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Familial atrophic rhinitis: a scanning electron microscopic study

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Introduction

THE aetiology of primary atrophic rhinitis, as opposed to atrophic rhinitis secondary to specific infection, granuloma, trauma (including surgery) or neoplasm, is unknown, although many theories have been put forwoard. These are reviewed in a related publication (Gray *et al.*, 1980). Isolated reports of members of the same family are noted, but they have not so far provided sufficient evidence to determine the specific mode of inheritance. Ssali (1973) reports a family with seven members affected, while Girgis (1966) quotes an instance of identical twins with atrophic rhinitis and Young (1967) mentions a mother and daughter both affected. However, Sibert and Barton (1980) have recently reported a family in which the father and 8/15 of his children have primary atrophic rhinitis. They conclude that the inheritance pattern in this particular family is autosomal dominant and that a wider genetic study is indicated.

Two of the children in the family underwent surgical closure of the nostrils in order to alleviate their symptoms, while a third child suffered a fractured nose and agreed that a biopsy should be taken while under anaesthesia for reduction of the fracture. A fourth member of the family with established atrophic rhinitis agreed to an elective biopsy.

Thus the opportunity arose to obtain specimens for scanning electron microscopy, a technique now proving to be of considerable interest in the study of nasal mucosa.

Materials and methods

Biopsies of the nasal mucosa of three daughters and one son of Mr. P.S., who himself has severe primary atrophic rhinitis, were taken under general anaesthesia. In one daughter biopsies were taken only after re-opening one of the nostrils; in the second, biopsies were taken before closure, and in the third a biopsy was taken when a coincidental fracture of the nasal bones was reduced. The son agreed to have an elective biopsy but declined any definitive procedure.

All biopsies were fixed in 3 per cent glutaraldehyde, processed and examined in a Coates and Welter Field Emmission Scanning Electron Microscope (SEM) at 16K volts.

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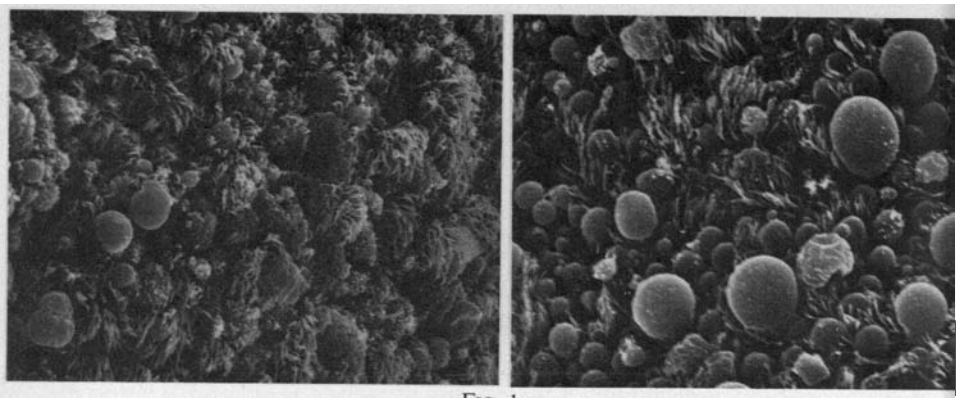


FIG. 1
Near-normal ciliary population. Some underdeveloped cilia. (×400).

FIG. 2
Mucus globules. (×1,000).

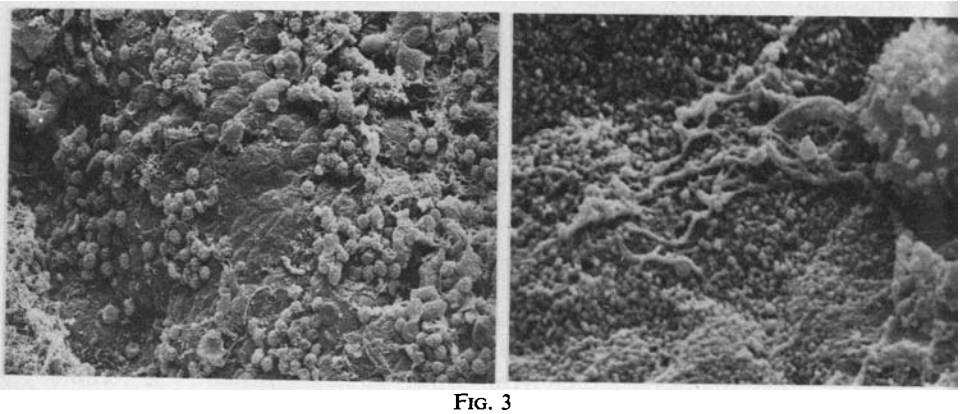


FIG. 3
Established atrophic rhinitis. (×250).

FIG. 4
Established atrophic rhinitis. (×300).

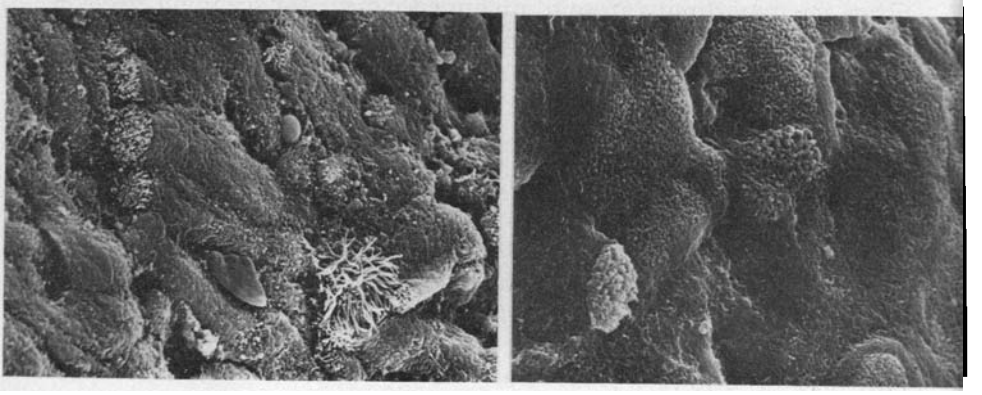


FIG. 5
Early atrophic rhinitis. (×1,000).

FIG. 6
Established atrophic rhinitis. (×1,000).

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Results

Case 1—Mrs. N.M. (b. 1946). Atrophic rhinitis was first diagnosed in 1965. Local medical treatment was not helpful, and in 1966 a cervical sympathectomy was carried out, but this gave no relief. An attempt was made to narrow the airways in 1972 by the insertion of silastic implants, but these were rapidly extruded. In 1974 the nostrils were closed surgically. After two years the nostrils were reopened with improvement in symptoms and in the appearance of the mucosa. At this stage biopsies of both inferior turbinates were taken. These show a near-normal ciliary population, although some are not fully developed (Fig. 1), but with the mucus forming globules rather than the normal coherent blanket (Fig. 2). Since 1976 her symptoms have recurred but, understandably, the patient is not anxious to undergo further surgery.

Case 2—Miss T.S. (b. 1957). Is the sister of Mrs. N.M. (Case 1) and also has severe atrophic rhinitis. Notwithstanding her sister's relatively short-lasting improvement following closure, she agreed to have, initially, one nostril closed surgically. This was carried out in December 1977 and biopsies were taken of the left inferior turbinate. These show complete absence of cilia with denuded epithelial cells and many more white blood cells than would be found from blood spilt at biopsy. These are enmeshed in debris and fibrin (Fig. 3). A single white blood cell adherent to the non-ciliated cell surface by fibrin-like strands is clearly seen (Fig. 4).

Case 3—Miss L.S. (b. 1953). A younger sister of the two previous patients was admitted for reduction of a nasal fracture. She complained of no previous nasal symptoms and examination showed the nasal mucosa to be apparently normal. She agreed to a biopsy of the nasal mucosa while under anaesthetic and surprisingly, in view of the clinical appearance, abnormalities were shown by SEM. The specimen was carefully searched before a small clump of irregular cilia were found. Attempts at ciliary formation can be seen nearby in stunted patches. There is no mucous blanket (Fig. 5). These appearances are compatible with early primary atrophic rhinitis, and it was of considerable interest that when the whole family was re-examined eight months later, this girl showed undoubted clinical changes of atrophic rhinitis with small inferior turbinates, airways markedly wider than normal, and a dry mucous membrane. After a further seven months, crust formation was noted.

Case 4—Mr. A.S. (b. 1959). The brother of the above patients had well-established atrophic rhinitis. While considering whether to undergo closure of the nostrils he agreed to biopsy of the inferior turbinates. He later declined further surgery, but the nasal mucosa shows typical changes of atrophic rhinitis. The epithelial cells are bare of cilia and the mucous blanket is depleted. A patch resembling stunted cilia is seen in the bottom right hand corner. (Fig. 6).

Discussion

Although there have been isolated reports of more than one member of a family with atrophic rhinitis, the family in this study is the first instance where a specific mode of inheritance has been shown. The genetic and clinical aspects of this family are presented elsewhere (Sibert and Barton, 1979) but the SEM reveals several interesting features.

In the two patients (Cases 2 and 4) with established severe atrophic

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rhinitis who had not undergone nostril closure the mucous membrane is similarly and severely damaged with complete loss of cilia and an abnormal mucous blanket. The significance of this is discussed in a related publication (Gray *et al.*, 1980). Case 1 who was severely affected prior to nostril closure had, on opening of the nostrils, evidence (by SEM) of good recovery on some epithelial cells with large numbers of apparently healthy cilia. However a high proportion of cells remained unciliated. Even if areas of cilial regeneration were functional, their very sparseness suggests that flow of a mucous blanket would be poor. Nevertheless the improvement on SEM fits with Young's findings of clinical improvement after nostril closure (Young, 1967). However in this patient gross clinical atrophic changes re-appeared within two years of reopening the nostrils.

It is of interest that in case 3, a female aged 13, SEM abnormalities in an apparently normal nose progressed to obvious clinical changes of atrophic rhinitis within eight months. It is noted elsewhere that atrophic rhinitis first became manifest in several members of the family around puberty (Barton and Sibert, 1980).

At present Mr. P.S.'s fifteen children themselves have a total of twenty-two children, all but five being under ten years old. One grandchild aged fifteen is said by the family, who are themselves well aware of the inherited nature of the condition, to have a 'smelly nose' but when examined by us showed no abnormality. Two other grandchildren do show abnormalities in the form of wide airways with dry mucosa but, as yet, the changes are not sufficient to merit the diagnosis of atrophic rhinitis. However the dominant pattern of inheritance is clear over two generations, and it will be interesting to note the outcome of the third generation in due course.

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