SFO COMMUNICATION

Pseudoexfoliation syndrome in Congolese patients

Le syndrome de pseudoexfoliation capsulaire chez les patients congolais

D. Kaimbo Wa Kaimbo

Département d’ophtalmologie, université de Kinshasa, BP 16540, Kinshasa 1, RD Congo

Received 2 March 2011; accepted 28 April 2011
Available online 19 October 2011

KEYWORDS
Pseudoexfoliation; Frequency; Cataract; Glaucoma; DR Congo; Central Africa

Summary
Purpose. — To determine the frequency of pseudoexfoliation syndrome (PEX) in Congolese patients and its association with cataract and glaucoma.
Methods. — We performed a cross-sectional and descriptive analysis of the data from patients diagnosed with PEX. Data were collected between February 2005 and June 2008 in a general practice of ophthalmology in Kinshasa. Patients aged 50 or above who attended the general practice of ophthalmology were included in the study. Each patient underwent complete ophthalmic evaluation, including visual acuity testing, slit-lamp biomicroscopy, applanation tonometry, gonioscopy, and ophthalmoscopy. The diagnosis of PEX was based on presence of typical pseudoexfoliation material on the anterior lens surface and/or the pupillary margin in either or both eyes.
Results. — Of 2142 patients seen during the study period, 37 (59 eyes) had PEX in either eye, for a frequency of 1.73%. The mean (±SD) age of the patients with PEX was 70.40 years ± 8 (range: 57–87 years). The frequency of PEX had a tendency to increase with age: 0.50% of patients aged 50–59 years had PEX, whereas 7.29% of those aged more or equal to 80 years were affected (P < 0.0001), with a female predominance (2.18% of women versus 1.22% of men, P = 0.035). PEX was bilateral in 22 (59.46%) of 37 patients (74.58% of eyes). Bilateral PEX was found more often in females (16 patients over 22, 72.72%) than in males (six patients over 15, 40%) (P = 0.05). PEX was significantly associated with cataract (P = 0.002) and glaucoma (P < 0.001).
Conclusion. — Despite the limitations inherent in a clinic-based study, this investigation provides an indication of the frequency of PEX in Central Africa. This shows that Congolese patients have a low frequency of PEX (1.73%), inferior to that of black people in South Africa.

© 2012 Elsevier Masson SAS. Tous droits réservés.
E-mail address: dieudonne_kaimbo@yahoo.com.
Introduction

Pseudoexfoliation syndrome (PEX) is an age-related condition characterized by a progressive and pathologic accumulation of polymorphic fibrillar material, including elastic fibrils, in the anterior segment of the eye [1,2]. It was first described by a Finnish ophthalmologist, Lindberg, in 1917 [3] and occurs in most races and countries; its prevalence is known to be high and common in many countries [4–7], particularly in Norway and in the Eastern European and Mediterranean countries. It is rare in African Negroes and in Negroes in the West Indies [8], and has not been reported in Negroes elsewhere outside South Africa [9–11].

Epidemiologic studies of PEX in populations of Central Africa are still lacking and there are no available data of PEX for Congolese people, accounting for about 60 million people. The aim of this clinic-based study was to estimate the frequency of PEX and assess whether PEX is significantly associated with cataract and glaucoma in Congolese patients.

Patients and methods

This study was a cross-sectional and descriptive study conducted in an outpatient clinic, a general practice of ophthalmology in Kinshasa, between February 2005 and June 2008. All the patients who were 50 years of age or older presenting to the general practice of ophthalmology were included in this study. Patients underwent a complete ophthalmic evaluation, included best corrected visual acuity, slit lamp biomicroscopy, Goldmann applanation tonometry, and gonioscopy. Repeat slit lamp examination after dilation was performed to look for pseudoexfoliative material. The study was conducted in accordance with the Declaration of Helsinki and verbal informed consent was obtained from all patients.

The diagnosis of PEX was based on slit lamp biomicroscopy by the presence of typical white, fluty material in the pupillary margin before dilation, on the anterior lens surface after dilation [1,12]. Phakic eyes with nuclear sclerosis, posterior subcapsular cataract, cortical or combination cataract were considered cataractous. Eyes with IOP greater than 21 mmHg with glaucomatous optic disc damage and/or any characteristic visual field defect and eyes with filtration surgery or use of antiglaucoma eyedrops were considered glaucomatous.

Statistical analysis

Chi² tests or Fisher exact test, as appropriate, were calculated to test for significance of a relation between dichotomous variables. Student’s t test was used to compare between means or ordered variables. Characteristics of
patients with PEX were compared with those without PEX. Analyses were based on individuals, not per eye. A P-value of less or equal to 0.05 was considered significant.

Results

We studied 2142 patients who were seen during the study period. Age ranged from 50 to 98 years (mean ± SD = 61.78 ± 9). One thousand two hundred and thirty-two (57.52%) patients were men and 910 (42.48%) were women. Table 1 shows the frequency of PEX among patients aged more or equal to 50 years, by sex, age and at least one eye affected. PEX was found in either eye of 37 (59 eyes) patients, giving a frequency of 1.73%. The mean ± SD age of patients with PEX was 70.43 years ± 8 while the mean age of patients without PEX was 61.78 years ± 9, the difference being significant (P < 0.0001). We found greater rates of frequency with increasing age: 0.50% of patients aged 50–59 years had PEX, whereas 7.29% of those aged more or equal to 80 years were affected. The difference between all age groups was significant (P < 0.0001). Women were more frequently diagnosed with PEX in at least one eye than men (2.18% versus 1.22% of men, P = 0.035).

We found unilateral PEX in 15 (40.54%) patients (in eight right eyes and seven left eyes) and bilateral PEX in 22 (59.46%) patients. The mean ages of patients with uni- and bilateral PEX were 70.80 years ± 7.96 and 70.57 years ± 8.35, respectively (P = 0.93). Bilateral PEX was found more in female (16 out of 22 patients, 72.72%) than in men (six out of 15 patients, 40%) (P = 0.05).

The frequency of cataract between the PEX and the non-PEX patients is shown in Table 2. Twenty (20 out of 37 patients; 54.05%) patients with PEX were found to have cataract, while in the non-PEX patients the frequency of cataract was found in 647 (647 out of 2105 patients; 30.74%) (P = 0.002). The frequency of glaucoma was 45.94% in patients with PEX and 16.48% in patients without PEX (P < 0.001) (Table 2).

Discussion

PEX was first described by the Finnish ophthalmologist Lindberg, in 1917, when he noticed bluish gray flakes located

<table>
<thead>
<tr>
<th>Variable</th>
<th>PEX n (%)</th>
<th>Non PEX n</th>
<th>Total n</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>37 (1.73)</td>
<td>2105</td>
<td>2142</td>
<td></td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
<td></td>
<td>0.035</td>
</tr>
<tr>
<td>Female</td>
<td>22</td>
<td>890</td>
<td>912</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>15</td>
<td>1215</td>
<td>1230</td>
<td></td>
</tr>
<tr>
<td>Age, years</td>
<td></td>
<td></td>
<td></td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>Mean</td>
<td>70.4</td>
<td>61.78</td>
<td>61.78</td>
<td></td>
</tr>
<tr>
<td>SD</td>
<td>8.09</td>
<td>8.9</td>
<td>8.9</td>
<td></td>
</tr>
<tr>
<td>Range</td>
<td>57–87</td>
<td>50–98</td>
<td>50–98</td>
<td></td>
</tr>
<tr>
<td>Age, years</td>
<td></td>
<td></td>
<td></td>
<td>0.0004</td>
</tr>
<tr>
<td>Females</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>50–59</td>
<td>3 (0.70)</td>
<td>426</td>
<td>429</td>
<td></td>
</tr>
<tr>
<td>60–69</td>
<td>9 (3.18)</td>
<td>274</td>
<td>283</td>
<td></td>
</tr>
<tr>
<td>70–79</td>
<td>5 (3.45)</td>
<td>140</td>
<td>145</td>
<td></td>
</tr>
<tr>
<td>≥ 80</td>
<td>5 (9.43)</td>
<td>48</td>
<td>53</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>22 (2.42)</td>
<td>888</td>
<td>910</td>
<td></td>
</tr>
<tr>
<td>Males</td>
<td></td>
<td></td>
<td></td>
<td>0.002</td>
</tr>
<tr>
<td>50–59</td>
<td>2 (0.36)</td>
<td>561</td>
<td>563</td>
<td></td>
</tr>
<tr>
<td>60–69</td>
<td>13 (2.99)</td>
<td>422</td>
<td>435</td>
<td></td>
</tr>
<tr>
<td>70–79</td>
<td>7 (3.66)</td>
<td>184</td>
<td>191</td>
<td></td>
</tr>
<tr>
<td>≥ 80</td>
<td>2 (4.65)</td>
<td>41</td>
<td>43</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>15 (1.22)</td>
<td>1232</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Males and females combined</td>
<td></td>
<td></td>
<td></td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>50–59</td>
<td>5 (0.50)</td>
<td>987</td>
<td>992</td>
<td></td>
</tr>
<tr>
<td>60–69</td>
<td>13 (1.81)</td>
<td>705</td>
<td>718</td>
<td></td>
</tr>
<tr>
<td>70–79</td>
<td>12 (3.57)</td>
<td>324</td>
<td>336</td>
<td></td>
</tr>
<tr>
<td>≥ 80</td>
<td>7 (7.29)</td>
<td>89</td>
<td>96</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>37 (1.73)</td>
<td>2105</td>
<td>2142</td>
<td></td>
</tr>
</tbody>
</table>

PEX: pseudoexfoliation.
on the pupillary border in a number of his patients with glaucoma [13–15]. PEX has been reported from all over the world in almost all races [4,5,16,17].

In Caucasian, it is generally thought to be most common in Scandinavian and northern European countries, where it appears in 18 to 22% in people 60 years of age and older [1,2,4,6,7,13,15,18–24]. In the Framingham study, it was found in 5% of patients aged 75 to 85 [25,26]. In southern Louisiana, its prevalence was found to be 2% in whites over the age of 50 [27]. In Australia, the prevalence of PEX in samples roughly 50 years and older has been found to be around 2% [1,28]. There is also a high prevalence of 35% among the Navajo Indians and in one study, it was found in no Eskimos over age 60 [5,25]. In Asian, prevalences of PEX ranged from 0.4 to 7.4% [1,29–38]. A higher prevalence of 9.6% was found in a study from Iran [39].

In Blacks, the prevalence of PEX has previously been reported to be low among African Americans. In southern Louisiana, the prevalence was found 0.3% in Blacks within a group of glaucoma patients over the age of 50, [27] compared with 2.7% among whites. In Negroes from Africa, the condition has been noted to be rare, except for the Bantu tribes of South Africa, where the prevalence was reported to be 11 [40] 12.5–16.1% for those 60 or older [40]. To the best of the author’s knowledge, there are no previous reports on the prevalence or characteristics of PEX in blacks for Central Africa. The frequency of 1.73% reported in our study in Congolese patients more or equal to 50 years is lower than that reported in black Bantu indigenous population from South Africa [11,40]. Our frequency is also lower than the prevalence reported from Mediterranean and Scandinavian countries in Caucasian [1,2,4,6,7,13,15,18–24].

Variations and differences in PEX prevalence in studies have not been well explained [41,42] and could be explained by differences related to examination techniques and diagnostic abilities [43] (methodology, age of population, definition of PEX, study design); they may be also related to genetic, geographical or racial variability [24,44].

As expected and reported in prior studies in Caucasian and Asian [3,6,19,26,28,31,32,45–50], the frequency of PEX, in our study, increased from 0.50% in patients between 50 and 59 years of age to 7.29% in those more or equal to 80 years. Our study confirms the idea that the prevalence of PEX increases with age. Although the reason for this age-related increase is unknown, it has been speculated that the changes in gene expression that occur with age may be responsible [51].

There are conflicting reports of gender differences in the prevalence of PEX [6,17,19,22,23,26,31,46,52]. Whereas some studies [1,6,16,23,26,53] report higher rates in women as found in our study, other studies [22,31,54] report higher frequencies in men and still others show no differences between the two sexes [24,28,33,37,40,46,52].

Of 37 patients with PEX in our study, we found that 15 had unilateral PEX and 22 had bilateral PEX. A proportion of unilateral cases is known to convert to bilateral over time [55,56]. Actually, the syndrome is rather bilateral with asymmetric presentation, as PEX fibers have been observed on electron microscopy in the conjunctiva [57] and orbital tissues [58] of the clinically uninvolved fellow eyes. Also, immunohistochemical and ultrastructural findings typical of PEX fibers have been revealed in the iris of all unaffected fellow eyes [59].

In this study, the frequency of cataract in PEX patients was higher (54.05%) than the frequency in patients without PEX (30.74%). Cataract has been reported in previous studies as more common in patients with PEX, as it is zonular weakness, compared with the general population [15,21,60]. In our study, we found glaucoma in 45.94% of patients with PEX, as opposed to 16.48% of those without PEX. The association of PEX syndrome to glaucoma is well-established [22,61–63].

PEX is considered to be the most common identifiable cause of open-angle glaucoma worldwide [64]. However, the majority of subjects with PEX do not have glaucoma [65]. The percentage of PEX patients with glaucoma is different for every population. Overall, studies indicate that about 40% PEX patients will develop glaucoma [66]. The reported prevalence of PEX among patients with OAG also shows geographic and racial variation [67], with 26% in Denmark [67,68], 75% in Sweden [69], 60% in Norway [70], 46.9% in the Mediterranean area of Turkey [71], and 44.5% in the Northwest of Spain [72], compared with reports of 1% [73], 3% [74], 6% [75], and 12% [76] in the United States. The prevalence for the Bantu tribes of South Africa was reported to be over 20% [10], which is also higher that the reported frequency of 3% of exfoliative glaucoma in a previous study in Congolese patients with glaucoma [77].

In conclusion, despite the limitations inherent in a clinic-based study, our study provides an indication (information) of the frequency of PEX in Central Africa. This shows that Congolese patients have a low frequency of PEX (1.73%), inferior to that of black people in South Africa (12.1 to 16%). This study confirms as in other races that PEX is an age-related condition and is associated with cataract and glaucoma.

### Table 2 Frequency of cataract and glaucoma in patients with and without PEX.

<table>
<thead>
<tr>
<th>Variable</th>
<th>PEXn</th>
<th>Non PEXn</th>
<th>Totaln</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cataract</strong></td>
<td></td>
<td></td>
<td></td>
<td>0.002</td>
</tr>
<tr>
<td>Yes</td>
<td>20</td>
<td>647</td>
<td>667</td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>17</td>
<td>1458</td>
<td>1475</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>37</td>
<td>2105</td>
<td>2142</td>
<td></td>
</tr>
<tr>
<td><strong>Glaucoma</strong></td>
<td></td>
<td></td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Yes</td>
<td>17</td>
<td>347</td>
<td>364</td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>20</td>
<td>1758</td>
<td>1778</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>37</td>
<td>2105</td>
<td>2142</td>
<td></td>
</tr>
</tbody>
</table>

PEX: pseudoexfoliation.

### Disclosure of interest

The author declares that he has no conflicts of interest concerning this article.
References