Single lens to lens duplication: The missing link

Rupal Bhatt, MS; Jitendra Jethani, MS; Praveen Saluja, MBBS; Vinay Bharti, MS

Congenital anomalies of the lens include a wide range from lens coloboma to primary aphakia and doubling of lens. There have been few case reports of double lens; the etiology suggested is metaplastic changes in the surface ectoderm that leads to formation of two lens vesicles and hence resulting in double lens. We report a case with bilobed lens, which raises the possibility of explaining the etiology of double lens.

**Key words:** Bilobed lens, lens coloboma, lens duplication

Indian J Ophthalmol 2008;56:67-8

Congenital coloboma of the lens is the most common congenital anomaly of lens. In the region of the coloboma, there is usually some defect in the zonules and hence the resultant coloboma. Cases with duplicated lens that is a single eye carrying two lenses, have been described.1-3 The exact mechanism for this anomaly is not known yet. Our case serves to explain the connection between lens coloboma and duplicated lens.

**Case History**

A young boy, 16 years old, presented with complaints of poor vision in the left eye since childhood. There was no history of any treatment or surgical intervention. In addition, no history pertaining to any trauma was given. Birth history was not significant. On examination, he had a vision of 20/200 in the right eye (RE) and 20/20 in the left eye (LE). The LE was clinically normal with a normal fundus. The RE revealed a horizontally placed bilobed lens sharing a common capsular bag. Zonules were lacking around the fissure between the two lobes [Fig. 1]. Also, there was an inferior iris coloboma. Fundus was normal in both eyes. An ultrasonogram of the right eye [Fig. 2] revealed a bilobed lens in the anterior segment of the eye with normal posterior segment.

**Discussion**

Congenital anomalies of the lens are varied. Partial congenital aphakia is a rare condition in which the lens appears to be partially divided by a deep furrow.1

In the case of congenital duplication of lens reported by

Cornea Unit, M and J Regional Institute of Ophthalmology, Ahmedabad-380 016, India (RB, VB); Pediatric Ophthalmology and Strabismus Clinic, M and J Regional Institute of Ophthalmology, Ahmedabad-380 016, India (JJ); M and J Regional Institute of Ophthalmology, Ahmedabad-380 016, India (PS)

Correspondence to Dr. Jitendra Jethani, Pediatric Ophthalmology and Strabismus Clinic, M and J Regional Institute of Ophthalmology, Civil Hospital, Ahmedabad - 380 016, India. E-mail: xethani@rediffmail.com

Manuscript received: 21.04.06; Revision accepted: 25.04.07

Figure 1: Slit-lamp photograph of the right eye showing horizontally placed bilobed lens. Note the lens is partially separated with a separate nucleus in each lobe

Figure 2: Ultrasonogram of right eye showing bilobed lens

Thakkar et al.2 two lenses were placed obliquely in a straight line with a clear area in between. Absence of zonules and capsule between the two lenses was also noted. Richardson3 reported a case in which the two lenses were asymmetrical, one being larger than the other. The patient also had corneal metaplasia, complete coloboma of iris and fundus.

The mechanism for this anomaly is not known yet. Whether it is part of a phyletogenic spectra or represents development from two embryonically separate lens remains a question. Duke Elder has tried to explain the occurrence of the two entities based on separate embryonic developmental sequences.1 He suggested that the metaplastic changes in the surface ectoderm leads the lens plate to invaginate at two place and forms two lens vesicles which results in double lens.1 This suggests that the lens is divided before the lens starts developing.

Our case has a deep furrow in the center [Fig. 1] with localized absence of zonules. We, therefore, suggest that once the deep furrow is formed due to absence of zonules, it may go further (deep) enough to divide the lens into two distinct parts. Therefore, the absence of zonules is significant and these furrows may be deep enough and progress to duplication. It is a possibility that the lens duplication might be occurring
When the lens vesicle is single and it is the laxity and abnormal stretching which causes the deep furrow and possibly duplication. Duplication may be the extreme form of such a bilobed, furrowed lens. It is interesting to note that both the cases previously reported\textsuperscript{2,3} had lens place in a straight axis at opposite ends. This gives further credence to our theory of the fissure dividing the lens into two and the remaining zonules pulling them away from each other.

Since lens coloboma occurs frequently such deep furrows (bilobed lens) are uncommon. We believe that it represents the missing link between the colobomatosus lens and duplicated lens. We understand that a single case report may not be suggestive of any mechanism but nevertheless it does indicate a possible mechanism of lens duplication.

**References**

1. Elder D. Anomalies of lens. In: System of ophthalmology. Henry Kempton; 1969. p. 688-741.
2. Thakkar H, Singh R. Congenital double crystalline lens. J Cataract Refract Surg 2003;29:405-6.
3. Richardson O. Congenital anomalies of the anterior segment. Trans Can Ophthalmol Soc 1951;14:102-3.

**Atypical presentation and diagnostic pitfalls: A case of rapidly progressive bilateral proptosis in a child aged 18 months**

Subrahmanya K Bhat, MS; Sachet P Shrestha, MS; Rimli Barthakur, MS; Maya Natarajan, DNB

We report an atypical presentation of non-Hodgkin lymphoma (NHL) in a child aged 18 months who presented with rapidly progressive bilateral proptosis. Computerized search using Medline did not reveal a similar presentation of NHL in such a young child. It stresses the need for an early histopathological study including immunohistochemistry and demonstrates the dramatic local response to combined radiotherapy and chemotherapy even in advanced stages without any ocular side-effects.

**Key words:** Bilateral, non-Hodgkin lymphoma, pediatric, proptosis

**Indian J Ophthalmol** 2008;56:68-70

There has been a progressive increase\textsuperscript{1} in the incidence of lymphoma over the years with frequent reports of varying and atypical presentations and a bewildering number of confusing classifications which still do not fit all the cases. This is especially true with respect to non-Hodgkin lymphoma (NHL). This case report discusses the difficulties encountered in diagnosis, the need for an early incisional biopsy and a complete immunohistopathological study with all the markers. It also highlights the fact that lymphomas respond dramatically to combined radiotherapy and chemotherapy.

**Case History**

An 18-month-old female child presented with a history of rapidly increasing prominence of both eyes of two months duration. It was first noticed in the left eye, the right eye was involved two to three days later. The child had occasional fever and did not have any major eye or systemic disease before the onset of prominence of the eyes. Her brother, five years old, was healthy.

The child weighed 9 kg and was afebrile. Examination of the eyes revealed an axial proptosis which measured 22 mm right and 23 mm left eye at an outer intercanthal distance of 82 mm. Lid edema, prominent vessels over the lids and lagophthalmos were more pronounced in the left than the right eye [Fig. 1]. There was gross congestion and chemosis of the conjunctiva and exposure keratitis in the left eye. The conjunctiva was mildly congested in right eye and the cornea was clear. Ocular motility was restricted in all directions of gaze in both eyes. Pupils were normal and reactive in both the eyes and vision appeared normal. Fundus examination of right eye revealed a normal optic disc but the left eye showed an edematous disc. Based on clinical findings, neuroblastoma, granulocytic sarcoma, multifocal eosinophilic granuloma and inflammatory pseudotumor were considered in the differential diagnosis.

Blood examination revealed hemoglobin of 10.9 gm%, erythrocyte sedimentation rate (ESR) of 35 mm, bleeding time of 1 min 25 sec and clotting time of 7 min 02 sec and blood counts were normal. Peripheral blood smear showed microcytic hypochromic picture without any abnormal or atypical cells and adequate platelets. Routine ENT and pediatric referral did not reveal any abnormality. Chest X-ray was normal. Computerized tomography [Fig. 2] showed diffuse infiltration of the orbit and erosion of medial wall of the orbit bordering the nose and lateral wall of the nose bordering the infratemporal fossa on both sides. Adjacent paranasal sinuses were normal. A malignant lesion was suspected and orbital biopsy was advised.

An incisional biopsy was done from both orbits to confirm a single bilateral etiology. Histopathological sections [Fig. 3] study showed solid sheets of predominantly undifferentiated