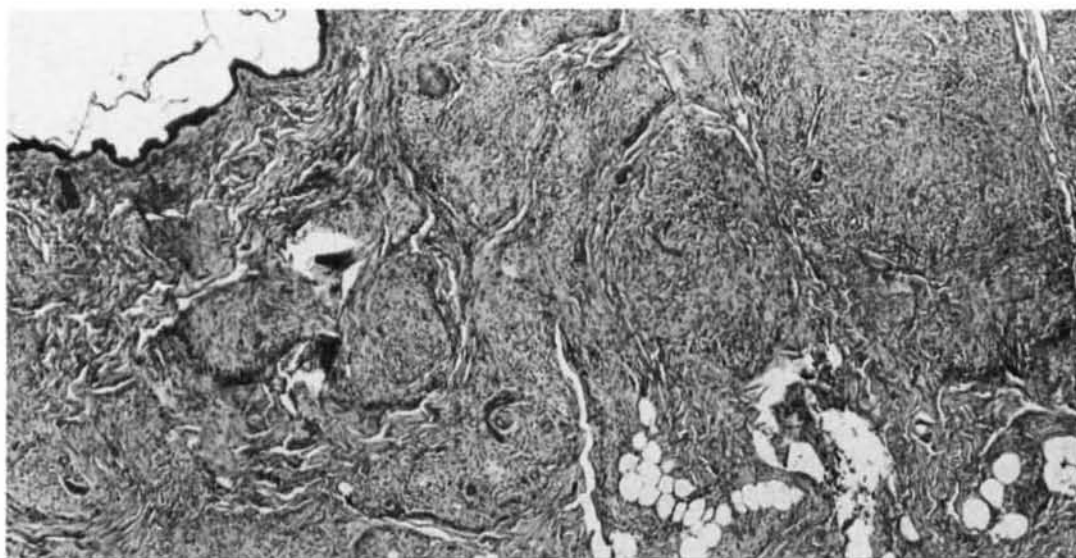


FIG. 2. H & E stain magnified X10. Epidermis thin without rete pegs. Dermis occupied by masses of leprosy cellular infiltrate, separated by narrow strands of collagen and set apart from the epidermis by the "free zone" of unaffected collagen.

and continues to be seen in the upper dermis.

Acute reaction, on the other hand, shows changes dominantly localized at the sites of the lesions. Both collagen and elastic tissue fibers are destroyed within this process, and in healing, the area is replaced by fibrosis (³). Here the histopathologic changes favor the vascular areas of the middle and lower dermis, though they may involve the entire thickness of the dermis besides subcutaneous tissue.

Other significant findings noted are elastotic changes which present a characteristic histologic picture; however, these are not related to leprosy but are instead attributed to solar degeneration (^{1,2}). In these areas the collagen is replaced by a disorganized material consisting of granules and coarse, thick, short fibers that are stained black by Verhoeff's stain (Fig. 3). These changes were observed more frequently and were more extensive in the facial skin *per se* as opposed to the eyelids and earlobes. They always occurred in the upper dermis, with the "free zone" separating it from the epidermis. In patients with lepromatous leprosy, the skin of the eyelids was dominantly redundant and histologically showed minimal or no elastotic changes but a definite decrease or absence of elastic tissue fibers.



FIG. 3. Verhoeff's stain magnified X10. Elastotic changes manifest by disorganized granular and coarse, thick, short fibers noted in the upper dermis separated from the epidermis by the "free zone."

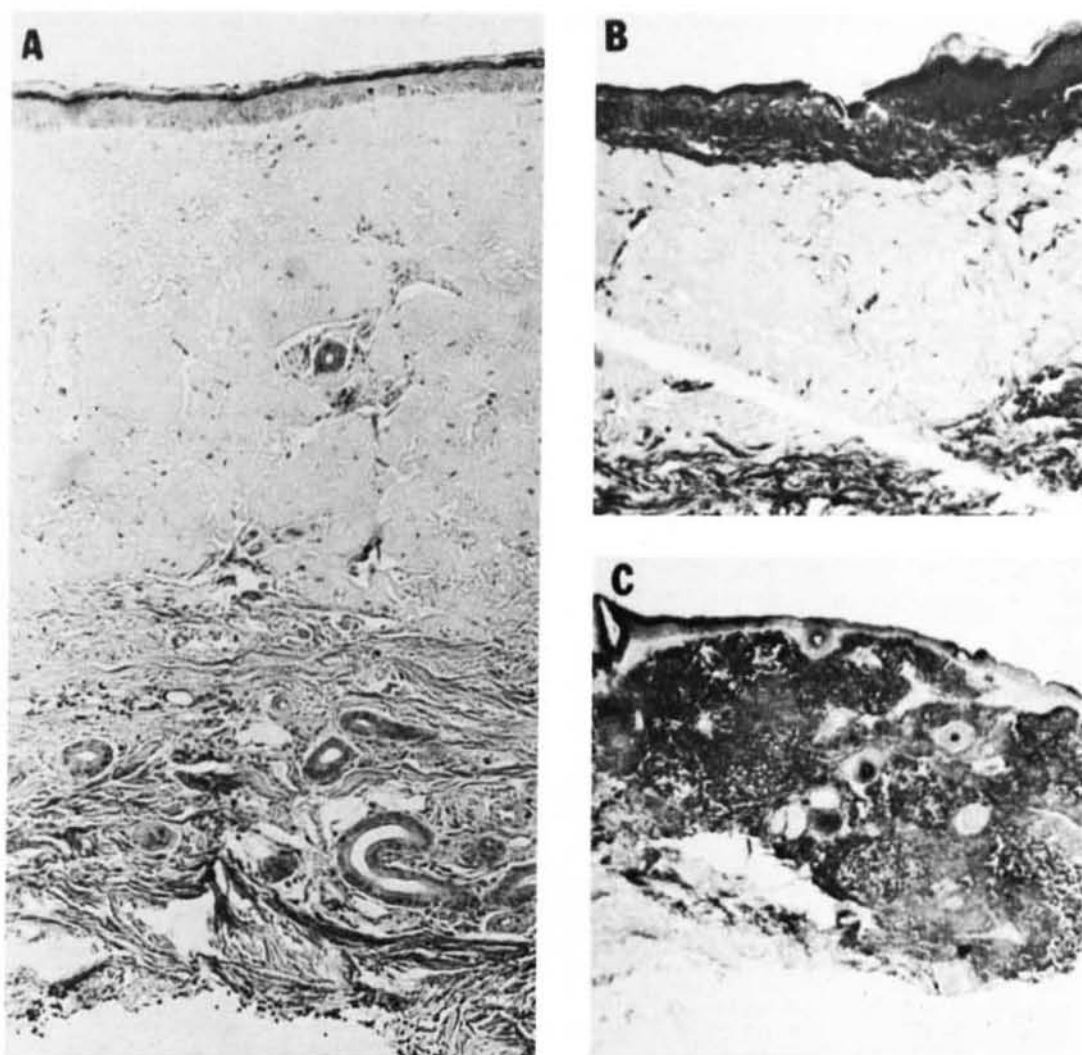


FIG. 4. Specimen from wrinkled facial skin of Figure 1B showing:

A. H & E stain magnified X10, extensive basophilic degeneration occupying a continuous broad band across the entire section.

B. Masson stain magnified X4 accentuating collagen especially in the "free zone."

C. Verhoeff's stain magnified X4, showing dominantly granular elastotic changes occupying the area of basophilic degeneration.

Severe elastotic changes were seen in greater degree than one would anticipate from senile and actinic effects; however, its correlation with lepromatous disease is not ascertained from the studies. It is of interest that although basophilic degeneration of collagen was observed in specimens stained by hematoxylin-eosin (Fig. 4), elastotic changes were not always demonstrated by Verhoeff's stain in corresponding areas of the tissue.

DISCUSSION

Degenerative changes are noted affecting

both collagen and elastic tissue fibers in facial skin of patients with leprosy. Whereas large areas of collagen are replaced and apparently destroyed by compression by masses of cellular infiltrate, collagen fibers were never observed to be entirely absent in any one specimen. Although collagen fibers escaped destruction within the "free zone," there were varying degrees of loss of collagen due to destruction by lepromatous disease below this level.

Loss of elastic tissue fibers ranged from a minimal decrease with fragmentation and

granular changes to an apparent total absence. As with the changes that affect collagen, initial loss occurs in the upper dermis where leprosy infiltrate is usually encountered. These changes are accompanied by varying losses of collagenous fibers which are the effect of lepromatous disease.

Clinically, skin infiltrated with lepromatous disease may appear thickened due to the infiltration of the cellular reaction whereby, in advanced cases, the patient acquires the so-called *leonine facies*. When the disease infiltrate regresses, the thickening is resolved, but the stretched skin remains thin and redundant. The sequelae of pendulous and sagging redundant skin commonly involves the earlobes and eyelids (Fig. 1A), in contrast to wrinkling which dominantly affects the facial skin *per se* (Fig. 1B), although both changes are often seen concomitantly in the same area (Fig. 1C). This manifestation is not unlike a *cutis laxa* in which there is a decrease in the amount of elastic tissue noted histologically (^{5,6}).

Opposite to the changes manifested by diminution or absence of collagen and elastic tissue fibers, attributed to lepromatous disease, are the findings of elastotic changes due to solar degeneration. These changes consistently occupy the upper dermis only. Here advanced degenerative collagenous changes appear as basophilic degeneration when stained by hematoxylin and eosin. Although such areas usually stain black with Verhoeff's stain to indicate that the collagenous degeneration corresponds to the presence of elastosis, the latter observation was not demonstrated in every case. However, this is not unusual, for according to Niebauer and Stockinger (⁴), degeneration of collagenous fibers is the first step in the formation of elastotic changes, and they must become electron dense before elastosis becomes evident by Verhoeff's stain.

As the majority of our patients are in the fourth and fifth decades of adulthood, it is not unusual to expect lepromatous facial skin manifesting both redundancy and wrinkling.

In knowing the underlying pathology of these changes, one is better able to establish the criteria for surgical correction. Definitive rhytidectomy should never be performed in the presence of active, progressive lepromatous disease. Surgery should be un-

dertaken only in those cases that are clinically and bacteriologically inactive. In retrospect, even in these cases rare foci of cellular infiltrate are observed sparsely distributed within the upper dermis of the operated specimen. The post-disease degenerative status of the collagen and elastic tissue and the possibility of reactivation of disease are factors to be considered as predisposing to unfavorable results from surgery with a proneness for recurrence of redundancy.

SUMMARY

Multiple collagen and elastic tissue fiber changes were noted in the majority of specimens of facial skin from patients with lepromatous leprosy.

Two gross manifestations affecting the skin are observed and appear to be correlated with specific histopathological findings.

Redundant sagging facial skin is associated specifically with a quantitative decrease or absence of elastic tissue fibers. This process may or may not be accompanied by loss of collagen. Clinically, skin infiltrated with lepromatous disease may appear thickened due to the infiltration by the cellular reaction whereby, in advanced cases, the patient acquires the so-called "leonine facies." When the disease infiltrate regresses, the thickening subsides, but the stretched skin remains thin and redundant. The sequelae of pendulous and sagging redundant skin commonly involves the earlobes and eyelids, in contrast to wrinkling which dominantly affects the facial skin *per se*, although both changes are often seen concomitantly in the same area. This manifestation is not unlike a *cutis laxa* in which there is a decrease in the amount of elastic tissue noted histologically, however, the changes in this instance are attributed to lepromatous disease. Wrinkling, on the other hand, is associated with elastotic changes and is attributed primarily to solar degeneration. Although the frequency and extent of elastotic changes were notable, it is interpreted as not unusual since it occurs in exposed facial skin. These findings therefore are considered incidental and are not related to leprosy from a causal standpoint.

RESUMEN

En la mayoría de una serie de muestras de piel facial de pacientes con lepra lepromatosa, se ob-

servaron múltiples alteraciones del colágeno y de las fibras de tejido elástico.

Se observan dos importantes manifestaciones que afectan la piel que parecen estar relacionadas con los hallazgos histopatológicos específicos.

La piel flácida colgante de la cara está asociada específicamente a una disminución cuantitativa o ausencia de las fibras de tejido elástico. Este proceso puede o no estar acompañado de pérdida de colágeno. Clínicamente, la piel infiltrada por la enfermedad lepromatosa puede aparecer engrosada debido a la infiltración por la reacción celular, a raíz de lo cual en los casos avanzados el paciente adquiere lo que se ha llamado "facies leonina." Cuando el infiltrado producido por la enfermedad regresa, el engrosamiento disminuye, pero la piel estirada permanece delgada y flácida. La secuela de piel colgante y flácida generalmente compromete los lóbulos de las orejas y los párpados, en contraste con las arrugas que afectan en forma dominante la piel de la cara "per se," aunque ambos cambios se observan a menudo en forma concomitante en la misma área. Esta manifestación tiene un cierto parecido con un *cutis laxa*, en el cual hay una disminución de tejido elástico diagnosticada histológicamente, sin embargo, en estos casos las alteraciones se atribuyen a la enfermedad lepromatosa. Las arrugas, por su parte, se asocian con cambios elastóticos y se atribuyen primariamente a degeneración solar. Aunque la frecuencia y extensión de los cambios elastóticos fue notable, esto se interpreta como no fuera de lo corriente, ya que ocurre en piel facial expuesta. Por lo tanto, estos hallazgos han sido considerados incidentales y no relacionados con la lepra desde un punto de vista causal.

RÉSUMÉ

Des modifications multiples du collagène et des fibres élastiques des tissus ont été relevées dans la majorité des échantillons de peau prélevés au niveau du visage chez des malades atteints de lèpre lépromateuse. Deux manifestations importantes qui affectent la peau ont été observées. Elles semblent être en relation avec des observations histopathologiques spécifiques.

L'effondrement de la peau du visage est associé de façon spécifique avec une diminution quantitative ou avec l'absence de fibres tissulaires

élastiques. Ce processus peut être ou non accompagné d'une perte du collagène. Sur le plan clinique, la peau infiltrée par la lèpre lépromateuse peut apparaître épaissie par suite de l'infiltration par une réaction cellulaire, ce qui aboutit chez les cas avancés à ce que l'on appelle le "facies léonin." Lorsque l'infiltration pathologique régresse, l'épaississement persiste, mais la peau qui a été étirée demeure épaisse et boursoufflée. Les séquelles de ce genre, avec peau pendante, s'observent généralement au niveau des lobes des oreilles et des paupières, ce qui contraste avec les rides qui touchent de façon prédominante la peau du visage. Néanmoins, les deux types de modification coexistent dans la même région. Cette manifestation n'est pas sans analogie avec la *cutis laxa*, qui se caractérise par une diminution dans la quantité de tissu élastique que l'on relève à l'examen histologique; néanmoins, dans le cas de la lèpre, les modifications sont attribuées au type lépromateux. Par ailleurs les rides sont associées avec des modifications élastotiques, attribuées essentiellement à une dégénération (solar degeneration). Quoique la fréquence et l'étendue des modifications élastotiques soient notables, on considère que cela n'est pas inhabituel, car cela peut survenir au niveau de peau faciale exposée. Ces observations des lors sont considérées comme marginales, et ne sont pas liées à la lèpre d'un point de vue causal.

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