DEGENERATIO PUNCTATA ALBESCENS*

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THIS condition was first described by Mooren (1882), since which date about fifty cases have been described in the literature. It is closely related to primary pigmentary degeneration of the retina, but has some fundamental differences. Duke-Elder (1940) describes it as "a familial disease of a degenerative type, commencing usually in early life, characterized by the presence of innumerable discrete white dots scattered over the whole fundus without pigmentary changes, and usually non-progressive and stationary". Although the disease is usually congenital or starts in very early life, a case was reported by Henderson (1934) which was first noted at the age of 17 years, after an attack of Vincent's angina. It is a familial disease, but has never been seen in more than one generation. A history of night-blindness and the presence of primary pigmentary degeneration in the same family is common.

Night blindness is usually congenital and sometimes depends more on a retardation of adaptation than on the lowering of the light threshold. A peripheral contraction of the visual fields most marked in dim light is present in more than half the cases, and central vision is good because the condition usually remains stationary. The fundus is peppered with discrete white dots which may be observed by the direct method of ophthalmoscopy. These dots lie deep to the retinal vessels, and are scattered all over the fundus, except for the foveal region. The pathology is unknown.

Case Report

A woman aged 48 years, of Portuguese descent, presented with difficulty with near vision for the past 8 years. She was unable to see well for the first hour of darkness, and the last symptom had been present since infancy. Her general health was good.

Family History.—The members of her family were very conscious of their disability to see adequately in dim light. Because of this trait, it was their custom to test their children with a white flower at dusk to ascertain whether they were affected or not. Although only the patient and her daughter had an ophthalmic examination (the others living in Madeira), this stigma was so well known that an accurate record was obtained of all those affected with one hour of night-blindness. The affected members were also stated to have blue irides (Fig. 1).

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FIG. 1.—Pedigree, showing association of night blindness with blue irides.

Examination.—The outward appearance of the eyes was normal. The irides were blue. Scattered throughout both fundi were innumerable whitish dots, some of which had coalesced to form a dumb-bell shape. These dots lay deep to the retinal vessels (Fig. 2). Pigmentation was absent, but the vessels and disc were normal.

FIG. 2.—Appearance of the right fundus. The dark areas in the photograph are due to chromatic error. Photographs of the left fundus were similar.

The visual acuity in each eye was 6/18, improved to 6/6 with +1.25 D sph., +0.25 D cyl. With an additional 1.25 D sph. to each eye the acuity was J.I.

The ocular tension was normal.

The visual fields showed slight peripheral constriction (Fig. 3).

FIG. 3.—Visual fields, showing peripheral constriction.

Laboratory Investigations.—No sugar or albumin was present in the urine, and the Wassermann reaction was normal.

Summary

A case of degeneratio punctata albescens, with an interesting family history of retardation of dark adaptation and peripheral constriction of the visual fields, is described.

REFERENCES

- DUKE-ELDER, S. (1940). "Text-book of Ophthalmology", vol. 3. Kimpton, London.
 HENDERSON, R. H. (1934). *Arch. Ophthalm. (Chicago)*, 11, 763.
 MOOREN, A. (1882). "Fünf Lustren ophthalmologischer Wirksamkeit", p. 216. Bergmann, Wiesbaden.