Increased Corneal Curvature with Posterior Nanophthalmos

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ABSTRACT

Posterior microphthalmos is a rare condition which is characterized by the disproportionately small size of the posterior segment of the eye. The corneal diameter, central anterior chamber depth and the lens thickness are all within the normal range.

We present a case of posterior microphthalmos with all normal anterior segment measurements except increased corneal curvature.

Key words: Posterior nanophthalmos. Corneal curvature. Microphthalmos.

INTRODUCTION

Microphthalmos is a structural malformation in which ocular globes exhibit a total axial length at least two standard deviations below age similar controls or more generally globes with an anteroposterior diameter of less than 20 mm in adults and less than 19 mm in a child of one year.

Two clinical sub-types of microphthalmos are identified; one with no other ocular abnormality is called pure microphthalmos or simply nanophthalmos and the one with other ocular or systemic defects is referred to as complex microphthalmos.

Posterior nanophthalmos is another less common variety in which reduced total axial length is associated with normal sized cornea. This condition is associated with high hyperopia and papillomacular folds.

CASE REPORT

A 16-year-old girl born from a non-consanguineous Pakistani family presented in April 2006 with the complaints of poor vision in both eyes since childhood. The patient had an average stature and built and general physical examination showed no systemic abnormality. The palpebral fissures were narrow with large bulging corneas. She was orthophoric with uncorrected visual acuity of counting fingers in both eyes. Cycloplegic retinoscopy with cyclopentolate one meter measured +16.0 DS of hyperopia. The best corrected visual acuity was 6/60 with +11.0 DS in both eyes.

A scan was performed which showed axial length of 14.5 mm and 15 mm in right and left eye respectively. Central lens thickness was 3.5 mm, central anterior chamber depth was 2.75 mm and central corneal thickness was 0.5 mm in both eyes. Intraocular pressures were within the normal range and gonioscopy showed open angles in both eyes. Autorefractokeratometer was used for keratometry. K reading was 48.25 and 51.0 diopters for the right eye and 48.75 and 50.75 diopters for the left eye.

Pupillary reaction was normal bilaterally. Dilated fundus examination showed shot-silk retina with crowded discs (a frequent finding in high hyperopia). Macular reflex was dull in both eyes.

Based on these clinical findings, a diagnosis of posterior nanophthalmos was made. The patient was prescribed hyperopic glasses. There was no other ocular abnormality but as the patients with posterior microphthalmos are prone to develop uveal effusions and non-rhegmatogenous retinal detachment, she was advised for follow-up after every 6 months.

Since autosomal recessive inheritance is reported, other family members were also subjected to the complete clinical examination and were found to be free of ocular and systemic abnormality. The parents were counselled regarding the hereditary nature of the disease and the possibility of later defects in her vision.

DISCUSSION

The term nanophthalmos, derived from a Greek word _nano_, meaning dwarf, is used for an eye, which is grossly normal in form and function but reduced in size. It is a form of pure microphthalmos that results from arrested growth of the globe after closure of the embryonic fissure. Posterior nanophthalmos is a type of nanophthalmos in which there is reduced total axial length (14-20 mm) but the anterior segment of the eye
has normal or near normal dimensions.\textsuperscript{5} In this regard, this case can be considered as posterior nanophthalmos because the anterior segment revealed no signs of retarded development.

Approximately 80\% of subjects with microphthalmos are associated with systemic malformations and more than 100 syndromes displaying microphthalmos have been recognized in the literature.\textsuperscript{2} Other ocular associations include disorders like microcornea, cataract, Aniridia and persistent hyperplastic primary vitreous. Systemic diseases include MIDAS (microphthalmos, dermal aplasia, sclerocornea) syndrome, mucolipidosis III, oculodentodigital syndrome, Aicardi syndrome, CHARGE syndrome, Goldenhar syndrome etc. This patient did not have any systemic abnormality.\textsuperscript{6}

The increase in corneal curvature in cases of posterior nanophthalmos is not a degenerative condition like keratoconus and keratoglobus. Corneal thickness remains normal in posterior nanophthalmos, while keratoconus and keratoglobus are associated with the localized or generalized corneal thinning. It could be the corneal compensation for the extremely short axial length in posterior nanophthalmos. Due to the increased corneal curvature, emmetropia and even myopia have also been reported in some cases of posterior microphthalmos.\textsuperscript{7} The small axial length of the eyeball does not affect the lens growth, which attains a normal size (3 mm) in this case.

Increase in the corneal curvature leads to a normal anterior chamber depth, which serves to prevent the angle closure glaucoma, commonly seen in eyes with small axial lengths.\textsuperscript{8}

As far as the visual acuity is concerned, the high refractive error of +11.0 DS leading to ametropia, amblyopia and dull macular reflex are the leading causes of decreased visual acuity as was in this patient.

Spitznas in 1983, described 5 patients with posterior nanophthalmos with a papillomacular fold and high hyperopia.\textsuperscript{9} Another report indicated elevated papillomacular retinal fold and absence of capillary free zone in patients with posterior microphthalmos. The sclera of nanophthalmic eyes have irregular collagen lamellae, absence of normal elastic fibers and abnormal collagen like deposits.\textsuperscript{10} These changes lead to decrease elasticity which results in altered blood flow dynamics. This can lead to uveal effusion and choroidal detachment especially after surgery. Rarely, nanophthalmos has been reported in association with pigmentary retinopathy but it was absent in this case.

The association of increased corneal curvature with posterior microphthalmos is a natural compensation to prevent complications like angle closure glaucoma. It may also be a compensatory shift towards emmetropia in patients with short axial length.

\textbf{REFERENCES}


