ESSENTIAL SHRINKING OF THE CONJUNCTIVA IN A HEREDITARY AFFECTION ALLIED TO EPIDERMOLYSIS BULLOSA

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With 1 Colour plate (I) and 11 Photographs.

As its name implies, Essential shrinking of the conjunctiva is aetiologically an ill-defined entity. The more precise alternative name of Pemphigus of the conjunctiva, though widely used, is valid only exceptionally, for few cases are due to pemphigus proper. Nor is vesicle formation, as distinct from pemphigus, always present. The vesicular skin affections do, however, play a great and possibly predominant part aetiologically. Shrinking of the conjunctiva has been observed as a rare complication in such affections as pemphigus malignus vulgaris, dermatitis herpetiformis, erythema exudativum multiforme, and blister formation after toxic reactions such as those arising from barbiturates, arsenic and possibly gold. It has also been recorded in isolated cases of epidermolysis bullosa.

The present report is of interest in that the shrinkage occurred in an unusual vesicular affection of the skin and mucous membranes, and in that it was observed in two sisters, the offspring of consanguineous parents.

CLINICAL RECORD

Miss Ethel H., aged 50 years, sought advice because of her fear of impending blindness. She also complained of irritation from in-turning lashes. She had no eye trouble until the age of 14 when "the eyes became inflamed". This inflammation subsided after a few weeks, but recurred frequently, generally at intervals of not more than a few months. She had a severe inflammation in the left eye at the age of 26, necessitating in-patient treatment, and a similarly severe attack in the right eye when she was 44 years old. Her sight has been declining steadily since the early attacks. On examination vision was less than 6/60 in each eye, not improved
by glasses. The corneae were irregularly scarred in their central and lower areas. There were inturning lashes at both lower lid margins. The lower fornices were somewhat shallow and the lower limbus in each eye was ill-defined owing to a thickened, pearly white broad fold of the conjunctiva encroaching on the cornea. Similar white thickenings, several in number, could be seen on each cornea, but they did not appear to be contiguous with each other or the main conjunctival fold. The bulbar conjunctiva was not abnormally moist, but was somewhat injected below; there was also some injection in the opposing palpebral conjunctiva. The upper fornices and conjunctiva were normal. With the slit-lamp no pannus or corneal vascularisation could be detected. It could also be confirmed that the thickenings seen on the cornea were independent of the conjunctival fold. Trachoma could clearly be ruled out and a provisional diagnosis of essential shrinking of the conjunctiva was made.

The speckled appearance of the skin of the patient's face and the striking deformity of her hands suggested the possibility of a generalised chronic condition. A history that an elder sister was blind and more severely affected by the same skin trouble and deformity of hands than the patient herself, led to a detailed study of both these women. This elder sister was admitted for in-patient treatment and the younger one has been seen frequently during the past 18 months. Their condition has remained substantially unchanged during that time. Their ready co-operation greatly facilitated this study.

**Family History**

As will be seen in the pedigree chart (Fig. 1), the two affected sisters had four normal brothers and one normal sister. Their father and mother were first cousins, their paternal grandfather and their maternal grandmother being brother and sister. Their father and mother were normal and died at the age of 83 and 84 respectively. The father's brother emigrated to Australia, his sister to New Zealand. They and their numerous children were all healthy. The mother's brothers and sisters emigrated to the United States. They too, and their numerous children, were healthy. A male member of the sibship containing the affected sisters has extensive symptomless vitiligo on the forearms and thighs. None in