Demographic and clinical profile of patients with juvenile onset open angle glaucoma in southwestern Nigeria

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Abstract

Background: This was a non-comparative, retrospective review of patients diagnosed with juvenile open angle glaucoma (JOAG) in the eye clinic of a tertiary hospital in southwestern Nigeria.

Objective: To document the demographic characteristics, clinical features and treatment outcome of the patients diagnosed with JOAG.

Materials and Methods: Data were extracted from the clinical record of patients diagnosed with JOAG in the eye clinic of the University College Hospital, Ibadan, Nigeria, between January 2001 and December 2005. Such data included the basic demographic data, the clinical characteristic of the patients and the outcome of their treatment.

Results: Twenty-nine patients were reviewed, which represents 3.4% of all newly diagnosed glaucoma patients seen in the out-patient section of the eye clinic of the University College Hospital, Ibadan, over the period reviewed. Eight (27.6%) patients were aged 20 years and below. The mean age was 25.1 ± SD 6.0 years. Eighteen (62.1%) had visual acuity of 6/18 or worse in the better eye at the time of presentation. The mean intraocular pressure (IOP) of the patients at presentation was 32.3 ± SD 15.2 mmHg. Eight (27.6%) patients defaulted within 6 months of presentation. The mean IOP for the 21 patients who were followed up on treatment for a mean period of 9.6 months was 17.0 ± SD 6.0 mmHg.

Conclusion: Most patients with JOAG in this review presented with advanced form of the disease. Early detection through parent-driven school eye health program and community-based case detection could help in reducing the scourge arising from JOAG among our population.

Key words: Disease, emerging, obesity

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Introduction

Glaucoma is a heterogenous group of optic nerve diseases that can be divided into congenital, juvenile onset and adult onset categories.[1] The juvenile onset open angle glaucoma (JOAG) is characterized by an early age of onset, usually between the ages of 5 and 35 years, severe elevation of intraocular pressure (IOP), and a strong genetic linkage with autosomal dominant pattern of inheritance.[2] JOAG is differentiated from late congenital glaucoma and other childhood glaucomas by the abscence of buphthalmos, Haab's striae and anterior segment dysgenesis.[2] There is limited information on the prevalence of JOAG among the Nigerian population. In the USA, it is estimated to occur in 1 in 50,000 individuals,[2] while Ellong *et al*.[3] reported a prevalence of 0.4% among Cameroonians. JOAG represents between 3.4% and 7.0% of glaucoma patients attending ophthalmic consultant clinics in some West African countries.[3,4] An important ocular finding that had been associated with JOAG is myopia.[2,5-7]

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Studies[3,4,8-10] have reported that considerable number of patients with glaucoma tend to present late with advanced diseases and JOAG patients are no exception.

There is still limited information in the literature on the clinical profile of patients with JOAG in West African subregion[3,4] and even more so in southwestern Nigeria. This may be due to the presumed rarity of the condition.

This study reviewed the demographic and clinical profile as well as the treatment outcome among patients with JOAG seen in an out-patient eye clinic of a tertiary hospital in southwestern Nigeria.

Materials and Methods

The medical records of 29 patients diagnosed with JOAG between January 2001 and December 2005 in the eye clinic of the University College Hospital Ibadan, Nigeria, were reviewed.

Data manually extracted from the case notes of the patients were the basic demographic data which included the age, sex and occupation, the indications for presentation, clinical characteristics of the patients which included the presenting visual acuity and IOP, the cup disc ratio and the treatment given with its outcome in terms of the IOP.

The diagnosis of JOAG was based on
- The presence of characteristic glaucomatous optic nerve head change;
- Visual field defect not attributable to other causes;
- Open anterior chamber angle in the affected eye on gonioscopy; and
- Age of diagnosis ranging between 5 and 35 years of age.

Excluded were
- Patients with ocular or systemic anomalies and
- Patients with features suggestive of secondary glaucoma based on the history or findings from ocular examination.

The best corrected visual acuity (pin hole, or refraction if one was done), the IOP and the cup disc ratio included in the analysis for each patient was that for the better eye. The cup disc ratio reported was based on dilated fundoscopy with biomicroscopy using 78 D Volks lens.

The presence of characteristic glaucomatous field loss affecting area within 10° from fixation is categorized as severe field loss, while characteristic glaucomatous field affectation not affecting areas within 10° from fixation is categorized as mild/moderate field loss.

The post-treatment IOP analyzed was for patients and eyes followed up for a minimum period of 6 months (range 6–13 months; mean 9.6 months).

Statistical analysis was performed using SPSS version 15. Descriptive statistics was used for continuous variables. Chi-square test was used to evaluate the difference between the treatment groups and the *P* value used was two sided. It was considered significant at a level ≤0.05.

Results

Eight hundred and fifty-three newly diagnosed glucoma patients were seen during the period under review. Twenty-nine patients met the inclusion criteria for the review; this represents 3.4% of the total glaucoma patients. Eight (27.6 %) were aged 20 years and below, while 7 (24.2%) were above 30 years of age. The mean age of the patients was 25.1 ± SD 6.8 years (range 12–35 years). There were 22 (75.9%) males.

Fourteen (48.3%) of the patients were students, while one (3.4%) was engaged in subsistence farming. Table 1 shows the socio-demographic profile of the patients.

Twenty-two (75.9%) of the patients presented on account of deterioration in vision and four (13.7%) were detected during routine ocular examination. Table 2 shows the indications for presentation of the patients to the clinic.

There was positive family history suggestive of glaucoma in

Table 1: Socio-demographic profile of the patients reviewed

Age distribution	*N* = 29	(%)
Age group (years)		
11–15	1	(3.5)
16–20	7	(24.1)
21–25	9	(31.0)
26–30	5	(17.2)
31–35	7	(24.1)
Mean age	25.1 ± SD 6.8 years	
Sex		
Male	22	(75.9)
Female	7	(24.1)
Occupation		
Student	14	(48.3)
Trader/artisan	11	(37.9)
Administrator	2	(6.9)
Farming	1	(3.5)
Unemployed	1	(3.5)

Table 2: Indications for presentation

Indication	*N* = 29	%
Deterioration in vision	22	(75.9)
Routine eye examination	4	(13.7)
Ocular ache	2	(6.9)
Refractive error	1	(3.5)

7 (24.2%) of the patients, while the history was negative in 13 (44.8%).

Eleven (37.9%) patients presented with visual acuity better than 6/18. Only 7 (24.2%) patients had field changes which could be categorized as mild to moderate. Table 3 shows the clinical characteristics of the patients.

Fifteen (51.7%) patients had medical treatment, while eight (27.6%) defaulted before any form of treatment could be commenced or within 6 months of presenting to the eye clinic. Six (20.7%) of the patients had surgical treatment. The surgery of choice in all the cases was trabeculectomy with adjuvant 5-flurouracil as the antimetabolite.

The mean IOP of the patients at presentation was 32.3 ± SD 15.2 mmHg (range 10.0–58.0 mmHg) and for those who had medical treatment was 33.1 ± SD 13.4 mmHg (range 18–51 mmHg). The mean IOP for all the 21 patients who were followed up on treatment for a minimum period of 6 months was 17.0 ± SD 6.0 mmHg (range 9.0–32.0 mmHg). Table 4 shows the IOP profile among the patients reviewed.

Table 3: Clinical profile of the patients reviewed

	n	%
Family history of glaucoma		
No	13	(44.8)
No record	9	(31.0)
Yes	7	(24.2)
Presenting visual acuity (in the better eye)		
>6/18	11	(37.9)
6/18–6/60	3	(10.3)
<6/60	12	(41.4)
NPL	3	(10.3)
Cup disc ratio (in the better eye)		
<0.4	3	(10.3)
0.4 – 0.7	7	(24.1)
>0.7	19	(65.6)
Visual field changes (in the better eye)		
Mild to moderate	7	(24.1)
Severe	15	(51.7)
No record	7	(24.1)

Table 4: Intraocular pressure profile of the patients reviewed

Pre-treatment IOP		
All patients (*N* = 29)	Mean	32.3 ± SD 15.2 mmHg
	Median	30.0 mmHg; range 10.0–58.0 mmHg
Medical treatment group (*n* = 15)	Mean	33.1 ± SD 13.4 mmHg
	Median	30.0 mmHg; range 10.0–51.0 mmHg
Surgical treatment group (*n* = 6)	Mean	42.7 ± 14.6 mmHg
	Median	46.5 mmHg; range 18.0–58.0 mmHg
	P	0.20
Post-treatment IOP		
All patients (*N* = 21)	Mean	17.0 ± SD 6.0 mmHg
	Median	15.0 mmHg; range 9.0–32.0 mmHg
Medical treatment group (*n* = 15)	Mean	19.1 ± SD 5.9 mmHg
	Median	18.0 mmHg; range 10–32.0 mmHg
Surgical treatment group (*n* = 6)	Mean	13.8 ± SD 6.3 mmHg
	Median	11.5 mmHg; range 9.0–26.0 mmHg
	P	0.11

IOP: Intraocular pressure

Discussion

JOAG, a rare subset of primary open angle glaucoma, remains an important cause of visual morbidity, even more importantly when one considers the age group affected and the late presentation of patients. In this review, 48% of the patients were students and 51.3% were already severely visually impaired at presentation. This obviously has strong socioeconomic implication when the damage resulting from JOAG is quantified in terms of blind years.

The age distribution of the patients in this review compares with what was reported among Cameroonians (mean age 26 ± 6.8 years; range 10–35 years).[3] The minimum age of 12 years recorded in this review which falls within the same age bracket of 12 years that was reported by Ellong *et al.*[3] may indicate the earliest time when the loss of retinal ganglion cells associated with JOAG become marked enough to affect the central vision and as such becoming noticeable. Screening at an early stage possibly as part of a school eye care program might have uncovered some of these cases. Futhermore, the rate of progression of JOAG may be more rapid among Africans compared to the Caucasians. This assumption may be substantiated by the mean age of onset of 18 years reported by Johnson *et al.*[11] and 21.3 years reported by Bayat *et al.*[12] These two reviews were among non-Africans. The age bracket of 12 years is also important as most children would have graduated from the elementary school to high school and as such be engaged in more visually demanding tasks of reading independently. This may unmask a covert deterioration in vision. The proportion of the patients that gave positive family history suggestive of glaucoma is rather low considering that JOAG has an autosomal dominant pattern of inheritance.[2] This is one of the limitations associated with retrospective reviews. Futhermore, in an African setting where blindness in a family is highly stigmatized, many patients are hardly forthcoming with history of blindness among family members. However, of interest is the fact that 88.3% in the review among the Cameroonians[3] gave positive family history. Eliciting the history may also have to do with the skill of the ophthalmologist.

Majority of the patients presented with advanced disease, indicating late presentation. Late presentation among glaucoma patients has been highlighted in many previous reviews.[3,4,8-10] The late presentation among these patients could be due to the asymptomatic feature of glaucoma in its early stage. So also is the health-seeking behavior of the people. Many factors that affect the health-seeking behavior of an individual have been documented in the literature and these include socioeconomic indicators.[13-16] Among patients with stroke, Kitko *et al.*[17] were also able to recognize factors such as lack of knowledge, fear of hospital, and denial as the factors affecting health-seeking behavior. In an African context, blinding eye diseases such as chronic glaucoma are usually associated with lot of myths, even more so when it presents early in life, and as such the tendency to seek unorthodox treatment options. Ashaye *et al.*[18] were able to show that there are several misconceptions about the causes of eye diseases among the Yoruba ethnic group, with fatalistic beliefs such as withcraft ranking highest in the cause of blindness. All these factors might have contributed to the late presentation among these patients.

The high clinic dropout rate of 26.7% is a cause for much concern. This could be a reflection of the way the patients perceived their eye problem. In a review of the characteristics of patients who dropped out of a glaucoma clinic in southwestern Nigeria, Ashaye *et al.*[19] found that factors such as younger age group, mild to moderate disease state and patients who do not perceive their problem as serious are positively correlated with high dropout rate.

The mean intraocular pressure of 32.3 ± SD 15.2 mmHg among the reviewed patients compares with the 28.2 ± SD 9.3 mmHg reported among the Cameroonians.[3] This is lower than the 38.5 mmHg (range 30–50 mmHg) reported by Wiggs *et al*.[7] in a clinical characterization of 23 patients with JOAG. Of interest is the fact that 37.9% of the patients had IOP which was less than 21 mmHg at presentation. This does not conform with what had been commonly reported in the literature[2] that patients with JOAG usually have high IOP (>30 mmHg). Among the Cameroonians,[3] only 5.3% had IOP that was less than 21 mmHg. However, a retrospective review in which the IOP documented was a one-time measurement may not be a total representative of the IOP profile for these patients. Merritt *et al.*[20] recorded the highest IOP between 6 p.m. and 12 midnight in 9 of 10 patients with JOAG reviewed. Twenty-four hour IOP phasing of these patients would have provided more accurate information about the IOP profile of some of these patients presumed to be having normal tension glaucoma.

The choice of treatment was at the discretion of the managing ophthalmologist, though as expected patients with higher IOP must have been offered surgical treatment as shown by the mean IOP of the eyes that had medical treatment (33.1 ± SD 13.4 mmHg) and the eyes that had trabeculectomy with 5-fluorouracil (42.7 ± SD 14.6 mmHg), although this difference was not statistically significant ($P = 0.20$). Fewer patients (eight eyes of six patients) took up surgery possibly because of patients' refusal borne out of fear of surgery and also due to the late presentation of the patients by which time the surgeon may not consider surgery as a better treatment option because of the risk of wipe out syndrome.

Koraszewka-Matuszewska *et al.*[6] reported gradual reduction in the effectiveness of trabeculectomy with time despite initial satisfactory IOP lowering effect among patients with JOAG, while Jacobi *et al*.[21] found that trabeculectomy in patients with JOAG shows satisfactory outcome comparable to the outcome in adult.

The small sample size of 29 patients seriously limits the generalization of the findings of this review. However, it could serve as a basis for a larger multicenter study on JOAG in Nigeria. Also, this may give room for exploring the interesting topic of genetics of JOAG among the Nigerian population.

Conclusion

The findings of this review show that patients with JOAG tend to present late with advanced disease, with a high dropout rate. The authors believe that early detection of cases during exercises such as parent-driven school eye health program and community-based health education on the benefit of early detection and good treatment compliance could help in reducing the scourge arising from JOAG.

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