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Case Report

Juvenile open angle glaucoma –A case series study

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ABSTRACT

Background: Juvenile open angle glaucoma is a rare form of glaucoma that differs from primary open angle glaucoma in its age of onset and magnitude of intraocular pressure.
Juvenile open angle glaucoma has its onset between 3-40 years of age with relatively high intraocular pressure.
Purpose: For the documentation and describe clinical manifestations management and outcome of management of the patients diagnosed as Juvenile open-angle glaucoma at the glaucoma department, CEITC, Chittagong, Bangladesh.
Materials and Methods: This is a hospital based prospective observational case series review. 20 patients who were diagnosed as Juvenile open-angle glaucoma from November 2010 to December 2011 were included in this study.
Patient particulars history with main causes of hospital presentations were recorded. Ophthalmic examinations and management given were documented. Similar relevant details were recorded for different follow-up periods.
Results: 40 eyes of 20 patients were included in this study. There were 16 male and 4 female. All cases were bilateral. Age more than 18yrs. (18-35) in 16 patients and below 18yrs. (5-18) in 4 patients. 15 patients came from rural area and 5 patients from urban. Pretreatment average IOP in the both eyes was 32±3mmhg, which was 15±1mmhg after treatment. 24 of 40 eyes were presented with advance field defects. 85% (17 patients) had myopic refractive error. In 18eyes pre treatment presenting visual acuity was <6/60 and >6/60 in the rest of the eyes. Visual acuity was improved after treatment. In 21 patients (53%) IOP was controlled with 2-3 medications. In 19 eyes (48%) IOP was controlled with filtration surgery.
Conclusion: As Juvenile open-angle glaucoma presented with high IOP and advance field defect, early diagnosis, appropriate investigations and medical or surgical management is mandatory to stabilize IOP and to prevent progression of field defects.

1. Introduction

Juvenile open-angle glaucoma (JOAG) which has an age at onset of (5-35) years tends to be more aggressive. It is usually resistant to medical therapy and is associated with more severe visual impairment than primary open angle glaucoma.[1]

The estimated prevalence of JOAG ranges from 0.38to 2 in 100000 in individuals between 4-20 years of age and 4 percent of child hood glaucoma.

Identifying risk factor are important because this information may lead to development of strategies for disease screening and prevention and may be useful in identifying persons for whom close medical supervision is indicated. Thick compact tissue in the angle represents an immature development of the trabecular meshwork and may be one of the primary cause of increase intraocular pressure in Juvenile glaucoma[2] the more extensive the immaturity, the earlier the glaucoma will become manifest. GLCIA, the first open angle glaucoma gene, was initially

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mapped in a large Juvenile glaucomafamily that localized to chromosome [1] the mutation in the gene, which are suspected to be responsible for open angle glaucoma, produce a protein, myocilin that is induced in trabecular meshwork. [3]

Male gender, positive family history, myopia, African ancestry and prominent iris process are some risk factors.

Primary glaucoma represents a significant public health problem. Although rare, untreated Juvenile glaucoma patients are ultimately diagnosed as primary open angle glaucoma after 35yrs. It is an important cause of blindness in the western countries and in blacks. It is also not uncommon in this subcontinent.

Management protocol is almost similar to POAG. All antiglaucoma can be used including prostaglandin analogues which is ineffective in the congenital glaucoma. Miotics should be used with caution due to it causes ciliary spasm and induced myopia with frontal headeche. Options for surgical therapy for JOAG include filtration surgery (trabeculectomy) drainage implants angle procedure in its early age (goniotomy and trabeculotomy) and cycloablation procedure.

The purpose of this study is to documentation and to see the management and its outcome of JOAG in a tertiary eye hospital of Bangladesh.

2. Method

This was hospital based combined non concurrent and concurrent descriptive case series study. Study was conducted in the glaucoma clinic of Chittagong eye infirmary and training complex which is one of the largest tertiary eye hospital in Bangladesh.. Cases were identified throughout a two years period from November 1^{st} 2008 to December 1^{st} 2010.

All patients were reviewed by a single consultant. Details of history was taken included the biographical details of patients (age, gender, address etc) and history of presentation.

Ophthalmic examination was done on patients and examination details included visual acuity (VA); intra-ocular pressure (IOP) measurement by Goldmann Applanation Tonometer; gonioscopic findings by Goldmann 2-mirror contact goniolens; fundoscopic findings by 90 diopter lens and any other notable ocular findings. Visual field was analyzed by Humphrey visual field analyzer. The method of management was recorded.

For previously diagnosed patients, their medical records were retrieved and relevant data were extracted and asked to come for follow-up as necessary. Newly diagnosed patients were duly processed and asked to return for future follow-up visits. At least three follow-up data were recorded, 1 month after diagnosis of joag, then 3 months and 6 months. On all visits ophthalmic examination was done by the same consultant. Medication was started according to height of IOP. Those patient presented with more than 30 intraoculr pressure we started 2 medications (b blocker and alpha 2 agonist). After one month we added carbonic anhydrase inhibitor if IOP was not controlled and progression of visual field defect. We decided to do surgery in non responsive cases. We did filtration surgery with mytomomicin in relatively younger age group and where we got thick tenons during operation.

After collection of data, they were then tabulated and analyzed. Outcomes of management were assessed mainly with regards to IOP control. Statistical analysis was done using SPSS v.13.

3. Results

A total number of 40 eyes of 20 patients were encountered during the study period. All of the cases were bilateral affected of these 10 were newly diagnosed and 10 were previously diagnosed. The ages of 16 patients (80%) were between 18-30 yrs. And same of 4 patients were (5-18) yrs. Mean age (23±7.13) years. 16 patients (80%) were male and 4 were female. 50% of the patients are student. Remaining 50% were either service holder or businessman or daily laborers. 15 patients (70%) came from rural area. 65% of them were from middle class family and 35% of them from poor family. (90%) 18 patients came with gradual decrease vision in the both eyes and 2 patients (10%) came with only headache.

Mean duration of symptom was 2yrs. 8 patients (40%) had a strong family history of glaucoma. 50% of them were previously treated by local ophthalmologist. Reasons of delayed presentation were the lack of knowledge (60%), lack of eye care facilities (20%) and poor economy (20%).

17 patients (85%) had myopic refractive error, 1patient (5%) had hyperopia and the rest 2 patients had no refractive error. Average IOP at presentation was around 35mmhg which was reduced to around 15mmhg after treatment (P=1.440).

24 cases had a cup disc ratio of more than.8:1 during presentation and had .7:1 in 10 cases and rest of them had.5-.6:1 during presentation. No other abnormalities were found during fundus evaluation.

24 eyes had advance field defects like total field loss (5.26%), biarcuate scotoma (21.05%) and tibular field (31.59%). IOP was controlled with either 2 or 3 medications in 21 eyes (52.5%), those patients (47.5%) resistant to medical treatment needed filtration surgery to control IOP (Table 5).

Presenting visual acuity was <6/60 in 18eyes (45%) and 6/9-6/60 in 22 eyes. Post management visual acuity was improved (Table 6).

4. Discussion

Kass and Becker were among the first to observe a strong correlation between family history and glaucoma. [4,5] Based

Table 1: Demographic features and presentation of the patients

	N	Percent
Age group		
5-18	4	20
18+	16	80
Mean age	23 years SD ± 7.13 years.	
Gender		
Male	16	80
Female	4	20
Occupation		
Student	10	50
Service	3	15
Business	4	20
Daily labor	3	15
Patient's residence		
Rural	15	75
Urban	5	25
Socio economic condition		
Poor	7	35
Middle	13	65
Rich	0	0
Presenting complain		
Decreased Vision	18	90
Eye ache	2	10
Duration of symptom (mean time) – 02 year		
Family History		
No	12	60
Yes	8	40
Reason of delayed presentation		
Economic	4	20
Lack of eye care facilities	4	20
Lack of Knowledge	12	60
Previous eye treatment		
Yes	10	50
No	10	50

Table 2: Status of refractive error

	N	Percent
Myopia	17	85.0
Hyperopia	1	5.0
No refractive error	2	10.0
Total	20	100.0

Table 3: Management of intraocular pressure

IOP	Right	Left
Before treatment	33	35
After treatment	15	16
	P = 0.440	

Table 4: Visual field test of patients

	N= 38	Percent
Nasal step	5	13.15
Bearcuate Scotoma	8	21.05
Tubular	12	31.58
Inferior Actuate Scotoma	2	5.26
Superior Actuate Scotoma	9	23.68
Total field loss	2	5.26
Total	38	100.0

Table 5: Treatment of the patients

Medical treatment	N	Percent
Two Medications	15	37.50
Three medications	6	15.00
Surgical treatment		
Trabeculectomy	11	27.50
Trabeculectomy with MMC	8	20.00
Total	40	100

Table 6: Visual acuity of the patients

VA	Presenting VA	Post management VA
6/6-6/18	19 (47.50%)	25 (65.0%)
6/24-6/60	3 (7.50%)	3 (5.0%)
6/60+	18 (45.0%)	12 (30.0%)
Total	40 (100.0%)	40 (100.0%)

on their observation, the researchers suggested that the most effective method of glaucoma detection would be to check family members.

40% of our patient had a strong family history of glaucoma. The percentage may be more as the rest of the patient did not know the cause of their relative's blindness.

It has been mentioned that, mutations of the trabecular meshwork glucorticoids genes could cause elevated IOP. This is called TIGR protein or myocilin was identified in Juvenile open-angle glaucoma families. We did not do any genetic analysis in our patient.

Juvenile open-angle glaucoma terminology often used when open-angle glaucoma diagnosed at young age (typically 10-30yrs.). Mean age of our study populations is (23±7.13)yrs. So it is strongly similar to other studies.

Although primary glaucoma's are more common in female, male are predominant in our study populations. Sensitive patients whose visual perfection is a factor usually present in the clinic due to their visual problems. 50% of our patients are student who presented earlier than others.

Most of our patients are from rural middle or poor class families. This may be due to lack of awareness and lack of health care facilities at rural area. Poor economy and remoteness may also play role. In our study, main reason of delayed presentation is lack of knowledge about the disease.

Electron microscopy specimens of anterior chamber angle reveal thick compact tissue consisting of cells with

fine processes and extra cellular substances. Thick compact tissue represents immature development.[2] We did not do any histopathology but gonioscopic examination shows abnormal processes over trabecular meshwork, concave iris insertion suggestive of immature development. Angle was open 360^0 areas in all patient.

According to different studies presentation of JOAG is aggressive. In this study most of the patients presented with high intraocular pressure (30-35)mm hg, enlarged C:D ration >.8:1 and with advance field defects.

Juvenile open-angle glaucoma is associated with more severe visual impairment than primary open angle glaucoma.[6] 45% of our case (eyes) presented with <6/60 vision. As 50% of our patients are students, they are visually sensitive and their presentation was quite earlier.

Aggressive Juvenile open-angle glaucoma is more resistant to medical therapy.[6] 47% (19eyes) of our cases were resistant to medical therapy; they were treated with 2-3 medications. Those patient who's IOP was not controlled even with two medications, filtration surgery was advised.

Trabeculectomy, a penetrating filtration procedure, is the treatment of choice in treating medically uncontrolled open angle glaucoma. However, intra operative and postoperative complications are not uncommon.[7–9] In our series, no intraoperative or post-operative complications were arrived.

The success rate of filtration surgery in young patient is believed to be lower than POAG.[10] To decrease the fibrovascular proliferation, we did 8 filtration surgeries with mitomycin c in relatively more advance cases.

Primary trabeculectomy in young adults may have a favorable outcome despite no antimetabolite therapy.[11] We also did 11 filtration surgery without antimetabolite which are still doing well.

5. Conclusion

Juvenile open angle glaucoma presents usually at an advance stage. Proper docementation with appropriate examination is important to diagnose JOAG. Those patient who have strong family history of glaucoma, should do a routine periodic eye checkup. Even at an advance stage appropriate medical or surgical treatment can stop further progression of the diseases. Filtration surgery with mitomycin c is recommended for very advance cases to assure longtime functioning bleb.

6. Source of Funding

None.

7. Conflict of Interest

The authors declare that there is no conflict of interest.

References

1. Ellis O. The etiology, symptomatology and treatment of juvenile glaucoma. *Am J Ophthalmol*. 1948;31:1589–96.
2. Tawara A, Inomata H. Developmental Immaturity of the Trabecular Meshwork in Juvenile Glaucoma. *Am J Ophthalmol*. 1984;98(1):82–97. doi:10.1016/0002-9394(84)90193-4.
3. Maurya RP. Is glaucoma a genetic disorders. *Ind J Clin Exp Ophthalmol*. 2016;2(3):167–8.
4. Epstein DL, Allmgham RR, Schuman JS. Chandler and Grant's Glaucoma. 4th ed. Balti¬more: Williams & Wilkins; 1997. p. 641–6.
5. Ritch RM, Shields MB, Krupin T. The Glaucomas. 2nd ed. St Louis: Mosby; 1996. p. 753–65.
6. Alexandros N. Primary viscocanalostomy for juvenile open- angle glaucoma. *Am J Ophthalmol* . 2005;140:490–6.
7. DeBry PW, Perkins TW, Heatley G, Kaufman P, Brumback LC. Incidence of late-onset bleb-related complications following trabeculectomy with mitomycin. *Arch Ophthalmol*. 2002;120:297–300.
8. Busbee BG, Recchia FM, Kaiser R, Nagra P, Rosenblatt B, Pearlman RB. Bleb-associated endophthalmitis. *Ophthalmology*. 2004;111:1495–1503. doi:10.1016/j.ophtha.2004.01.028.
9. Siegfried CJ, Rosenberg LF. Hypotony after glaucoma filtering surgery: mechanisms and incidence of glaucoma. *J Glaucoma*. 1995;4:63–9.
10. Lanzl IM, Wilson RP, Dudley D, Augsburger JJ, Aslanides IM, Spaeth GL. Outcome of trabeculectomy with mitomycin-C in the iridocorneal endothelial syndrome. *Ophthalmology*. 2000;107(2):295–7. doi:10.1016/s0161-6420(99)00077-9.
11. Costa VP, Katz LJ, Spaeth GL, Smith M, Gandham S. Primary Trabeculectomy in Young Adults. *Ophthalmology*. 1993;100(7):1071–6. doi:10.1016/s0161-6420(93)31536-8.

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