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Corneal Mosaic Patterns—Morphology and Epidemiology

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Summary
The corneas of 1485 consecutive patients were examined for the presence of a mosaic pattern. One hundred and ninety-nine peripheral mosaic patterns, ten posterior crocodile shagreens and two cloudy dystrophies were found. Peripheral mosaic pattern is age related and distinct from corneal arcus. Posterior crocodile shagreen is also age related and in one case was found to be unilateral. The morphology of the mosaic patterns is discussed.

The presence of a mosaic pattern in the cornea was described by Vogt in 1930. Since then five conditions where this type of pattern is evident have been documented:
(a) Posterior crocodile shagreen—at the level of the deep stroma and Descemet's membrane.
(b) Anterior crocodile shagreen—at the level of Bowman's layer.
(c) Cloudy dystrophy (François)—centrally from deep stroma to Bowman's layer.
(d) A peripheral crisscross appearance (peripheral mosaic pattern)—seen in corneal arcus.
(e) Anterior corneal mosaic—seen on applying pressure to the normal, fluorescein stained cornea.

In this paper we describe the prevalence of the conditions a–d mentioned above in a selected hospital population. The main features of each of the conditions are described, and their differences clarified. The nature of the mosaic appearance is discussed.

Method

During a five-week period in the summer of 1988, all new patients attending the Ophthalmic Accident and Emergency department (1294) and consecutive patients (191) from the Ophthalmic Outpatients department (total=1485) were examined. All patients unsuitable for slit-lamp examination or with marked corneal opacification were excluded from the study (54 patients). Examination of the cornea was undertaken on a Haag-Streit 900 slit-lamp using direct illumination. The presence of any mosaic pattern was recorded diagramatically with special note of its depth, distribution, shape and degree of symmetry between the two eyes. The age and sex of the patients was also recorded.

The distribution of the mosaic pattern was recorded on a diagrammatic cornea divided into quadrants and three concentric zones. The distribution was scored for the peripheral mosaic pattern (PMP) according to the number of quadrants and zones involved.

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Results

The age of the 1485 patients ranged from 3 to 95 years, with a mean age of 47 years (Fig. 1). The total number of males was 867 (mean age=45) and females 618 (mean age=51). The mosaic pattern was distributed either centrally (ten patients with posterior crocodile shagreen and two patients with cloudy dystrophy) or peripherally (199 patients); in only two patients did the central and peripheral mosaic pattern coexist.

The prevalence of PMP was found to be age related, no cases found below the age of 43 years, with a maximum prevalence beyond the 8th decade (Fig. 2). In all patients the involvement was symmetrical. The PMP was located in the region of Descemet's membrane, forming a confluent layer which faded as it was traced centrally from the limbus. There was always an associated 'granularity' of the anterior stroma overlying the area involved. The most frequent location was the infero-temporal quadrant; those patients with minimal involvement had the pattern confined to this quadrant. Those with greater central extension of the mosaic pattern had more quadrants involved (Fig. 3).

The prevalence of posterior crocodile shagreen was also found to be age related, no cases found below the age of 33 years, with a maximum prevalence beyond the 8th decade. The changes were confined to a well defined layer in the posterior stroma, consisting of a regular arrangement of polygons separated by lucent zones. In some cases a marked degree of asymmetry was noted, and in one case with only a faint mosaic pattern in one eye the other eye showed no visible changes.

Two cases of cloudy dystrophy were found, both being bilateral and asymmetrical in their distribution between the two eyes. They were easily distinguished from posterior crocodile

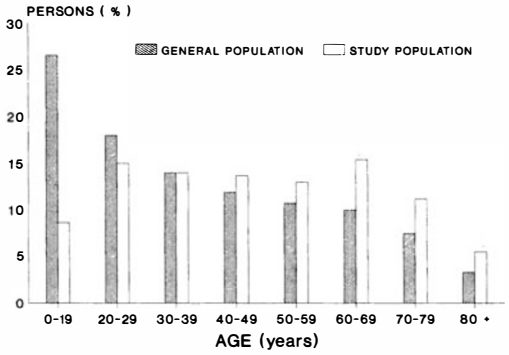

Fig. 1. *Age structure of general population of England & Wales (mid 1986) and study population (per cent).*

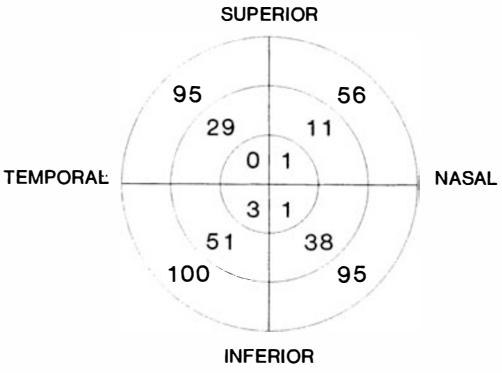

Fig. 3. *Corneal distribution of peripheral mosaic pattern. Figures (per cent) represent the proportion of cases in which each area is involved.*

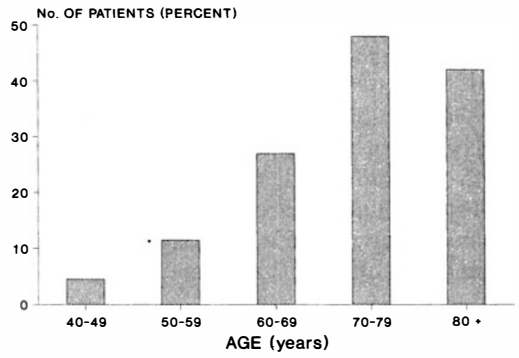

Fig. 2. *Age distribution of peripheral mosaic pattern.*

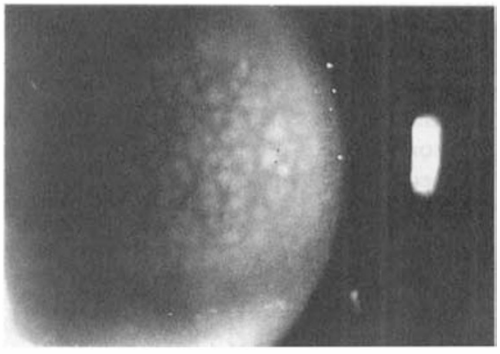

Fig. 4. *Posterior crocodile shagreen.*

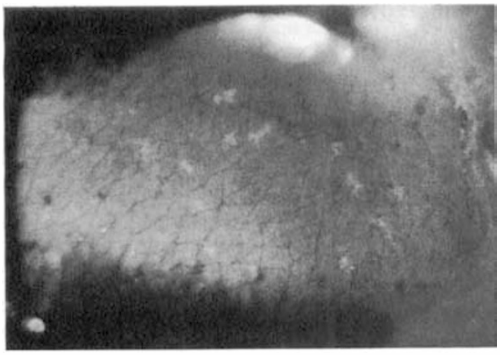

Fig. 5. *Anterior crocodile shagreen.*

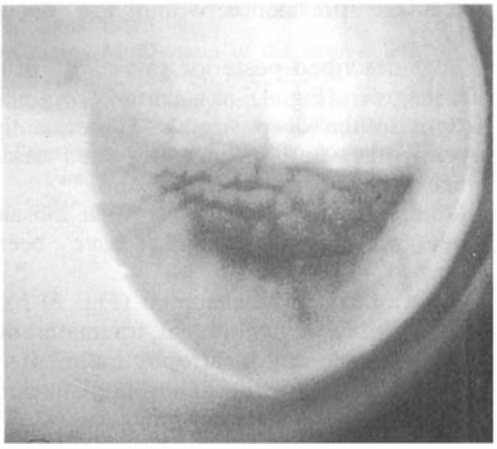

Fig. 7. *Anterior corneal mosaic. Produced by applanation of the fluorescein stained cornea, viewed through a Goldmann tonometer head.*

Fig. 6. *Peripheral mosaic pattern.*

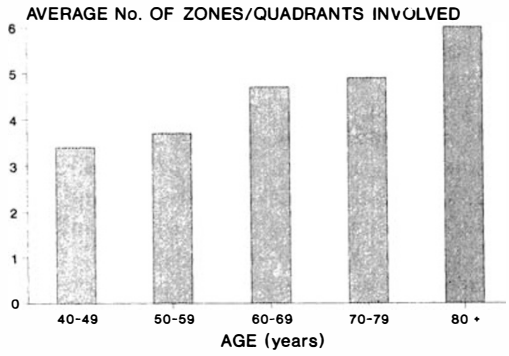

Fig. 8. *Age distribution of the extent of involvement in peripheral mosaic pattern.*

shagreen by virtue of their anterior stromal extension.

In addition to a PMP in the posterior stroma some cases with extensive involvement also showed a faint mosaic pattern at the level of Bowman's layer. This was apparently independent of the posterior changes and was less extensive in its encroachment centrally, the changes were confined to the peripheral zone.

The shape of the PMP changed as it was traced centrally. In the periphery the mosaic was composed of elongated irregularly arranged quadrilaterals with their long axes parallel to the limbus, separated by lucent zones. Centrally the pattern became more regular, resembling that seen in posterior crocodile shagreen; in no case did one type blend into the other.

No case of anterior crocodile shagreen was found in our study.

Discussion

The group under study was selected from a hospital population, and this accounts for the deviation from the general population age structure (Fig. 1).[1] Our population contains a greater proportion of elderly persons, with fewer below the age of 20 years as compared to the general population. Therefore, we have presented our data as absolute numbers, or

percentage prevalence within age bands (Fig. 2).

Vogt described posterior (mosaic) crocodile shagreen (Fig. 4)[2] as a central polygonal pattern in the deep stroma. Subsequently several other authors have reported cases which have invariably been bilateral.[3,4]

Other conditions having a similar mosaic appearance in the cornea have been described.

Anterior crocodile shagreen (Fig. 5) has been described in involutional, traumatic and juvenile forms, and in atrophy bulbi.[5] It is seen centrally, consisting of fibrous plaques in areas of interruption of Bowman's layer.

Cloudy dystrophy (François) is a bilateral condition thought to have autosomal dominant inheritance.[6] It is located centrally, consisting of diffuse ill-defined posterior stromal opacities which fade as they extend anteriorly. Posteriorly the apearance resembles posterior crocodile shagreen, but the pattern is poorly defined and it extends anteriorly rather than forming a discrete layer.

PMP (Fig. 6) has been described in association with corneal arcus,[4,5] consisting of numerous crisscrossing lines of relative clarity throughout the arcus. We have been unable to find any detailed description of the condition.

Anterior corneal mosaic (Fig. 7) is frequently noted when pressure is applied to the normal fluorescein stained cornea; experimental evidence points towards a subepithelial origin for the pattern.[7]

We found PMP to be age related, bilateral and symmetrical. Grayson[5] regarded the pattern to part of the corneal arcus, however we found the distribution of the early mosaic pattern not to mirror that of early arcus. Arcus is first identifiable at the inferior and superior corneal periphery,[5] only later is it seen to involve the temporal and nasal regions. PMP however is first seen infero-temporally, later extending supero-temporally and infero-nasally, less frequently involving the supero-nasal quadrant (Fig. 3). In many patients the two conditions coexisted but their distributions did not coincide, neither was there any relationship between the extent of the PMP and the density of the corneal arcus. The extent of the corneal involvement was also found to increase with age (Fig. 8), in both the number of quadrants involved and central zones affected.

Posterior crocodile shagreen differed from PMP in its distribution and symmetry. Posterior crocodile shagreen was often found to be asymmetrical and in one case unilateral, all previous reports describing it as bilateral.[3-5] Two cases of cloudy dystrophy were found, these were easily distinguished from posterior crocodile shagreen by their ill-defined mosaic pattern and their anterior extension. These morphological differences suggest separate conditions rather than variations of one disease, as has been suggested.[3]

PMP represents a distinct disease process, not related to corneal arcus, posterior crocodile shagreen or cloudy dystrophy. We consider it to be an age related corneal degeneration. The mosaic pattern common to all these conditions raises the possibility of an underlying structural arrangement in the cornea as the basis for its morphology. PMP, posterior crocodile shagreen and anterior crocodile shagreen are all located at or near boundaries between basement membranes (Descemet's and epithelial) and stroma, and cloudy dystrophy's mosaic pattern disappears as it is traced anteriorly away from the region of Descemet's membrane. The arrangement of the collagen bundles within these basement membranes or boundary regions may form a common basis for the mosaic pattern in these conditions. A histological description of posterior crocodile shagreen suggested the stromal opacification seen clinically is the result of the lack of orderly collagen arrangement.[4] The case described was almost certainly cloudy dystrophy as the opacification occupied the posterior two thirds of the cornea, and the authors made no distinction (apart from inheritance) between PMP and cloudy dystrophy. In addition polymorphic amyloid degeneration was present, and a history of previous surgical procedures complicated the histological interpretation. Until histological studies on PMP and posterior crocodile shagreen are available the true basis for the mosaic pattern remains obscure.

We would like to acknowlege the help of Mr. P. Bloor with the photography.

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[1] Estimated resident population at mid 1986 by single year of age and sex. In key population and vital statistics. *Series DH2 no.13 Table 1, OPCS.*

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