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Pseudo-capsular exfoliation in the Bantu of South Africa

I. Early or pre-granular clinical stage

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The first comprehensive clinical description of pseudo-capsular exfoliation of the lens was published by Vogt (1923, 1925) under the name "superficial exfoliation of the anterior capsule of the lens."

The appearance is described as a deposit of granular material likened to hoar frost or coarse white powder occurring on the anterior lens capsule, pupil margin, zonule, ciliary body, and persistent pupillary membrane, in the anterior chamber angle, and floating free in the aqueous. Pigmentary disturbances occur. Free pigment is often seen in the aqueous and a transient pigment cloud may be seen in the anterior chamber after dilatation of the pupil. Deposition of pigment on the trabeculae produces a dense band seen gonioscopically. Sampaolesi (1959) and Tosi (1964) considered that a pigmented line lying on the corneal side of Schwalbe's line was a very early sign of pseudo-exfoliation.

In the early stages the changes are confined to the anterior lens capsule and occur in two zones. A central zone, absent in 18 per cent. of cases (Tarkannen, 1962) corresponds to the pupillary aperture, it has a matt homogeneous appearance and may be missed unless the pupil is dilated, revealing a distinct edge with or without a few larger granular deposits. The peripheral zone, which is always present, is more distinct. It varies in width and may extend to the zonules. The outer margin has a radial pattern and the inner edge is sharply delineated. This deposit is granular. The intermediate area is clear but may be bridged by isolated deposits or by a continuous sheet. Gifford (1958) believed that the earliest change might be the formation of a continuous sheet of material which broke down in the intermediate area to produce the two distinct zones.

Pseudo-capsular exfoliation is a chronic disease and has a slow and insidious beginning which is difficult to recognize clinically. Early diagnosis depends upon adequate conditions of examination with pupils dilated, using a slit lamp in a darkened room. A further requirement is an examiner who is aware of, and interested in, the condition (Aasved, 1969).

Such a condition may be expected to be diagnosed more frequently at an advanced stage, and it is said to be a disease of old age, average 69·4 years in 418 cases (Tarkannen, 1962). Only eight cases under 50 years of age have been described (Trantas, 1929; Hørven, 1935; Gradle and Sugar, 1940; Gifford, 1957; Joannides, Katsourakis, and Velissaropoulos, 1961; Tarkannen, 1962); the two youngest subjects were aged 31 and 35 (Hørven and Hutchinson, 1967). Early cases of pseudocapsular exfoliation may be detected in mass surveys, but the pupils are seldom dilated; surveys to detect pseudocapsular exfoliation are generally confined to elderly age groups as in old peoples' homes where the disease is likely to be already advanced.

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It is surprising that pseudo-capsular exfoliation, even in the early stages, should spring suddenly into being as an obvious granular deposit. If deposited as granules of this size it should be detectable elsewhere and if it arises slowly *in situ*, either as minute granules or in a pre-granular stage, it should be detectable in younger subjects by careful observation.

This paper presents observations made during a glaucoma population study (Bartholomew, 1971). Pseudo-capsular exfoliation was especially looked for under favourable conditions and it is believed to have been detected at a very early stage. The clinical appearances are described and the evidence leading to this conclusion is presented and discussed.

Material and methods

In a population study undertaken among the Pondos, a tribe of the Cape Nguni-speaking Bantu of South Africa (Fig. 1), 2,584 persons over the age of 30 years were examined. The examination included biomicroscopy, using a Haag-Streit 900 slit lamp, in suitably darkened rooms, the pupils being dilated in all cases.

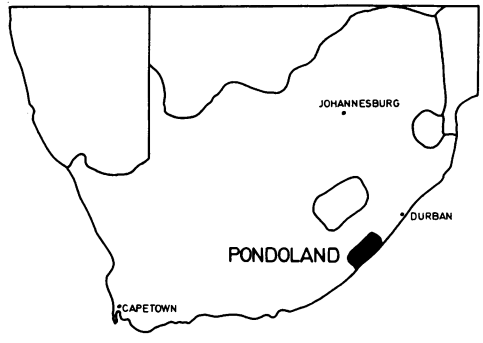

FIG. 1 *Map of South Africa, showing position of Pondoland*

The generally accepted changes of pseudo-capsular exfoliation are described as granular. The early changes detected in the survey are described as pre-granular.

Results

Table I shows the population sample for men and women in 10-year age groups. Fig. 2 (opposite) shows the percentage of granular and pre-granular psuedo-capsular exfoliation by 10-year age groups for men, women, and all cases together. Table II shows the total number of cases and compares the unilateral and bilateral occurrence of granular and pre-granular exfoliation. Table III (opposite) shows cases of particular interest.

Table I *Population sample by age and sex*

Age group (yrs)	*Male*	*Female*	*Total*
30—39	67	90	157
40—49	453	461	914
50—59	338	321	659
60—69	234	315	549
70+	155	150	305
Total	1,247	1,337	2,584

Table II *Analysis of stage of disease*

Side	*Unilateral*	*Bilateral*	*Totals*
Pre-granular	23	53	76
Granular	50	82	132
Total	73	135	208

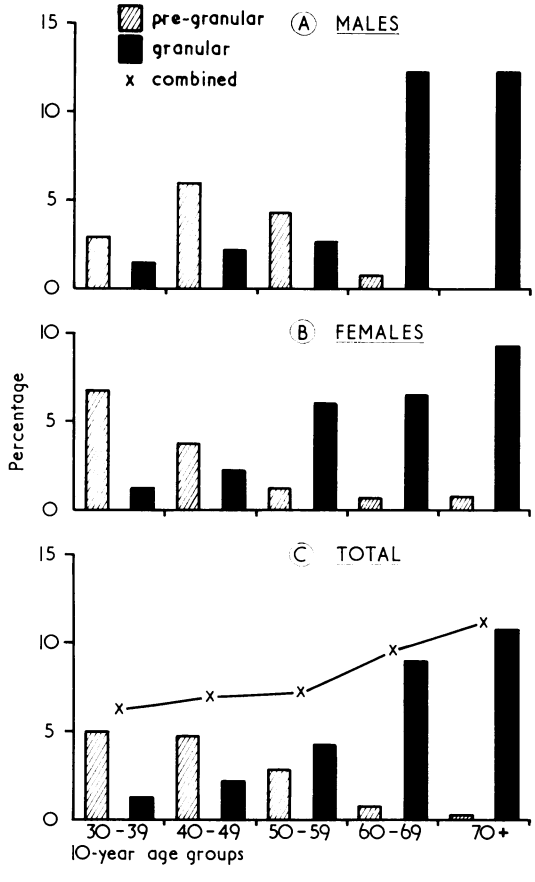

FIG. 2 *Percentage of granular and pre-granular pseudo-capsular exfoliation, by 10-year age groups*

Table III *Cases of particular interest*

Type of case	*No. of cases*
Granular and pre-granular in the same eye	1
Granular in one eye, pre-granular in the other	5
Pre-granular with a pigment cloud	4

CLINICAL APPEARANCE OF THE PRE-GRANULAR STAGE

A gradual progression could be traced from the very earliest to the full picture of pseudo-capsular exfoliation. It is convenient to describe this in stages (Figs 3, 4, 5, 6, overleaf).

In Stage 1 (Fig. 3), greyish radial non-granular striae are visible on the mid-third of the anterior capsular surface, entirely behind the iris. The striae are thin and slightly fusiform, vary a little in length, and are evenly distributed in all quadrants. They number some eighty in all.

The striae are difficult to see. They are most easily seen when a 0·5 mm. width slit beam is focused on the lens surface at an angle of 45° and the whole slit lamp is traversed from side to side. With diffuse illumination the striae tend to disappear and with static

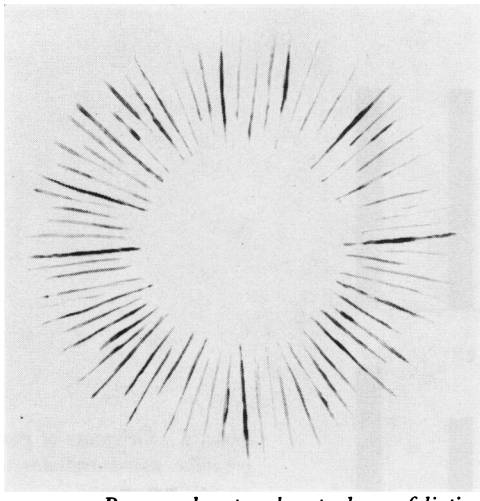

FIG. 3 *Pre-granular pseudo-capsular exfoliation. Stage* 1

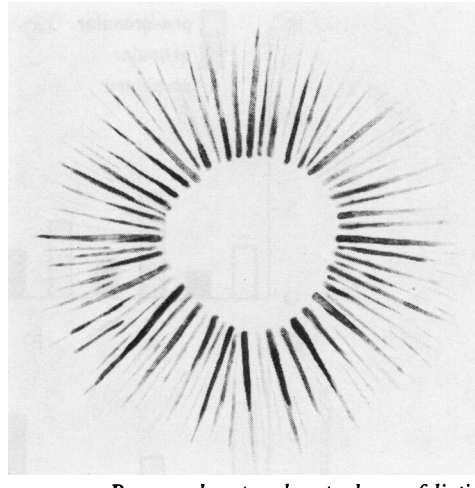

FIG. 4 *Pre-granular pseudo-capsular exfoliation. Stage* 2

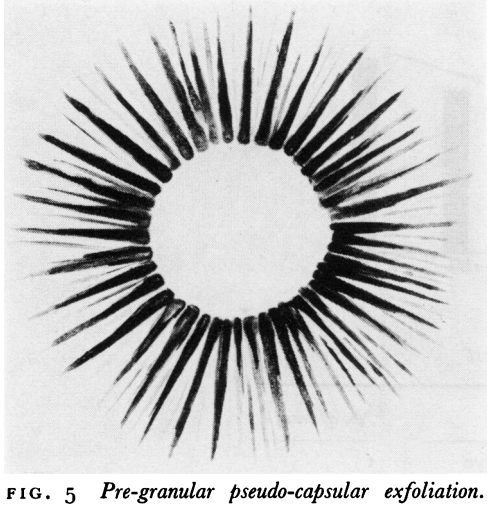

FIG. 5 *Pre-granular pseudo-capsular exfoliation. Stage* 3

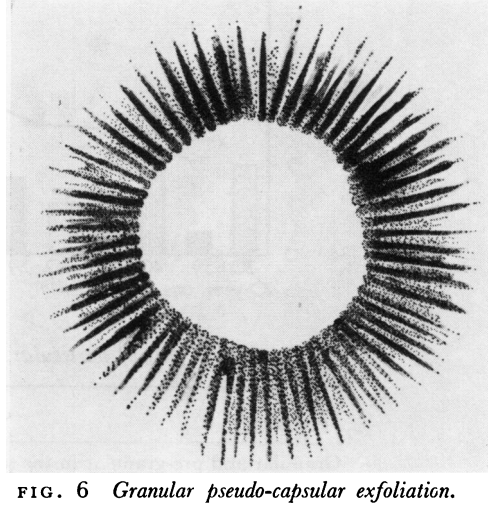

FIG. 6 *Granular pseudo-capsular exfoliation. Stage* 4

illumination the lines are easily missed. Only a segment of each stria can be seen in this way at any time.

On occasion the appearance of the striae is so faint as to be almost imagined, but it grades continuously into Stage 2 (Fig. 4) where they are more easily seen. They become slightly broader and thus lie closer together. The outer ends become a little longer while the inner ends become blunted and broader.

In Stage 3 (Fig. 5), the striae broaden and the blunted inner ends begin to touch one another, thus forming a continuous dentate line. No visible granular deposits occur (with 25 times magnification). With the addition of fine granular deposits, this stage merges into Stage 4 (Fig. 6) which becomes the generally accepted early stage of pseudo-capsular exfoliation.

No central deposit was noticed in Stages 1 or 2 but some cases in Stage 3 gave the

impression of a faint greyish matt deposit; lacking the visual contrast of the striae and their background this could not be confirmed.

Discussion

Evidence that this appearance was in fact an early stage of pseudo-capsular exfoliation came from a consideration of the morphology of the deposits and of certain individual cases, the prevalence of granular and pre-granular stages in the population sample, and their unilateral or bilateral occurrence. Other causes which might produce this appearance were considered.

The morphological resemblance between pre-granular and granular pseudo-capsular exfoliation is strong. The radial pattern is similar, each having some eighty lines. This equals the number of structural folds on the posterior iris surface which may play some part in the production of the radial pattern.

Movement of the iris and pupillary margin against the capsule is responsible for the width of the outer band. In Stage 1, the striae are too peripheral to be affected by the iris and the inner ends of the striae remain pointed. As the striae lengthen centrally they come within range of the iris effect and the pointed inner ends become blunted (Stages 2 and 3).

A 40-year-old man presented one eye with the appearance shown in Fig. 7. One quadrant showed definite granular pseudo-capsular exfoliation which merged in the opposite quadrant to the radial striae of Stage 1. His other eye showed striae in Stage 1. This case was unusual as progression in an eye is usually radially symmetrical in the absence of iris abnormalities. This case provided a major clue in confirming that the radial striae were due to pseudo-capsular exfoliation.

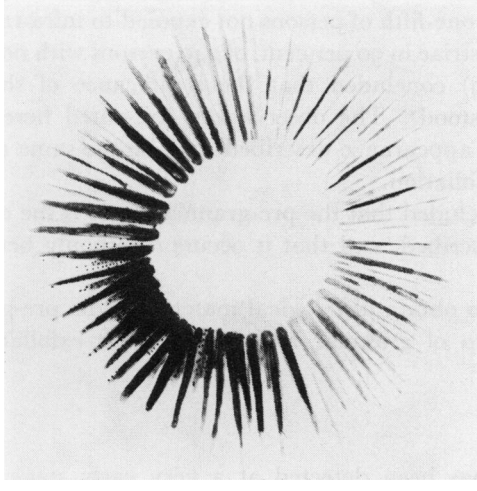

FIG. 7 *40-year-old male. Pre-granular and granular pseudo-capsular exfoliation in the same eye*

The association of granular pseudo-capsular exfoliation in one eye with pre-granular pseudo-capsular exfoliation in the other was seen in five more cases. This may be due to chance, but is readily explained by the fact that pseudo-capsular exfoliation is commonly bilateral.

Four patients with pre-granular pseudo-capsular exfoliation produced a pigment cloud in the anterior chamber after dilatation of the pupil. This is a characteristic feature

of granular pseudo-capsular exfoliation. Pigmentary disturbances are a common senile change; their occurrence in such young cases favours a generalized ischaemic process rather than a localized lens abnormality as a cause for pseudo-capsular exfoliation.

These ten cases strongly suggest on clinical grounds that the pre-granular appearance is an early stage of pseudo-capsular exfoliation.

The prevalence of the pre-granular stage in the sample of the population (Table I and Fig. 2) is highest in the 30 to 39-year age group (5·1 per cent.) and falls with increasing age to the lowest level in the over 70-year age group, in which only one case was seen (0·3 per cent.).

The prevalence of granular pseudo-capsular exfoliation is lowest in the 30 to 39-year age group (1·3 per cent.) and rises steadily through the age groups to its highest level in the over 70-year age group (10·9 per cent.). The falling prevalence of pre-granular pseudo-capsular exfoliation can be explained if these cases steadily progress and become granular until, in the over 70-year age group, nearly all have become granular.

Granular pseudo-capsular exfoliation is unilateral in 45 to 48 per cent of cases (Gifford, 1957; Tarkannen, 1962). In the population studied, 132 cases were seen, fifty (38 per cent.) unilateral, and 82 (62 per cent.) bilateral, 76 pre-granular cases were seen: 23 (31 per cent.) were unilateral and 53 (69 per cent.) were bilateral. This again suggests that the two groups are stages in the same entity.

Pseudo-capsular exfoliation may be confused with true exfoliation of the lens capsule, remnants of exudation following iritis, iris atrophy, and heterochromic cyclitis. None of these conditions were seen in any of these cases. Retro-iridal pigmented lines and stripes (Berliner, 1949), which have a radial pattern on the anterior lens surface, are thin pigmented lines and are readily recognizable. Superficial anterior grey radial striae (Rehsteiner and Schläpfer, 1930) in description and illustration very closely resemble the appearance described here, although localized as being subcapsular (Vogt, 1931). They were seen in over half of 59 glass blowers and one-fifth of persons not exposed to infra-red irradiation. Abramowicz (1933) reported these striae in 90 per cent. of 240 persons with no occupational exposure to heat. Berliner (1949) concluded that the significance of these radiating structures was not entirely understood. The observations presented here suggest that anterior grey radial striae and the appearance described may be the same and that both may be due to pseudo-capsular exfoliation.

From these observations it is concluded that the pre-granular stage is the earliest sign of pseudo-capsular exfoliation yet described, and that it occurs commonly between 30 and 50 years.

Further studies are in progress to obtain histological material of the pre-granular stage and to investigate the relationship of granular and pre-granular exfoliation to intra-ocular pressure.

Summary

(1) Pseudo-capsular exfoliation has been detected at a very early stage, termed pre-granular, during a population study among Pondos of South Africa.

(2) It occurred in 5·1 per cent. in persons between the ages of 30 and 39 years of age and fell to 0·3 per cent. in persons over the age of 70 years.

(3) The clinical appearances are described. Morphological, clinical, and statistical evidence that this appearance is pseudocapsular exfoliation is presented and discussed.

(4) This appearance may be the same as anterior grey radial striae.

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