Dominant inheritance in a family with primary atrophic rhinitis

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SUMMARY Primary atrophic rhinitis is an uncommon condition which presents with crusts in the nose. The nasal mucosa is dry and atrophied and the nasal cavities are abnormally wide.

We report a large London Irish family with an affected father with fifteen children. Eight of these have primary atrophic rhinitis. Symptoms appear around puberty, and there was one case in the third generation with an affected mother. The nasal appearances of the affected members varied considerably and many hid their disease well.

The family fits well with dominant inheritance. A familial aetiology for primary atrophic rhinitis is a more attractive theory than those previously postulated.

Primary atrophic rhinitis is an uncommon condition which presents with crusts in the nose. The smell of these is offensive to others and may cause much social distress to the patient. The nasal mucosa is dry and atrophied and the nasal cavities are abnormally wide. Paradoxically, the patient may complain of nasal obstruction, probably because of the changes to the nerve endings causing lack of appreciation of the air currents. There have been many suggestions for the aetiology of this condition including hormonal, infective, and dietary theories. None, however, has been proved. There have been sporadic reports of familial cases in published reports. We report a large family with a father, eight of his fifteen children, and one grandchild with the condition.

Case reports

The S family is a large Irish family living in West London. The father (case I.1) comes from Belfast. He denied symptoms but on examination he had gross changes of atrophic rhinitis. The family tree is illustrated in the figure, and the case histories are shown in the table. There is no reliable information on the more distant family and the sibs of case I.1. There is no consanguinity and his wife (case I.2) is normal.

Discussion

The family fits well with dominant inheritance; eight of fifteen sibs and their father are affected. Moreover, there is an example of the condition being

FIGURE S family tree.

TABLE Case histories of S family

Case	Sex	Age (July 1978)	Diagnosis
I.1	M	68	Marked atrophic rhinitis with crusting
II.1	M	35	Normal
II.2	F	34	Normal
II.3	F	33	Small turbinates; dry mucosa; mild signs of atrophic rhinitis
II.4	F	32	Extensive surgery; severe changes of atrophic rhinitis
II.5	F	30	Affected; severe crusting of atrophic rhinitis
II.6	F	30	Severe rheumatoid arthritis; nose normal
II.7	M	28	Nose normal
II.8	F	25	Mild changes; dry mucosa; small turbinates
II.9	F	25	Severe atrophic rhinitis
II.10	F	21	Atrophic rhinitis
II.11	M	20	Normal
II.12	M	19	Severe atrophic rhinitis
II.13	M	17	Normal
II.14	F	15	Small turbinates with abnormal biopsy; mild atrophic rhinitis
II.15	F	14	Normal
III.1	F	6	Not examined; thought to be normal
III.2	F	4	Not examined; thought to be normal
III.3	M	2	Normal
III.4	F	16	Not examined; thought to be normal
III.5	M	15	Not examined; thought to be normal
III.6	M	14	Not examined; thought to be normal
III.7	F	6	Normal
III.8	M	5	Not examined; thought to be normal
III.9	M	15	Early changes of atrophic rhinitis
III.10	M	14	Normal
III.11	M	13	Normal
III.12	F	8	Normal
III.13	M	5	Normal
III.14	F	8	Not examined; thought to be normal
III.15	F	6	Not examined; thought to be normal
III.16	F	1	Not examined; thought to be normal
III.17	F	3	Not examined; thought to be normal
III.18	M	6 months	Not examined; thought to be normal
III.19	M	3	Normal
III.20	F	2	Normal
III.21	M	2	Normal

transmitted through three generations. There is variability of expression of the condition in the family with some members being severely affected, for example case II.4, and some members with only mild changes, for example case II.14. It appears that the condition does not develop significantly until puberty in this family.

Young¹ reported a mother and daughter affected, and Ssali² a family with seven affected members. Girgis³ reported the condition in identical twins. There are no other published reports of familial cases of which we are aware.

The way that several members of our family concealed their disease well suggests that an accurate family picture is difficult to obtain unless specifically searched for. There may be reduced fitness because of this condition, with unpleasant nasal odours making patients unattractive to the opposite sex. There is no evidence for this, however, in this family which clearly shows a dominant pattern of inheritance. A familial aetiology for primary atrophic rhinitis is a more attractive theory than those previously postulated. A larger genetic survey of this condition would be justified.

References

- ¹ Young A. Closure of the nostrils in atrophic rhinitis. *J Laryngol Otol* 1967;**81**:515-24.
- ² Ssali CH. Atrophic rhinitis. A new curative surgical treatment. *J Laryngol Otol* 1973;**87**:397-403.
- ³ Girgis IH. Treatment of ozaena by dermofat graft. *J Laryngol Otol* 1966;**80**:615-27.

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