Clinical Approach to Nanophthalmus: Diagnosis and Management Features

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ABSTRACT

Purpose: To evaluate the results of diagnostic features and management approaches in 4 cases with nanophthalmos.

Material and Methods: Four cases with nanophthalmos were evaluated retrospectively in terms of diagnostic features and treatment approaches. Three of the cases were siblings. Three siblings were female and the other was male. Bilateral laser iridotomy (LI) was applied to one of the sibling cases before being sent to our clinic. Argon laser peripheral iridoplasty (ALPI) was performed in two siblings in our clinic. The youngest sibling case was closely followed for glaucoma development. A trabeculectomy was performed in another center in the eye of the male patient with glaucomatous optic atrophy. Phacoemulsification (Phaco) + viscogoniosynechialysis surgery was performed in the other eye of this patient, whose intraocular pressure (IOP) was not under control with maximal medical treatment.

Results: No complications were encountered in the interventions to the cases. Additional medication (fix combination) was needed during follow-up.

Conclusion: Patients with nanophthalmos should be carefully monitored for glaucoma development with advancing age. Before the peripheral anterior synechia (PAS) develops, ALPI is valuable for IOP control. According to the degree of development of PAS in angle, synechialysis combined with phaco can provide significant benefits in IOP control. However, all interventions to these cases are open to complication development, and close follow-up is required. It should be considered additional medication and/or surgery may be required during follow-up.

Keywords: Microphthalmos, nanophthalmos, glaucoma, argon laser peripheral iridoplasty, phacoemulsification, synechialysis.

INTRODUCTION

Nanophthalmos a rare form of microphthalmia and denotes clinical entity with normal lens size despite smaller ocular volume (two-third of normal ocular volume). It is often bilateral. It can be sporadic or inherited.[1,2]

Sclera is thick in these eyes with high hypermetropia. It is though that resistance of abnormal collagen structure to ocular growth play role in the pathogenesis. In eyes with nanophthalmos, axial length is rather short (<20.5 mm). While lens volume comprises 4% of total ocular volume, it ranges from 10% to 30% due to decreased ocular volume in eyes with nanophthalmos.[1-3] The disproportion caused by lens leads abnormal configuration of anterior segment. By advancing age, alterations in lens trigger angle closure and resultant glaucoma by increasing aggregation in anterior segment. In addition, abnormalities such as loss of foveal reflection, retinal folds, retinoschisis, increased choroidal thickness and choroidal folds and optic disc drusen can be seen in nanophthalmic eyes.[1]

In nanophthalmos, ophthalmologists can face challening problems such as glaucoma, or uveal effusion. The treatments and interventions in these cases have potential for complications. Here, we discuss cases with nanophthalmos, clinical approach and diagnostic and therapeutic characteristics in these cases in the shed of literature and our experiences.

CASES

Case 1

A 48-years old woman referred to our clinic due to failure in controlling IOP, who had been followed with diagnosis

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of glaucoma in another facility. In examination, visual acuity was 0.2 in right and 0.1 in left eye (Snellen cards) with bilateral +16 D hypermetropia correction. Central corneal thickness (CCT)was 610 µ in both left and right eyes. Corrected IOP was 26 mmHg in right and 27 mmHg in left eye with fixed brimonidine plus timolol maleate combination. In anterior segment examination, anterior chambers were narrow and bilateral LI was observed at direction of 2 o'clock. In gonioscopy, there was bilateral closed angle and PAS in iridotomy fields (Picture 1). The concavity: disc (c:d) ratio (c/d) was 0.4 in both eyes while macular folds were observed in fundus examination. Axial length was 14.9 mm. The patient was diagnosed as nanophthalmos and overall 360° ALPI was performed in 2 distinct sessions (180° in each session). During ALPI, 24 pulses (12 pulses per quadrant) were applied using following parameters: 500 µm, 0.5 second, 300 mw argon laser parameters. No complication was recorded during and after procedure.

Picture 1. *Case 1,closed angle in gonioscopy and diffuse synechia in LI field.*

Low dose steroid and brimonidine (twice daily) over one week were recommended. Significant opening occurred after ALPI while angle elements could be observed occasionally on gonioscopy (Picture 2). In the follow-up IOP remained at 17-18 mmHg without medical therapy over one year; however, additional treatment (brinzolamide plus timolol maleate, 2x1) was prescribed due to tendency to increase thereafter. During 2-years follow-up, IOP was maintained at 18-19 mmHg with above-mentioned therapy. No progression was observed in glaucoma during follow-up.

Picture 2. *Case 1, significant opening in angle in gonioscopy after ALPI*

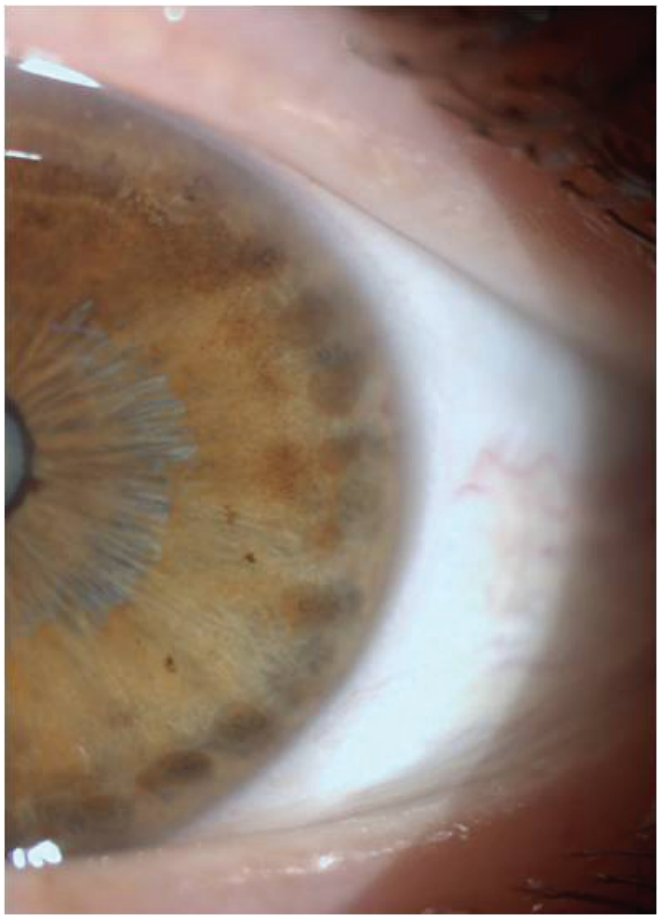
Case 2

Case 2 was sister of first case and 45 years old. The visual acuity was 0.15 with +17 D correction in right eye and 0.15 with +18 D correction in the left eye (Snellen cards). The IOP was 28 mmHg in right eye and 36 mmHg in the left eye. The CCT was measured as 560 µ in right and 570 µ in left eye. c/d ratio was 0.5 in both eyes. Axial length was 15.2 mm. In anterior chamber examination, anteriro chamber was narrow while angle was closed bilaterally in gonioscopy. There was macular folds and loss of foveal reflection in fundus examination. The patient was diagnosed as nanophthalmos and overall 360° ALPI was performed in 2 distinct sessions (180° in each session) in both eyes using same argon laser parameters in first case (Picture 3). No complication was recorded during and after procedure. Low dose steroid and brimonidine (twice daily) over one week were recommended.

In the follow-up IOP remained under control (17-19 mmHg) without medical therapy; however, brinzolamide plus timolol maleate fixed combination (2x1) was prescribed due to tendency to increase after 1.5 years. During 1.5 years of follow-up, IOP was maintained at <20 mmHg in both eyeswith above-mentioned therapy. No progression was observed in glaucoma during follow-up.

Case 3

The case was sister of first two cases and 27 years old. Visual acuity was 0.2 in right eye with +13 D correction and 0.2 in the left eye with 13.5 D correction. The IOP was 18 mmHg in right eye and 16 mmHg in left eye. The CCT was measured as 530 µ in both eyes. Although anterior chambers were narrow, they were wider than her sisters. The c/d ratio was 0.2 bilaterally. Axial length was 16 mm



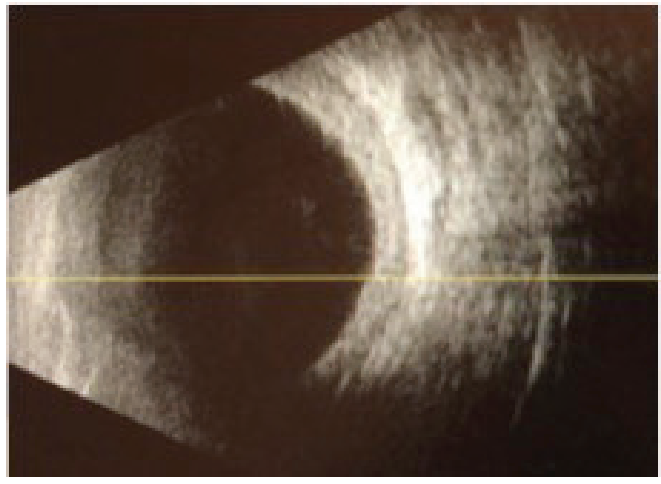
Picture 3. *Case 2, anterior segment image following 180° ALPI.*

at right and 16.2 mm in left eyes. There was maculad folds and loss of foveal reflection in fundus examination. The patient was diagnosed as nanophthalmos. During follow-up, the IOP remained at level of 17-19 mm Hg and no progression to glaucoma was observed.

Case 4

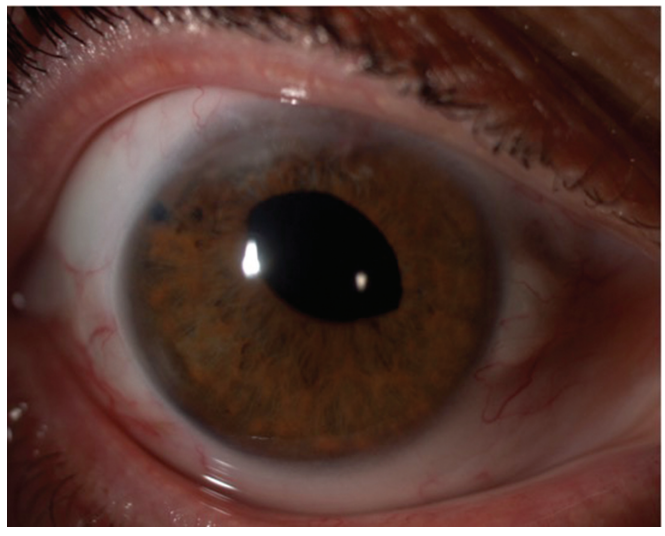
A 39-old man had undergone left trabeculectomy with diagnosis of glaucoma in another facility. In the examination, visual acuity was 0.15 in right eye and "finger counting" at 30 cm in the left eye underwent filtration surgery. The IOP was 32 mmHg with dorzolamide plus timolol maleate fixed combination (2x1), brimonidine (2x1), latanoprost (2x) and acetazolamide (tablet, 2x1) in right eye and 12 mmHg without drug in left eye. Axial length was 17.1 mm in right nd 17.3 m in left eyes (Picture 4). The CCT was 575 µ in right and 570 µ in the left eyes.

In anterior segment examination, anterior chamber was narrow in right eye and there was iridotomy at direction of 1 o'clock. In left eye, anterior chamber was extremely narrow and posterior synechia and iridotomy filed at superior nasal region were observed. There was a shallow bleb formation.



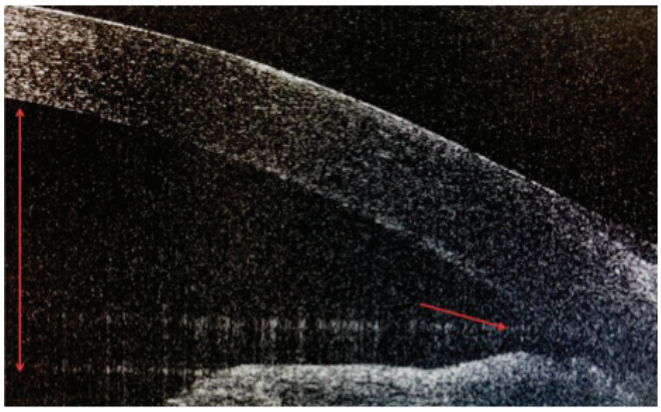
Picture 4. *Case 4, short axial length, thick sclera and choroid in B mode sonography.*

In gonioscopy, angle was closed in right and left eyes. In right, small PAS areas seen barely were present in superior half of eye. The c/d ratio was 0.9-1.0 in right and 1.0 in left eye. There was bilateral loss of foveal reflection and retinal folds. The patient declined intervention in left eye. Phaco plus synechialysis was recommended in right eye. IOL degree was calculated as +45 D for right eye using SRKT and Holladay II formulas. During phaco plus IOL implantation under general anesthesia, it was aimed to release small PAS areas by viscogonio-expansion. No complication was detected during and after surgery. The IOP was measured as 10 mmHg immediately after surgery while visual acuity was 0.15 (Picture 5). In control visit on month 1, IOP was 12 mmHg and visual acuity was 0.2. on anterior segment optic coherence tomography and Pentacam evaluation, marked improvement was detected



Picture 5. *Case 4, anterior segment image immediately after phaco plus viscogonio-synechialysis in right eye; PI at superior temporal region.*

in angle and anterior chamber parameters (Picture 6). In the control on month 4, it was seen that IOP tended to increase; thus brinzolamide plus timolol fixed combination (2x1) was prescribed. The IOP was <20 mmHg without progression.



Picture 6. *Case 4, marked improved in anterior chamber angle and depth on AS-OCT (red arrow).*

DISCUSSION

In cases with nanophthalmos, normal lens size despite small ocular volume causes angle closure at varying degrees due to aggregation in anterior segment. Acut angle-closure can be seen in different decades.[4] However, intermittent angle closure episodes predispose to PAS development. Progression to chronic angle closure can be observed by diffuse PAS development.

In cases with nanophthalmos, LI and ALPI can be beneficial in relieving angle and preventing PAS development.[4] In two cases presented, it was found that LI was performed before presentation to our clinic. However, LI itself may trigger anterior synechia formation in narrow anterior chamber. In fact, we observed wide-based synechia development in first case.

In these cases, LI contributes to relieve angle and to reduce IOP by removing iris from angle and trabecular network. In first 2 cases, we observed that ALPI was extremely helpful in controlling glaucoma and IOP. However, alterations in lens can make IOP and glaucoma control by changing anterior chamber configuration and angle. In such case, additional agents and/or surgical treatment may be needed.

Gonioplasty-assisted lens surgery of filtration surgery can be performed in cases with failure to control IOP despite laser therapy and additional medical therapy. In case 4, there was refractory glaucoma with PAS in right eye. Trabeculectomy was performed previously in the left eye in another facility. IOP was under control with shallow bleb formation in this eye with optic atrophy. However, there was extremely narrow anterior chamber and diffuse synechia. It was planned to perform phaco plus IOL implantaiton with viscogonio-synechialysis in the right eye. The surgery was conducted under general anesthesia; IOP was reduced by mannitol before surgery. No complication was observed during or after surgery. Although different results have been reported in lens extractions performed in nanophthalmic eyes, better results have been achieved after introduction of phaco technique.[5-10] However, there are cases experiencing severe complications.[2] In a study by Steijns et al.[10] phaco was performed in 43 nanophthalmic eyes, no complication was detected in 71.1% of cases. Authors reported that most common complications were uveal effusion by 9.3% and cystoid macular edema by 7%.

It was reported that pars plana vitrectomy (PPV) plus pars plana lensectomy (PPL) can be helpful in controlling glaucoma in nanophthalmic eyes. Zhang et al.[11] performed combination surgery in 21 eyes of 21 patients and reported success rate as 71.4%. Authors reported that complication was observed in 3 cases (uveal effusion, iris hemorrhage and retinal rupture) but no permanent problem threatening vision was seen.

The degree (dioptre) is rather high in IOL implanted to nanophthalmic eyes. Often, IOL>+40 D is required.[9] SRK/T, Hoffer Q and Holladay II formulas are found to be successful for calculation of IOL degree in eyes with microphthalmia or nanophthalmos.[12,13] In our case, IOL degree was calculated as +45 D using SRK/T ve Holladay II formulas. In these cases, pigyback IOL should be avoided since it increases aggregation in anterior segment and may cause pigment dispersion due to friction to iris.[9]

In eyes with nanophthalmos, surgical complications may be associated with complications. It may be possible to experience clinical presentations such as corneal decompensation, severe iritis, anlge-closure glaucoma, uveal effusion, retinal detachment, CME, choroid and/or vitreous hemorrhage and malignant glaucoma in lens surgery or filtration surgery in these cases.[7,10] Uveal effusion is a clinical entity difficult treat and may cause severe loss of vision. Some authors proposed that scleral resection or sclerotomy aiming to attenuate sclera and reduce compression of thick sclera with reduced permeability on vortex veins can be beneficial in cases with nanophthalmus.[14,15]

Singh et al.[2] reported severe loss of vision on 13 of 15 eyes underwent filtration surgery and that complication rate was high in cases with nanophthalmos. The vision was improved in only 3 of 6 eyes underwent lens extraction.

Authors emphasized that laser and additional medical therapy is a good option in angle closure in these cases.. Yalvac et al.[16] reported outcomes in 20 patients underwent trabeculectomy plus inferior sclerotomy. Authors reported that success rate was 85% at year one and 47% at year 5. However, success definition was IOP of 5-22 mmHg without loss of light perception and need for glaucoma surgery; need for additional medical therapy was not included to success criteria. They also reported choroid detachment in 50% of case at early period whereas in 5% of case at late period

In conclusion, glaucoma development should be closely monitored in cases with nanophthalmos. In particular, ALPI is valuable in relieving angle and conrolling IOP. Lens extraction and gonioplasty surgery can provide significant clinical benefit in cases in which laser therapies and medical treatment failed to control IOP. It is essential to monitor patients closely following laser or surgical treatment. Glaucoma control may become challenging by increasing IOP and additional medical treatment and/or surgery may be required. In refractory cases, trabeculectomy can be considered with additional surgical interventions (such as sclerotomy) despite all risks.

REFERENCES

1. Carricondo PC, Andrade T, Prasov L, et al. Nanophthalmos: A review of the clinical spectrum and genetics. J Ophthalmol. 2018:2735465:1-9.
2. Singh OS, Simmons RJ, Brockhurst RJ, et al. Nanophthalmos: a perspective on identification and therapy. Ophthalmology 1982; 89: 1006–12.
3. Stewart DH 3rd, Streeten BW, Brockhurst RJ, et al. Abnormal scleral collagen in nanophthalmos. An ultrastructural study. Arch Ophthalmol 1991;109:1017–25.
4. Ritch R, Chang BM, Liebmann JM. Angle closure in younger patients. Ophthalmology 2003; 110: 1880–9.
5. Utman SA. Small eyes big problems: is cataract surgery the best option for the nanophthalmic eyes? J Coll Physicians Surg Pak. 2013;23(9):653-6.
6. Wu W, Dawson DG, Sugar A, et al. Cataract surgery in patients with nanophthalmos: results and complications. Cataract Refract Surg. 2004;30:584-90.
7. Yuzbasioglu E, Artunay O, Agachan A, et al. Phacoemulsification in patients with nanophthalmos. Can J Ophthalmol 2009;44:534–9.
8. Jung KI, Yang JW, Lee YC, et al. Cataract surgery in eyes with nanophthalmos and relative anterior microphthalmos. Am J Ophthalmol 2012;153:1161– 8.e1
9. Singh H, Wang JC, Desjardins DC, et al. Refractive outcomes in nanophthalmic eyes after phacoemulsification and implantation of a high-refractive-power foldable intraocular lens. J Cataract Refract Surg 2015;41:2394-2402.
10. Steijns D, Bijlsma WR, Van der Lelij A. Cataract surgery in patients with nanophthalmos Ophthalmology. 2013:266-70.
11- Zhang Z, Zhang S, Jiang X, et al. Combined 23-G Pars Plana Vitrectomy and lensectomy in the management of glaucoma associated with nanophthalmos. Ophthalmic Res. 2018;59:37-4.
12- Wladis EJ, Gewirtz MB, Guo S. Cataract surgery in the small adult eye. Surv Ophthalmol 2006;51:153–61.
13. Narváez J, Zimmerman G, Stulting RD, Chang DH. Accuracy of intraocular lens power prediction using the Hoffer Q, Holladay 1, Holladay 2, and SRK/T formulas. J Cataract Refract Surg 2006;32:2050–3.
14. Brockhurst RJ. Vortex vein decompression for nanophthalmic uveal effusion. Arch Ophthalmol 1980; 98: 1987–90.
15. Jin JC, Anderson DR. Laser and unsutured sclerotomy in nanophthalmos. Am J Ophthalmol 1990; 109: 575–80.
16. Yalvac IS, Satana B, Ozkan G, et al. Management of glaucoma in patients with nanophthalmos. Eye (Lond). 2008;22:838-43.