ELASTOSIS PERFORANS SERPIGINOSA
A CASE REPORT WITH HISTOCHEMICAL
AND
ENZYME DIGESTION STUDIES*
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In 1952 Lutz (1) described a peculiar serpiginous keratotic eruption that was present on the sides of the neck of a 21-year-old man for 8 years. He named the lesion keratosis follicularis serpiginosa. Histologic studies of the keratotic wall showed that it was composed of a cystic structure containing horny lamellae situated just beneath the epidermis and that it appeared to have arisen from the hair follicles. Under the same title, Beening and Ruiter (2) in 1954 described the eruption in a 13-year-old girl who had developed ring-formed and semi-circular lesions on the side and nape of her neck. When first seen 3 years before, these lesions consisted of a number of closely-set hailstone-sized, hard, reddish papules. The summit of each papule revealed a keratotic plug which when removed revealed a deep depression. The center of the rings appeared depigmented but not atrophic. Three years later most of the lesions had undergone involution. Histologic study of specimens removed by biopsy showed a markedly acanthotic epidermis which contained horny lamellae. Serial sections demonstrated that the horny masses penetrated into the cutis in some places. Where they remained in contact with connective tissue, inflammatory infiltrate containing small round cells, histiocytes and occasional giant cells were found. The hyperkeratosis appeared to have a direct relationship to the hair follicle and the cyst formations were apparently connected with the sweat glands. Neither of the aforementioned authors elucidated the true nature of the horny lamellae found in the cysts.

Miescher (3) was first to do elastic staining and recognize the presence of elastic fibers in the lesion. He felt that the lesion was caused by the pushing out of the necrotic mass through the epidermis which reacted to it with acanthosis, hyperkeratosis and parakeratosis. This suggested an intrapapillary hyperplasia of elastic tissue as the primary disorder. He applied the name of elastoma intrapapillare perforans verruciforme to this disease. Hitch et al (10) found that the elastic tissue penetrated into the epidermis at the depth of rete ridges and into the sides of follicles. The present paper is a report of histologic and enzyme digestion studies carried out on a recently discovered lesion in an 18-year-old boy.

CASE REPORT
The patient, an 18-year-old healthy schoolboy, had noted the appearance of a few keratotic papules on the back and sides of his neck, 17 months prior to his first examination. The lesions had gradually increased in number and become confluent and had formed semi-circular and annular configurations with clear non-atrophic centers. Observation over a period of seven months revealed no essential change in the character of the lesions (Figures 1 & 2).

EXPERIMENTAL STUDIES
Materials and Methods
The material studied consisted of four specimens of skin from the right side of the neck of the patient presented above. They were removed under local procaine anesthesia. The procaine was infiltrated in an area 1 cm. away from the lesion to avoid distortion of the histology. One biopsy specimen measuring 0.5 x 0.5 x 0.3 cm. was fixed in 10% formalin and embedded in paraffin. Sections were cut and examined both unstained and stained with hematoxylin and eosin, Verhoeff elastic tissue stain, Congo red, periodic acid-Schiff, Masson and van Gieson stains. The second piece of skin taken from approximately the same area of the patient measured 1.5 x 0.5 x 0.3 cm. It was frozen, sectioned, and exposed to elastase (prepared by Worthington* within 5 minutes

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Histologic Studies

Results

The epidermis showed spotty areas of hyperkeratosis, parakeratosis, and keratotic plugging, with a tissue defect extending through the whole thickness of the epidermis due to a perforation by fibrous material invading from beneath (Figure 3). In the vicinity of the perforation, the epidermis showed acanthosis, especially of the rete ridges. Fibrous material was present in the epidermis, sometimes occupying small, round, cystic spaces one above the other; thus, apparently, forming a spiral (Figure 5). Although there were similar changes in the hair follicles, as will be described below, none of these spirals could be traced down to hair follicles or sweat ducts. This fibrous material was stained black by Verhoeff, light pink by Congo red, and negative with PAS and Sudan III stains. The Masson and van Gieson stains were negative for collagen.
Fig. 3. Low power view of intrapapillary and intradermal increase of Verhoeff positive fibers. On the right there is one perforated lesion. Formalin fixation. Verhoeff's elastic tissue stain. × 25.

Fig. 4. Higher magnification of the intrapapillary lesion shown in the extreme left of Figure 3. Fine and coarse fibers are seen in the papilla. One projection of lesion is close to the surface epidermis, which is thinned out. Verhoeff's elastic tissue stain. × 100.

The dermis contained an increased amount of short, wavy, fairly thick, curled fibers. In the hematoxylin and eosin preparations, these varied in color between greyish blue and pink. The fibers showed the same tinctorial characteristics as those described in the epidermis, i.e. black with Verhoeff's stain, negative with the periodic acid-Schiff, and pink with the Congo red stains. They ran parallel to the surface epidermis, were sparse in the deep dermis, and dense in the middle and upper portions where they partially lay in clumps (Figure 3). In these foci, there were found lymphocytes. Some of the tips of the rete ridges in the involved areas were invaded by those fibrous
FIG. 5. Tortuous spiral formation in the epidermis. In the lower field at the left there are several hair follicles which underwent degeneration. Frozen section. Hematoxylin and eosin. × 50.

FIG. 6. Invasion of three tips of papillae by coarse reddish fibers from subpapillary dermis. Hematoxylin and eosin. × 100.

FIG. 7. Higher magnification of the perforated area shown to the extreme right in Figure 3. At the right corner of the bottom of the crater, there is a tangled mass of pinkish red coarse fibers surrounded by red blood cells. Hematoxylin and eosin. × 100.
Enzyme Digestion Studies

1/2 hour incubation in elastase, followed by Verhoeff stain: A considerable amount of Verhoeff positive material was found dissolved out in comparison to the control sections incubated in Michaelis buffer without elastase. The remaining material showed swelling, segmentation, clumping, and uneven staining. It appeared largely amorphous and structureless. However, some swollen remnants of fibrous structures could be seen (Fig. 8). Interestingly enough, the epidermis was simultaneously digested, leaving only faint
Fig. 10. 1½ hour incubation in elastase solution. The epidermis is largely digested. Projecting dermal papillae are devoid of Verhoeff positive material except for one small area in subpapillary dermis (see arrow). Frozen section. Verhoeff’s elastic tissue stain. X 40.

DISCUSSION

Since Lutz in 1952 reported the first case of what in retrospect was elastosis perforans scrofuligiosa, more than ten case reports have appeared (2-13). Clinically this disorder is characterized by the onset in late childhood or early adulthood, affecting an otherwise healthy individual. The outer configuration of the lesion frequently changes, and spontaneous remission may occur. No subjective symptoms or systemic manifestations have been observed. The clinical picture resembles tinea, verruca vulgaris, or porokeratosis. There is no evidence of pseudoxanthoma elasticum or senile skin changes in any one of the previously reported cases.

Although the etiology is unknown, the basic lesion appears to involve the elastic fibers. The positive Verhoeff, and Congo red stains, the negative Masson and Van Gieson stains, and the selective digestion of the fibers with elastase seem to confirm this. The changes in the elastic tissue are present in the subpapillary region of the dermis, and this process is seen in the papillary bodies and epidermis. The tinctorial reaction of these elastic fibers is strongest in the subpapillary layer and weakest in the cystic spaces in the upper epidermis.

Intrapapillary increase in elastic tissue varied
Fig. 11. 2 hour incubation in elastase solution. Advanced degree of dissolution of the Verhoeff positive fibers in the lesion in the epidermis and two completely empty spaces. Partially digested epidermis is left with a number of shreds of Verhoeff positive fibers which appear broken and smudgy. Frozen section elastic tissue stain. X 100.

Fig. 12. Control 2 hour incubation in plain buffer solution. Two intrapapillary lesions contain distinctly fibrous, Verhoeff positive components in them. Frozen section Verhoeff’s elastic tissue stain. X 100.
from place to place according to the fiber size; from fine mesh-work (of probably young fibers) to a clumping of thicker ones in the bulged papillae. It was almost always possible to see a direct communication of the intrapapillary elastic fibers with those situated beneath in the upper dermis. The elastic tissue in several areas invaded the lower poles of rete ridges. On the other hand, direct invasion of appendages by the elastic tissue was not seen, although several degenerated hair follicles in the epidermis were observed.

The elastase digestion experiment demonstrated progressive disruption and digestion of the fibers in the lesion, while the controls showed essentially no change. Marshall and Lurie (7) showed that the elastic material lost its staining reactions after digestion with elastase, and Dammert and Putkonen (9), demonstrated that trypsin removed the elastic material. Epidermal cells were also digested by the elastase preparation which is known to contain minor protein contaminants. The elastase was prepared from porcine pancreas and presumably a small amount of a proteolytic enzyme that escaped destruction in the repeated crystallization of the elastase digested the epidermal cells. This contaminant enzyme, however, does not affect elastic tissue (14).

The present lesion has been variously named by several authors. Originally it was named Keratosis Follicularis Serpiginosa by W. Lutz. Subsequently, the terms Elastoma Intrapapillare Perforans Verruciforme and Elastosis Perforans Serpiginosa were suggested by G. Miescher and by Dammert and Putkonen, respectively. As Hitch has pointed out, the elastic tissue increase is not primarily or necessarily "intrapapillary" or "follicular" but in the cutis in general. There is no evidence that it is a tumor, as the suffix "oma" suggests, but a morbid process of abnormal increase in elastic tissue, as the name elastosis might convey. Clinically it takes serpiginous configuration with eruptive individual lesions. Therefore, we feel that the term suggested by Dammert and Putkonen Elastosis Perforans Serpiginosa is more suitable than the others.

**SUMMARY**

1. Histochemical and enzyme digestion studies have been carried out on biopsy material from a patient with elastosis perforans serpiginosa.

2. The nature of the lesion, as representing primarily a proliferation and degeneration of elastic tissue, has been confirmed.

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