

Syndroma: Glaucoma Pigmentatum Primarium – Pigmented Glaucoma

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ABSTRACT

In this study pigmented glaucoma was diagnosed in 29 out of 4350 patients suffering from glaucoma. In clinical analysis of those cases main attention was put on Krukenberg spindle with emphasis on the process and reason of its development. In all cases of Krukenberg spindle moderate myopia and myopic astigmatism were present. The main axis of the spindle lies in the principal meridian of corneal astigmatism. The principal meridian of the anterior cornea surface corresponds with the axis of the principal meridian of the posterior corneal surface. The higher degree of astigmatism more pronounced Krukenberg spindle. This corresponds to a number of cases in which the spindle is hardly recognisable in the astigmatism, of only ± 0.25 to ± 0.50 Dcyl. The axis of meridian corresponds to direct astigmatism (from 110° to 70°). The authors conclude that there is no pigmented Krukenberg spindle without myopic astigmatism. It is for this reason and because of the inheritance factor that this type of glaucoma should be considered primary glaucoma and a separate clinical entity.

Introduction

The pathognomonic sign of pigmented glaucoma is Krukenberg spindle – a spindle shaped deposition of pigment on the corneal endothelium. In 1973. Vojniković referred to the pattern of formation of pigmented spindle developing because of the myopic astigmatism. More precisely, pigmented spindle is being formed on the endothelial edge of the myopic astigmatism positioned between 110° and 70° (Figures 1,2). Therefore the pigmented spin-

dle is being formed only in the presence myopic astigmatism. The higher myopic astigmatism, more marked and pigmented Krukenberg spindle. This can be explained by a steeper meridian of astigmatism that causes faster deposition of pigment on the spindle edge^{1,2}. The anterior corneal surface astigmatism corresponds to the posterior corneal surface astigmatism. This form of glaucoma is associated with moderate myopia, myopic astigmatism and anterior chamber malformation (gonyodysgenesis). Since ante-

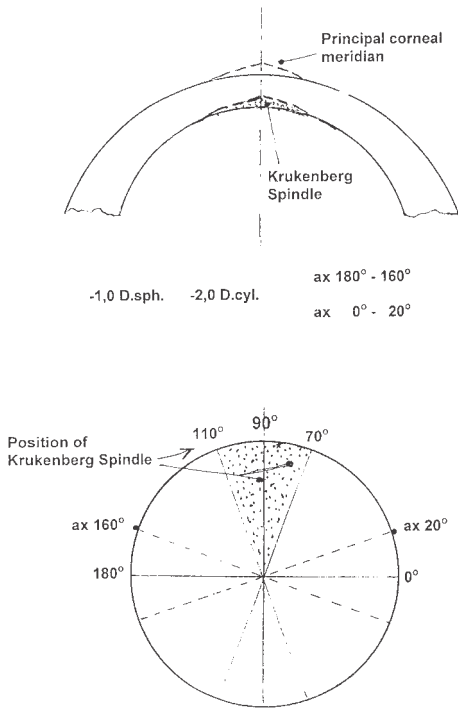


Fig. 1. The position of the principal corneal meridian defines the Krukenberg spindle formation in pigmented glaucoma – glaucoma pigmentatum (by Vojniković)

rior chamber is deeper than usual, acute attack does not occur even in the presence of very high intraocular pressure^{3,4}. As the process is associated with the pigment formation and dissipation as secondary changes, the appropriate term would be pigmented glaucoma – glaucoma pigmentatum instead of pigmentary glaucoma – glaucoma pigmentosum⁵. The atrophic changes of iris stroma, myopia and astigmatism as well as endothelial degeneration of the trabeculum are considered to be primary processes. This phenomenon is more frequent in women than in men, emphasizing the importance of the sex-linked inheritance. This type of glaucoma is highly resistant to medical and surgical treatment.

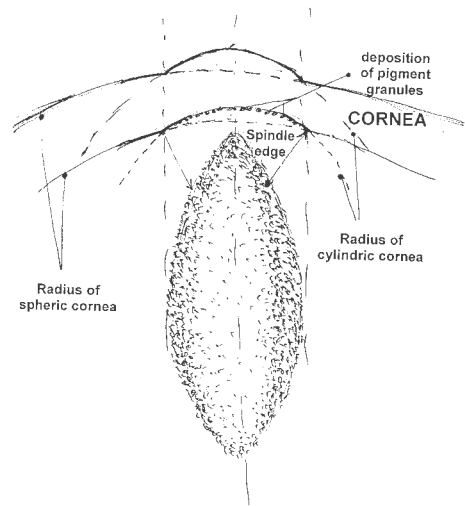


Fig. 2. Configuration of Krukenberg spindle

Sugar does not recognize Krukenberg spindle as a pathognomonic sign. He does not consider it either a unique nosologic group nor a separate clinical entity (especially not from the aspects of genetic determination and inheritance), but a broader group of pigment dispersion symptoms in the anterior segment.

Patients and Methods

We conducted a retrospective study among a total of 4350 glaucoma patients, 29 of them suffering from diagnosed pigmented glaucoma. All the patients were adults (age range from 38 to 72), 18 females and 11 males. All of them were already taking antiglaucoma medications. Complete ophthalmologic examinations were performed including biomicroscopy, gonioscopy with Goldmann triple-mirror goniolens, tonometry with Goldmann applanation tonometry, tonography with Schiötz indentation tonometry, and visual field examination with Goldmann perimetry. The measurement of corneal

astigmatism and examination of the eye refraction was performed using autorefractometer.

Results and Discussion

The values of myopic astigmatism are shown in Table 1. The most frequent ones were low, ranging from -0.25 Dcyl to -0.50 Dcyl in 6 cases, -0.75 Dcyl to -1.0 Dcyl in 10 cases, -1.25 Dcyl to -2.0 Dcyl in 9 cases, -2.25 Dcyl to -3.0 Dcyl in 3 cases and above -3.0 Dcyl in only 1 case. The same was with myopia. The most frequent finding was low myopia of -0,25 to -2,0 Dsph in 22 cases. Five patients had myopia of -2,25 to -4,0 Dsph (Table 2). The principal corneal meridian was most frequently positioned between 90° and 70°, mostly in the direct position of the 90° astigmatism (Table 3).

In pigmented glaucoma the anterior chamber is deeper (more than 3.0 mm), there is obligatory moderate myopia and

myopic astigmatism and sex-linked pattern of inheritance^{6,7}. For all those reasons the authors consider pigmented glaucoma a separate clinical entity of primary congenital glaucoma and not the secondary one. The authors consider the term pigmented glaucoma – glaucoma pigmentatum more appropriate than pigmentary glaucoma – glaucoma pigmentum. When comparing with the authors' previous study, in this one all pigmented glaucoma patients were additionally analysed for corneal astigmatism using autorefractometer. Only minimal deviation in the position of the principal corneal meridian was found.

This study represents the summary of all up to date studies of the author and argues against the classification of glaucoma given by the European Glaucoma Society⁸, which does not separate glaucoma pigmentatum from pigment dispersion syndrome and therefore incorrectly classifies it as secondary glaucoma.

TABLE 1
MYOPIC ASTIGMATISM IN PIGMENTED GLAUCOMA

Dcyl	-0.25 to -0.50	-0.75 to -1.0	-1.25 to -2.0	-2.25 to -3.0	-3.25 to -3.50
No of Patients	6	10	9	3	1

TABLE 2
THE MEAN VALUES OF MYOPIA IN PIGMENTED GLAUCOMA

Dsph	-0.25 to -1.0	-1.25 to -2.0	-2.25 to -3.0	-3.25 to -4.0
No of Patients	10	12	5	2

TABLE 3
POSITION OF THE PRINCIPAL CORNEAL MERIDIANS

Position Degree	70–80°	80–90°	90–110°	110–150°
No of Patients	4	11	10	4

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SINDROM: GLAUCOMA PIGMENTATUM PRIMARIUM – PIGMENTNI GLAUKOM

SAŽETAK

U ovo ispitivanje uključeno je 29 pacijenata s dijagnosticiranim pigmentnim glaukomom od ukupno 4350 pregledanih glaukopskih pacijenata. U kliničkom ispitivanju ovih slučajeva posebna pažnja posvećena je Krukenbergovu vretenu s naglaskom na način i razlog njegovog nastanka. U svim slučajevima s Krukenbergovim vretenom prisutna je komponenta miopije i miopnog astigmatizma. Glavna os vretena uvijek leži u glavnom meridijanu kornealnog astigmatizma. Glavni meridijan prednje površine rožnice odgovara osi glavnog meridijana njene stražnje površine. Što je viši stupanj astigmatizma, naglašenije će biti i Krukenbergovo vreteno. To vrijedi i u brojnim slučajevima gdje je vreteno teško prepoznatljivo kod astigmatizma od svega ± 0.25 do ± 0.50 Dcyl. Os meridijana odgovara direktnom astigmatizmu (110° do 70°). Autori zaključuju da nema pigmentacije Krukenbergovog vretena bez miopnog astigmatizma. Upravo stoga, kao i zbog faktora nasljeđa ovaj bi se oblik glaukoma trebao smatrati primarnim i klasificirati kao odvojeni klinički entitet.