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The Missed Glaucoma: A Study on Accidentally Discovered Cases with Juvenile Glaucoma

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ABSTRACT
Background: The diagnosis of juvenile glaucoma can be missed, as this age group seldom seeks ophthalmological consultation in the lack of ocular symptoms
Objective: This study aimed to evaluate incidence of missed/late diagnosed glaucoma in the juvenile age group.
Methods: A retrospective case series on 38 cases in the juvenile age group (age 8-45 yeas) who were diagnosed with glaucoma. The incidence of missed/delayed diagnosis of glaucoma in these cases were reported and evaluated.
Results: 22 (57.9%) patients presented with advanced glaucoma with a cup/disc (C/D) ratio of 0.8 or more and significant visual field (VF) changes in one or both eyes. 4 (10.5%) of these patients had total glaucomatous optic atrophy in 1 eye at the time of diagnosis. The remaining 14 patients (36.8%) were diagnosed as mild/moderate juvenile glaucoma with C/D ratio less than 0.8 at time of diagnosis. Clinical findings of 7 of these cases are described here.
Conclusion: Juvenile glaucoma is an often missed potentially blinding glaucoma.
Keywords: Glaucoma, Juvenile glaucoma, Glaucoma detection.

INTRODUCTION

Rare and poorly defined, juvenile glaucoma (JG) is viewed by some as an early-onset variant of primary open-angle glaucoma (POAG) and by others as a developmental glaucoma.

Asymptomatic in its beginning, its course is more progressive than that of POAG, and it is frequently recalcitrant to treatment [(1-3)].

The diagnosis of juvenile glaucoma can be missed, as this age group seldom seeks ophthalmological consultation in the lack of ocular symptoms. The aim of this work was to check how often the diagnosis of new cases of juvenile glaucoma was missed.

CASE SERIES

This was a retrospective case series. Medical files of 38 patients (59 eyes) diagnosed with JG (ages 8-45) at the Glaucoma Service of the Research Institute of Ophthalmology (RIO) Hospital and a private glaucoma practice between January 2014 and December 2019 were reviewed.

Included in this analysis are demographic data, preoperative angle evaluation, intraoperative and postoperative complications, IOP, number of drugs, C/D ratio (CDR), and visual acuity (VA) data were all recorded and saved before surgery for analysis.

Perkins handheld applanation tonometer was used to assess intraocular pressure (Haag-Streit, Koniz, Switzerland). Snellen charts were used to assess VA. The +90 slit-lamp biomicroscope lens was used to rate CDR on a scale from 0.0 to 1.0. There was just one examiner who took all the measurements.

RESULTS

A total of 38 eyes were diagnosed with juvenile glaucoma during the review period. 22 (57.9%) patients presented with advanced glaucoma with a cup/disc (C/D) ratio of 0.8 or more and significant visual field (VF) changes in one or both eyes. 4 (10.5%) of these patients had total glaucomatous optic atrophy in 1 eye at the time of diagnosis.

The remaining 14 patients (36.8%) were diagnosed as mild/moderate juvenile glaucoma with C/D ratio less than 0.8 at time of diagnosis. Clinical findings of 7 of these cases are described here.

CASE 1

A 19 years old girl was diagnosed as glaucoma a week before presentation. She was prescribed bilateral topical CAI/beta-blocker drops. Her visual acuity (VA) Rt 1.0 while Lt: finger counting (FC) 30 cm. Her intraocular pressure (IOP) was Rt 16, Lt 16 mmHg (on bilateral medications), while her cup/disc ration (CDR) was Rt 0.85 (Figure 1A), Lt secondary optic atrophy (OA). Right pupil was round and reactive, while left pupil was dilated and sluggishly reactive (figure 1B). Gonioscopy revealed bilateral advanced trabeculodysgenesis (Figure 1C).

The case was subjected to bilateral consecutive surgery of adjusted trabeculotomy [(4)]. Postoperatively, her intraocular pressure was well controlled at 10-14 mmHg without medications for the follow up period of 7 years. Her vision in the markedly affected left eye, however, did not show any improvement.

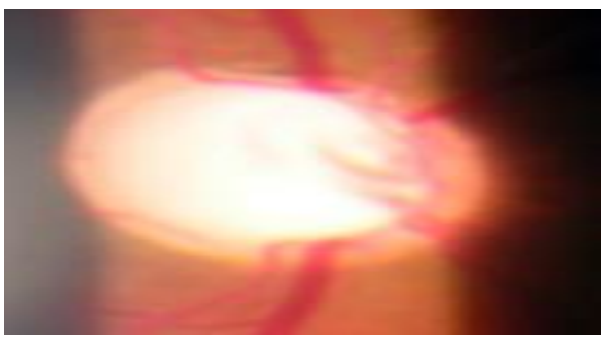

Figure 1A

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Figure 1B

Figure 1C

CASE 2

A 23-years-old man was accompanying his 25-year-old brother who was diagnosed with glaucoma in infancy, with several surgical interventions in both eyes, with a lost left eye and a right eye controlled on full medications. The younger brother sought ophthalmic consultation for a potential possibility of glaucoma like brother. His best corrected visual acuity (BCVA) was 1.0 bilaterally, his IOP was found to be 36 and 34 mmHg in the right and left eyes respectively, with 0.6 optic disc cupping.

He was prescribed medical therapy, which controlled the glaucoma, and later subjected to successful surgery to evade the topical side effects of medications.

CASE 3

A 9-years-girl usually accompanied her younger 1-year-old brother diagnosed and operated for primary congenital glaucoma. Being in the eye clinic, the father asked to have a glasses prescription for the elder sister. Her IOP was found to be Rt: 40 and Lt: 38 mmHg, with a subtotal optic disc cupping of 0.85 and 0.9 at Rt and Lt successively.

CASE 4

After being declined refractive surgery because of subclinical keratoconus at another facility, a 23-years-old man sought out our expertise. His right eye had a refraction of 0.9 (sphere 3.00, cylinder 2.75 and axis 25), while his left eye had a refraction of 1.0 (sphere -3.50, cyl -1.25 and axis 170). His right eye's IOP was 28 mm Hg and his left eye's IOP was 30 mm Hg. Both eyes had normal, rounded optic discs, with a 0.6-disc cupping in the right eye and a 0.7-disc cupping in the left eye, as seen during fundus examination. No known risk factors for subsequent glaucoma were present, including trauma, steroid usage, or other factors. Bilateral faint lower arcuate scotomata were present on the visual field assessment, suggesting early glaucoma (Figure 2 A). Retinal nerve fibre layer (RNFL) thickness was below normal inferiorly in both eyes as measured by optical coherence tomography (OCT), with an overall decrease in rim area (Figure 2B). Medical treatment for glaucoma was started, and then consecutive surgeries were done in both eyes utilising adjusted trabeculotomy technique.

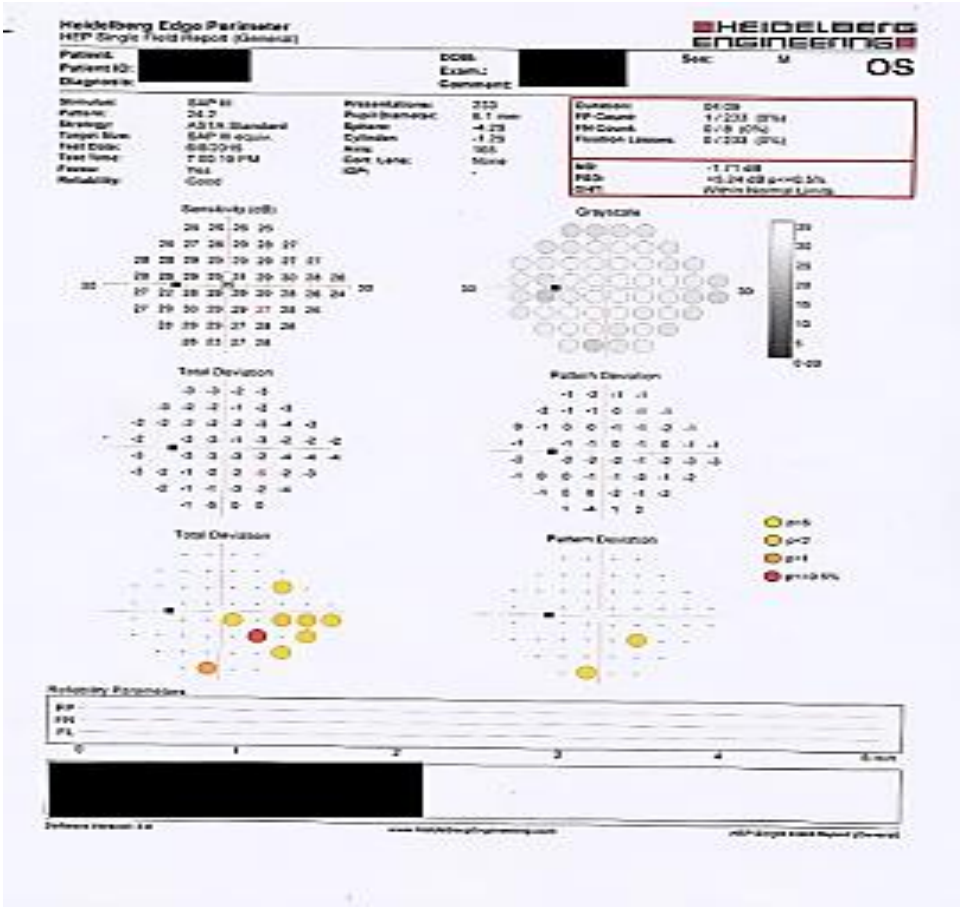

Figure 2A: Visual field of the left eye of case 4

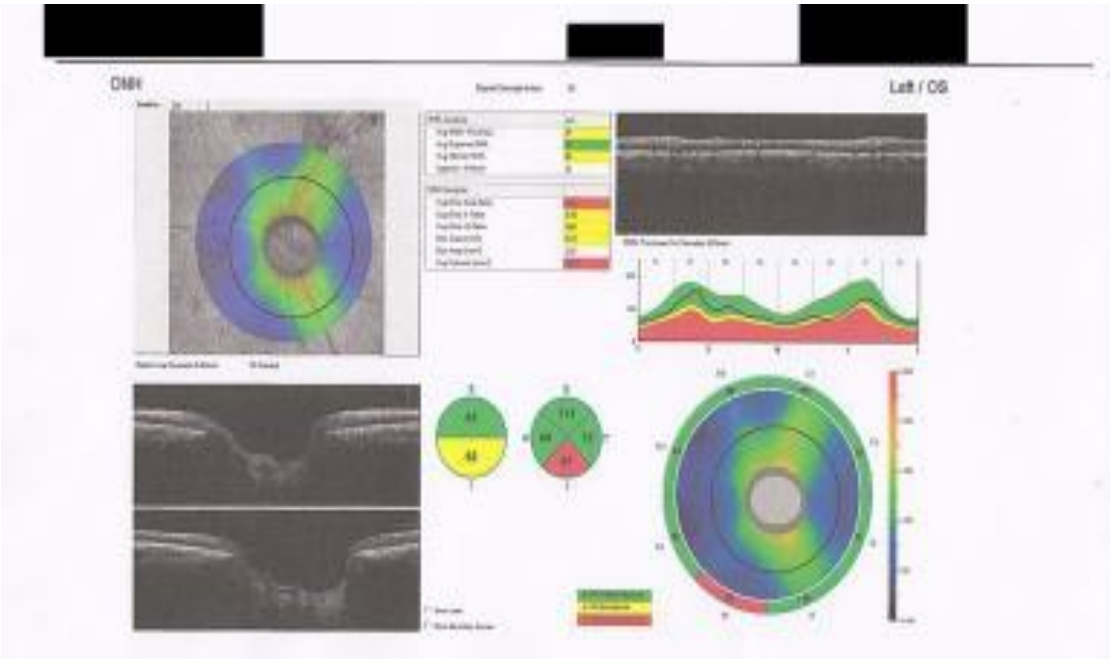

Figure 2B: OCT of the left eye of case 4

CASE 5

A 13-years-old girl presented with her myopia for glasses renewal. She has been having new glasses yearly for the past 6 years, and had a recent marked reduction of Rt eye vision. Examination showed BCVA of no PL in the right and 0.7 in the left. Her IOP was Rt 34 and Lt 28 mmHg, and CDR Rt was 1.0 and Lt was 0.7. Both eyes were operated, with IOP spikes after surgeries, followed by eventual control at low teens. Right eye could see a partial 0.05, and left eye 0.9.

CASE 6

A girl presented at the age of 11 years with a weak eyesight in the right eye, and giving history of congenital cataract surgery at the first year of life (Figure 3). Her VA was Rt FC 3 meters and Lt was 0.5. Her IOP was found high in both eyes at Rt was 30 and Lt was 32 mmHg, with CDR of 0.9 and 0.7 for Rt and Lt successively.

Figure 3

CASE 7

A 14 years old boy was referred for the management of his right eye glaucoma. On presentation, he gave a history of previous 2 glaucoma surgeries in the left eye at the ages of 4 and 5 years. His VA was 0.8 at Rt (spherical equivalent refraction: -1.5) and FC 1.5 m at Lt. IOP was 26-30 and 14-21 mmHg at Rt and Lt respectively. His CDR was 0.9 and 0.95 at Rt and Lt respectively.

DISCUSSION

According to the onset of diagnosis, glaucoma can generally be divided into 3 age groups: birth-infancy (pediatric glaucoma), childhood-young adulthood (Juvenile glaucoma), and late adulthood. Pediatric glaucoma, especially its most common form of primary congenital glaucoma (PCG), is more than often diagnosed timely because of the symptoms it produces (corneal cloudiness, photophobia, etc., and is diagnosed proportionally more often than in adults, albeit with a delay [5, 6]. Late adult glaucoma comes in an age when people are inclined to have their eyes checked, either in conjunctions with other senile diagnosis like cataract or retinal conditions, or as part of a yearly routine check. Still, even in this higher age group, the rate of undiagnosed glaucoma cases, at least for open-angle glaucoma, is 49–53% [7-9].

There is a lack of data on the prevalence of juvenile glaucoma in the literature. Prevalence of juvenile glaucoma based on family history was 0.01% in a recent cohort study. Between the ages of 18 and 40 years, prevalence rate was an order of magnitude higher

(0.16%) [10]. Patients with glaucoma were found to make up to 0.4% of female participants and 0.6% of male participants aged 40–49 years in the Egna-Neumarkt Study [11].

Juvenile glaucoma typically affects people in an age category, which seldom seek medical advice if there are no symptoms, and remains eventually the least examined age group (the healthy young!). Even when they present to eye doctors for other reasons like requiring glasses for refractive errors, or for the treatment of seasonal allergies, ophthalmologists are often reluctant to measure IOP for this age group, and not infrequently miss a glimpse at the optic disc. This age category can also be prescribed topical steroids for some allergic conditions common in this age group like vernal keratoconjunctivitis, which can play a role in causing a steroid-induced glaucoma. Causes of glaucoma in the juvenile age group can be classified as developmental either a late diagnosed PCG or with a true late onset, and secondary, including steroid-induced, aphakic, and those associated with other anomalies [12].

The accidental discovery of bilateral advanced glaucoma with one absolute eye in a 13 and 19-years-old girl (cases 1, 5) implies that this was quite a late diagnosis, and that bilateral glaucoma was an ongoing process for long previous years in that girl with bilateral significant goniodysgenesis. The finding of established glaucoma case 2 and 3, who had siblings previously diagnosed with pediatric glaucoma, highlights again and again the familial and hereditary nature of glaucoma in this age group, and calls for the importance of routine check of siblings of glaucoma patients [13].

Similarly, some of the case described had an associated myopia, which is a known association in literature [14]. The question remains on the relation between myopia and glaucoma in this age group. Whether it is a cause and effect, or a co-incidence.

Case 6 demonstrated the accidental discovery of glaucoma in an aphakic young girl years after pediatric cataract surgery. Aphakic glaucoma in children is a known entity [15], and cataract extraction in children warrants a life-long monitoring of IOP in these patients.

The partial apparent regaining of sight in the right eye of the 13-years-old girl (case 5) who could not perceive light with her right eye, with partial regaining of ambulatory vision postoperatively, demonstrates the importance of intervention in this age group if the IOP is high, even in eyes, which apparently have no vision. Whether this was the result of child response to visual acuity testing not being reliable, or because of relief of an actual significant pressure on an already highly compromised disc, remains to be answered.

CONCLUSION

Juvenile glaucoma is a commonly missed glaucoma. With the modern age of sub-specialization in Ophthalmology, awareness has to be spread in all presumably unrelated Ophthalmology subspecialties about the importance of routine IOP measurements in all age groups, and more and more so in patients with a family history of the disease.

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