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SHORT COMMUNICATION

Ultrasound biomicroscopy and OCT findings in posterior microphthalmos

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PURPOSE. *Posterior microphthalmos is a rare ocular abnormality. The authors report two siblings with bilateral posterior microphthalmos.*

METHODS. *Two siblings (aged 10 and 13) were admitted with a history of low degree of vision. Ophthalmologic examinations, ocular ultrasonography, ultrasonic biomicroscopy (UBM), and optical coherence tomography (OCT) investigations were carried out and the results were evaluated. All results were also compared with their normal sibling.*

RESULTS. *Two siblings with posterior microphthalmos have normal and near normal anterior segment dimensions, shortened axial lengths (smaller than 17 mm), low vision with high hyperopias, papillomacular folds, and crowded optic discs. OCT investigation showed bilateral papillomacular retinal fold and UBM examination showed that ciliary body moved behind the iris towards the pupilla and the iridocorneal angle anomalies in two siblings. These findings were not observed in the normal sibling.*

CONCLUSIONS. *Posterior microphthalmos can exist in the presence of some ocular anomalies. In this case report, UBM provided new information about the structure of iridocorneal angle and ciliary body. (Eur J Ophthalmol 2008; 18: 479-82)*

KEY WORDS. *Optical coherence tomography, Posterior microphthalmos, Retinal folds, Ultrasound biomicroscopy*

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INTRODUCTION

Microphthalmos is an ocular abnormality of the eye in which all tissues are significantly smaller than normal. It is subdivided into two categories as simple and complex microphthalmos. The complex type is associated with several ocular abnormalities (1). Posterior microphthalmos is another subtype of microphthalmos which is characterized by normal or near normal anterior segment dimensions, yet with abnormal dimension in the posterior segment (2). Several reports have been published on posterior microphthalmos. The most frequent abnormalities reported for posterior microphthalmos are papillomacular folds (3), retinoschisis, retinal dialysis (4), avascular area on the peripheral retina (5), uveal effusion syndrome, pigmentary retinopathy, and crowded optic disc (6). To our knowledge, no study reporting ultrasonic biomicroscopy (UBM) findings of this anomaly has been published in the literature.

In the two siblings covered in this report, we aimed to present all the findings of posterior microphthalmos including ultrasonic biomicroscopy.

Case reports

Two children were admitted to our clinic with a history of low degree of vision. They were two siblings of a Turkish family with three children. They had no systemic anomaly; however, we diagnosed posterior microphthalmos during their ophthalmologic examination. The third sibling and the parents had normal or near normal ocular dimensions. Table I shows the results of the examination of the two siblings.

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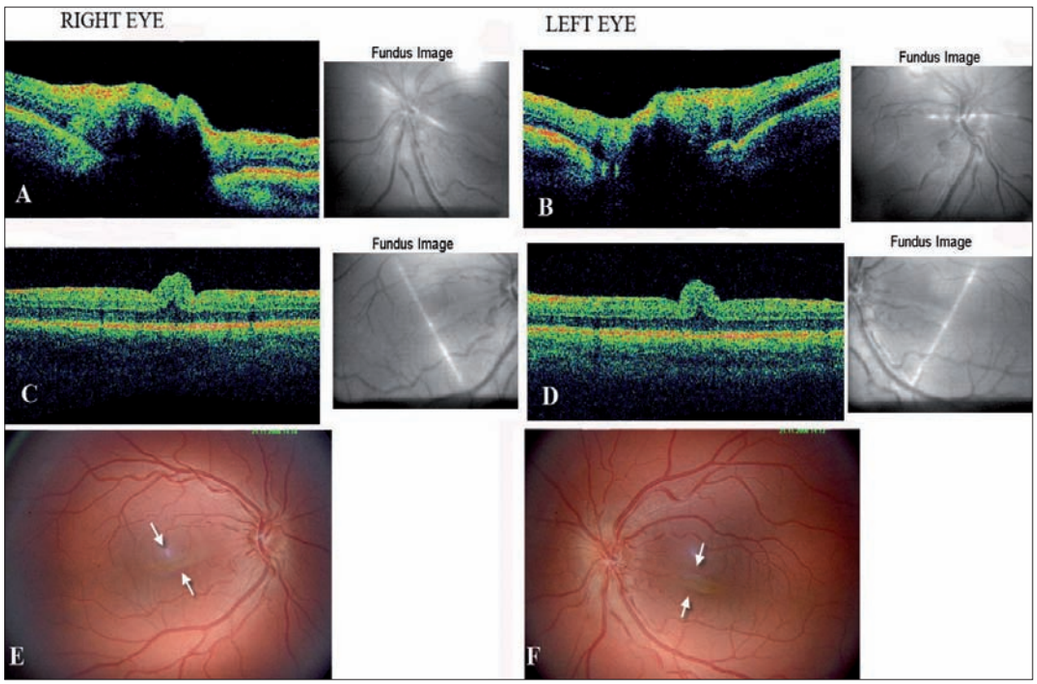

Fig. 1 - *Optical coherence tomography and fundus images of 10-year-old child. Lack of cupping of the optic discs* **(A, B)**, *retinal folds* **(C, D),** *fundus picture, folds are between the arrows* **(E, F)**.

On complete ophthalmic examination, we did not find any evidence of strabismus, nystagmus, or microphthalmos in any of the children. Their slit-lamp examination revealed normal anterior segment findings and dilated fundus examination showed thick retinal folds in the papillomacular bundle including the fovea, slightly elevated papilla with mild blurred border appearance, and normal peripheral retina in both eyes of the children (Figs. 1 and 2).

On ultrasonic investigation, both A- and B-mode ultrasonography revealed obviously shortened eye globe whereas the anterior segment was normal in both eyes of the children.

On optical coherence tomography (OCT) investigation of the thick retinal folds in the papillomacular bundle in both

TABLE I - THE FINDINGS OF THE TWO CASES WITH POSTERIOR MICROPHTHALMOS

		Case 1	**Case 2**
Age, yr/gender		10/Male	13/Female
Best-corrected visual acuity	Right eye	20/200	20/200
	Left eye	20/100	20/200
Refractive error	Right eye	+11.00 +1.00 95°	+15.50 +1.25 95 °
	Left eye	+11.00 +1.00 79°	+16.25 +0.75 163 °
Anterior chamber depth, mm	Right eye	3.00	2.80
	Left eye	3.10	2.85
Lens thickness, mm	Right eye	3.80	4,27
	Left eye	3.90	4.40
Axial length, mm	Right eye	16.68	15.50
	Left eye	16.40	15.40
Intraocular pressure, mmHg	Right eye	16.7	16.9
	Left eye	16.8	17.2
Corneal diameter, mm	Right eye	11.00	11.00
	Left eye	11.00	11.00

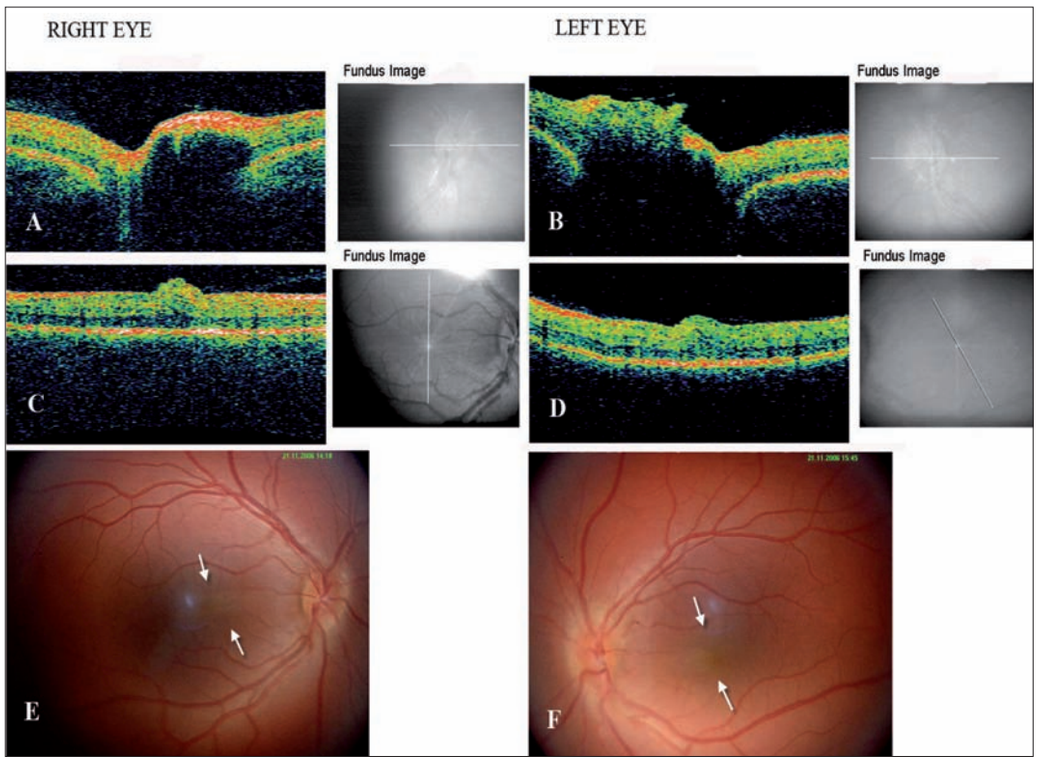

Fig. 2 - *Optical coherence tomography and fundus images of 13-year-old child. Lack of cupping of the optic discs* **(A, B)**, *retinal folds* **(C, D)**, *fundus picture, folds are between the arrows* **(E, F)**.

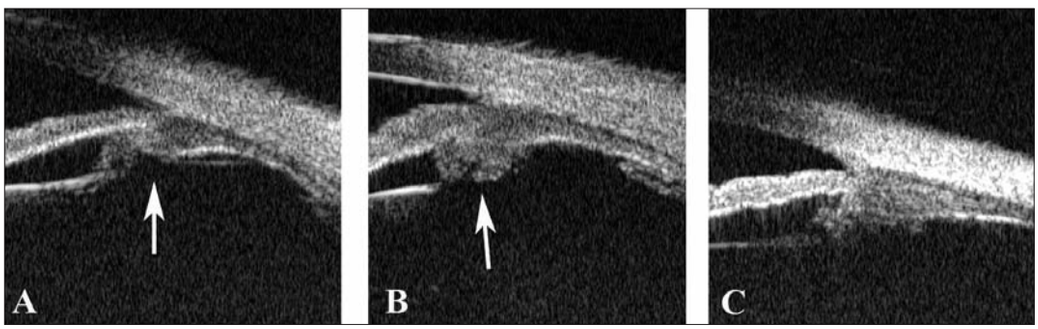

Fig. 3 - *Ultrasound biomicroscopy (UBM) findings of anterior segments in the three siblings.* **(A, B)** *Ciliary body had moved forward behind the iris in two siblings with posterior microphthalmos.* **(C)** *UBM findings of the anterior segments of the normal sibling.*

eyes of the children, we found that the folds included only neural retina without retinal pigment epithelium and choroid. In the section performed parallel to the retinal folds, we observed an increase in the retinal thickness. In the evaluation of the optic disc, the cupping of the disc was absent and the disc was elevated in the OCT sections (Figs. 1 and 2).

On UBM of the anterior segment in both eyes of the children, we observed that the ciliary body moved towards the pupilla behind the iris, and that there was fluid in the suprachoroidal area. These findings were not observed in the normal sibling on UBM (Fig. 3).

DISCUSSION

Posterior microphthalmos with retinal folds is a rare anomaly affecting posterior segment and presents with normal or near normal anterior segment. Usually, there is no associated systemic abnormality. The differential diagnosis from pure microphthalmos is established with the presence of normal findings for the anterior segment. The diagnosis of posterior microphthalmos is further supported by the presence of a normal anterior segment and shortened posterior segment with papillomacular folds including the neurosensory retina.

Posterior microphthalmos is considered as a congenital anomaly in which the sclera is abnormally thickened, limiting the growth of the choroid and retinal pigment epithelium (RPE), while leaving neuroretinal growth and development unrestrained. Thus, papillomacular folds develop and they have been well-documented (4-7). OCT clearly demonstrated these changes in the retina and that only neural retina had the folds without the inclusion of RPE or the choroid.

Based on determinations in some siblings of a family, it had been proposed that this disease had autosomal recessive inheritance (6). In our cases, we observed posterior microphthalmos in two of the three siblings, and the other sibling and the parents had normal ocular findings.

As has been reported, several pathologies can be associated with posterior microphthalmos, such as esotropia, avascular zone on the periphery of the retina without ridge formation (5), uveal effusion syndrome, pigmentary retinopathy (6), retinoschisis, and retinal dialysis (4). We did not observe any of these pathologies in our cases. The most frequently observed findings are papillomacular folds, crowded optic disc, and high hyperopia; we also observed these pathologies in our cases. OCT is the most effective method for determining the retinal changes; it especially helps us differentiate between neural retina and RPE changes. In addition, crowded optic disc can be subjectively determined by OCT.

We could not find any report evaluating the anterior segment of eye with posterior microphthalmos with UBM. In the UBM examination of two children we determined that ciliary body moved behind the iris towards the pupilla and that the trabecular angle was slightly different from the normal. Moreover, we observed some liquid in suprachoroidal space with UBM.

Finally, in posterior microphthalmos not only posterior segment but also ciliary body shows abnormalities which are detectable with UBM. In addition, posterior microphthalmos possibly has an autosomal recessive pattern and further genetic investigation is required.

The authors do not have any proprietary interest.

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