Pigmentary dispersion

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SUMMARY A long-term study of primary pigmentary dispersion has shown that the condition is commoner in males than females and appears most frequently in the third decade. After 10 years there may be a significant reduction in the amount of pigment deposited on the cornea, and the condition of several patients receiving treatment for glaucoma has remained under control when treatment has been stopped.

Primary pigmentary dispersion is seen predominantly in young myopic males and is often associated with open-angle glaucoma. Recent pathological studies^{1,2} have shown fragmentation of the neuroectodermal pigment of the iris and hyperplasia of the dilator muscle fibres. Mechanical rubbing of the pigment epithelium against bundles of radially disposed zonular fibres may be a factor in causing a release of pigment.³ A mesodermal defect in the angle has been postulated to explain some cases of glaucoma in which pigment release is minimal. In other patients there appears to be a direct relationship between the degree of pigment dispersion and the presence of glaucoma.

The long-term behaviour of the pigment in pigmentary dispersion has not been studied in detail. It is possible the pigment dispersion occurs steadily over a lifetime, or the release could be self-limiting. A reduction of pigment in the angle has been reported in 1 patient over some time,⁴ and in another an improvement in intraocular pressure occurred.³ The future management of a patient with pigmentary dispersion will be influenced by what happens to the release of iris pigment.

In this study a group of patients with pigmentary dispersion have been observed for some years to evaluate the role of pigment in the production of glaucoma and to determine what happens to the pigment and glaucoma.

Material and methods

Forty-five patients with primary pigmentary dispersion were available for review, consisting of 29 males and 16 females. The presence of pigment on the cornea and in the angle and the extent of iris transillumination were recorded. A diagnosis of

Table 1 *Age at diagnosis*

Age	20-29	30-39	40-49	50-59	60-69	Total
Male	2	11	9	5	2	29
Female	1	4	3	7	1	16

glaucoma was made when an individual had an intraocular pressure over 30 mmHg, and over half this group had glaucomatous cupping and field loss. Of 20 males and 7 females with glaucoma, cupping and field loss were found in 15.

The role of pigment in the production of glaucoma was investigated by comparing the degree of pigment dispersion in both eyes in patients with unilateral glaucoma. In 9 patients with unilateral glaucoma there was a definite increase in the severity of the pigment dispersion in the eye with glaucoma. In 6 patients a difference could not be seen clearly. In no patient was the pigment dispersion less in the eye with glaucoma.

Results

A change in pigment dispersion with time was assessed by comparing the findings at the initial examination with those of the most recent follow-up. The great majority of patients who were followed up for 5 years showed no change. Between 5 and 10 years 3 of 8 patients reviewed showed a reduction in pigment and after 10 years 5 of 7 patients showed a reduction. Only 1 patient showed an increase in

Table 2 *Changes in pigment dispersion*

Follow-up in years	0-5	5-10	10-15
Pigment less	0	3	5
Pigment same	8	4	2
Pigment more	0	1	0

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Table 3 *Response to stopping glaucoma treatment*

<i>Duration of treatment</i>	<i>0-4</i>	<i>5-9</i>	<i>10-14</i>	<i>Pigment same</i>	<i>Pigment less</i>
Intraocular pressure raised	1	0	2	1	1
Intraocular pressure normal	1	4	0	0	4

pigment dispersion. This individual when first seen had heavily pigmented angles but no corneal pigment was recorded. The intraocular pressure was raised in the right eye. When the patient was re-examined 5 years later corneal pigment was present in both eyes, more in the right than the left. At this time the intraocular pressure was even higher in the right eye, and early cupping and a field defect had developed.

A group of 8 patients on treatment for glaucoma had their medication stopped and the intraocular pressures reassessed. For most of them the glaucoma was not severe, as only 3 had a field defect.

Five patients did not show a rise in pressure above 30 mmHg, and treatment was discontinued. Three of the 5 patients had a reduction in pigment dispersion, 1 showed no change, and for the fifth patient the pigment change was not recorded. Three patients developed a rise in pressure after stopping treatment, and 1 of these patients showed a reduction in pigment dispersion. No patient whose intraocular pressure stayed below 30 mmHg immediately after stopping treatment had to restart again after a follow-up varying from several months to several years.

CASE REPORT

The following case report summarises these observations. A 32-year-old white male was first seen in 1971 for assessment of uniocular pigmentary glaucoma. At that time treatment consisted of pilocarpine 4% daily in the right eye and 4 times daily in the left together with epifrin twice daily in the left eye and acetazolamide 250 mg twice daily. The clinical findings showed a pigment dispersion which was mild in the right and marked in the left. In addition the left disc was cupped and there was an extensive inferior Bjerrum scotoma. The initial intraocular pressure readings were 15 mmHg in the right eye and 25 in the left. Treatment was stopped in the right eye and switched to ecothiopate iodide 25% and epifrin 1%, both twice daily in the left eye. Over a period of a year the intraocular pressure fell below 20 mmHg in the left eye, and in 1975 the frequency and strength of the ecothiopate iodide was reduced and in 1976 was discontinued. In 1978 epifrin was stopped, and for a year the

intraocular pressure has remained at normal levels. Over this period a moderate deposit of pigment on the posterior surface of the left cornea has disappeared. The angle still shows excessive pigmentation, and the field defect has not changed.

Discussion

The age and sex distribution in this series confirms that pigmentary dispersion is commoner in males than females. The initial diagnosis is usually made in the third and fourth decades. Since most of these patients come under regular observation because of myopia, it seems likely that pigment dispersion begins most frequently in the third decade.

The patients with glaucoma were evenly divided between those with intraocular pressure elevation alone and those with cupping and field loss. There was a significant increase in the severity of pigmentary dispersion in the involved eye of the majority of patients with uniocular glaucoma, which suggests that the pigment is a significant factor in the development of glaucoma. Another possibility is that the excessive pigment dispersion in the eyes with glaucoma reflects the presence of other factors responsible for glaucoma but is not itself the primary cause. Since in 6 of the patients with uniocular glaucoma the pigment dispersion appeared to be similar in both eyes, there must have been some additional factor responsible.

The presence of pigment represents a balance between the release of fresh pigment and the destruction of old pigment. The long-term follow-up of these patients indicates a significant reduction in the extent of pigmentary dispersion after a period of 10 years. This suggests a gradual reduction in the production of fresh pigment or, less likely, an increased rate of destruction. Pigment which has been incorporated into endothelial cells (for example, after a traumatic pigment dispersion) is often very slow to disappear and may remain visible long after its active release has stopped. It is possible, therefore, that primary pigment dispersion takes place over a relatively short period even though it can still be seen many years later. Only 1 patient showed an increase in pigment, and this developed over a 5-year period, after the diagnosis was made. For all the other patients it appeared that the bulk of pigment dispersion had taken place by the time the condition was identified.

Haloes after exercise are associated with a heavy release of pigment and are an indication of active dispersion. These symptoms occur early in the course and stop after several years. Pigment circulating in the anterior chamber, and a marked

release after pupil dilatation, are additional signs of active pigment dispersion, which gradually disappear as the patient grows older. It is difficult to be precise, but the evidence suggests that most pigment dispersion takes place over a period of about 5 years.

Five of 8 patients on treatment for glaucoma failed to show a pressure increase when the treatment was stopped and 3 of these patients showed a decrease in pigment dispersion. This is a further indication that for some people pigment is responsible for glaucoma, and for many of these individuals both the pigment release and the glaucoma are self-limiting. One patient who had a reduction in corneal pigment showed a persistent rise in pressure when treatment was stopped. It is possible that factors other than pigment were responsible for the glaucoma, or it may be that pigment remaining in the angle was sufficient to produce an obstruction to outflow. Patients who show a reduction in corneal pigment still have excessive pigmentation of the trabecular meshwork on gonioscopy.

The management of patients with pigmentary dispersion needs to be reassessed in view of the evidence that this condition is self-limiting. Patients seen for the first time with no evidence of glaucoma should be examined at regular intervals for at least 5 years because of the possibility that active pigment dispersion is still taking place which might cause a rise in pressure. The great majority of patients do not show evidence of a progressive increase in pigment dispersion, and only 3 had a rise in pressure

develop within 5 years of making the diagnosis. After 10 years there is increasing evidence of a reduction in pigment dispersion, and no patients showed a rise in pressure after this interval.

Patients who require treatment for glaucoma should be encouraged to persist with medical therapy because of the chances of remission as the pigment dispersion slows down. This is often possible with the uniocular and less severe bilateral cases. However, the younger patients who require miotics are usually unable to tolerate the side effects. When medical treatment has been employed for several years and pressure control is satisfactory, medications can be reduced and often stopped altogether. Many of these eyes show a reduction in pigment dispersion.

Conclusion. After a period of several years patients with pigment dispersion syndrome may show a reduction in the amount of dispersed pigment, and glaucoma, if present, may go into remission.

References

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