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Case Study

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CASE REPORT – ISOLATED BILATERAL LENS COLOBOMA WITH BERGMEISTER PAPILLA

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ABSTRACT

Purpose: Lens coloboma is a rare congenital disorder of crystalline lens characterized by notching of the equator of the lens. Coloboma may associated with other ocular malformation or in isolation. The zonules are usually absent or maldeveloped in the region of lens coloboma, so the purpose of this case report is to find out other pathologies of this coloboma with other ocular structure.

KEYWORDS: lens coloboma, bergmeister papilla, Coloboma.

INTRODUCTION

Coloboma of lens usually occur unilaterally, and if bilaterally, is symmetrical coloboma. It is an autosomal dominant condition occurring in the 4th month of development. The characteristic notching of lens tissue as a result of an incomplete formation of lens at the equator due to incomplete closure of fetal fissure. The zonules may absent or maldeveloped in the lens coloboma regions. Coloboma usually associated with iris coloboma, choroidal coloboma, Disc coloboma, localized or total cataract.

CASE REPORT

A 33 year old man was referred to our hospital with poor visual acuity in his both eyes since his childhood. His vision in RE is 6/60 and with glass+1.00D (180) it improve up to 6/12, in LE his vision is 6/36 and with glass +1.00D spherical correction it improve up to 6/24. IOP in RE 12.2mmhg and in LE 15.9 mmhg which is totally normal. Slit lamp examination after pupillary dilatation revealed found notching of crystalline lens on nasal side in both eyes.(FIg1, Fig2) His iris is normal no iris coloboma found. In fundus examination there is no presence of choroidal coloboma and Disc coloboma. Central fundus and peripheral fundus are within normal limit. Presence of whitish remnants of hyaloid artery was seen in both eyes around disc which is called bergmeister papilla. (FIG 3, FIG 4)

In Optical Coherence tomography macula of both eyes are normal, Apart from ocular defects patient does't have any systemic illness like Marfan syndrome and Marshall syndrome. 2D echo also revealed no anomalies.

At the end of total ophthalmic examination the patient was diagnosed with congenital bilateral lens coloboma with bergmeister papilla without any systemic illness.

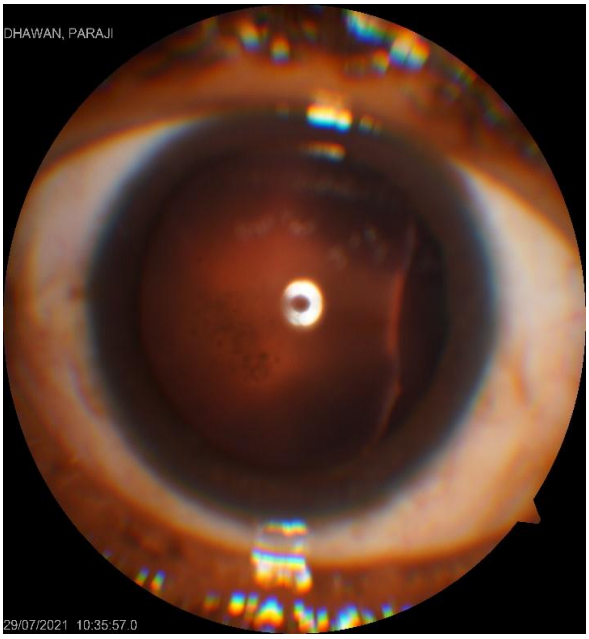

FIG -1 Right eye nasal lens coloboma.

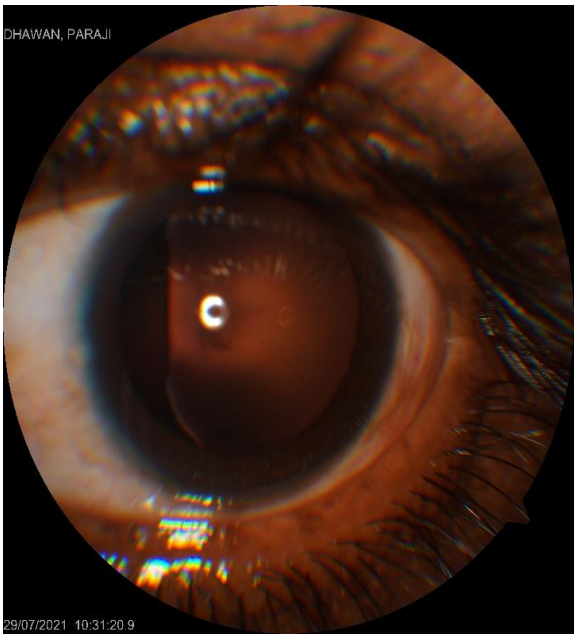

FIG -2 left eye nasal lens coloboma.

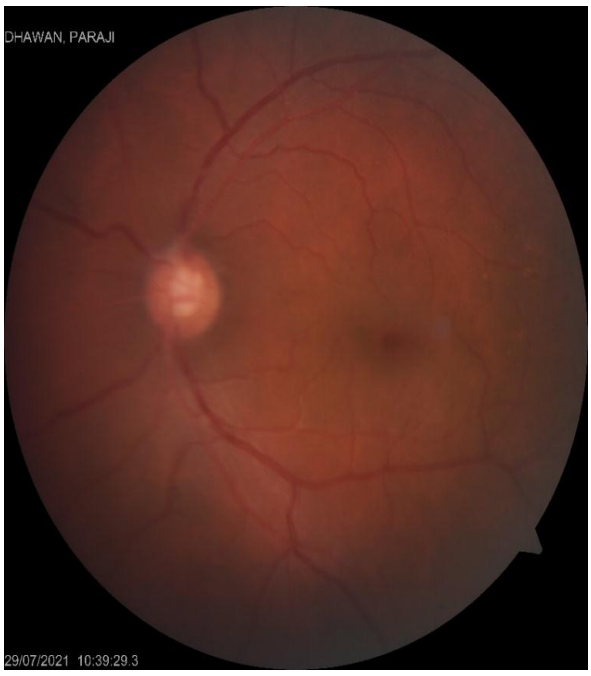

FIG 3 - Right eye fundus showing Bergmeister papilla around optic disc.

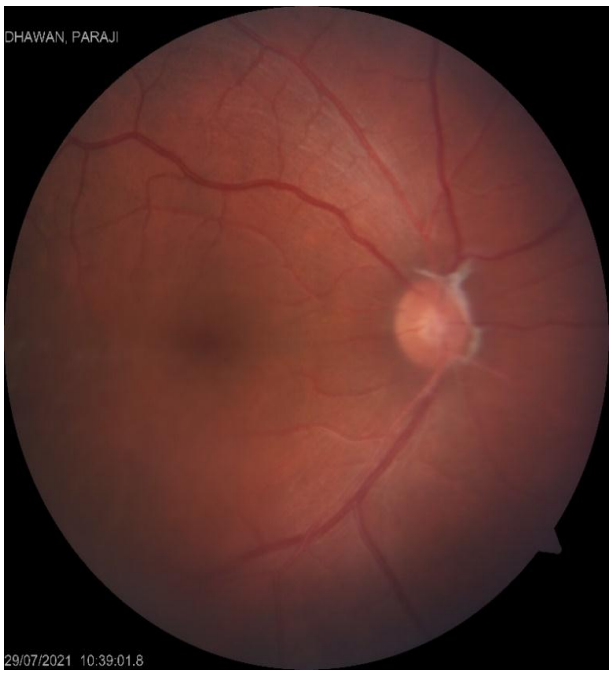

FIG4 Left eye fundus showing Bergmeister papilla superonasal to optic disc.

DISCUSSION

The eye develops from embryological layers, includind the neural ectoderm, neural crest, and the surface ectoderm. During the 4th week of embryogenesis, the optic vesicle derived from the neural ectoderm invaginates and fuses through the lower nasal margin, leading to the formation of optic cup. Any defects in closure of fissures at this developmental stage leads to the occurrence of coloboma.[1] The lens coloboma is extremely rare, associated with absence of zonules or weak zonule at that site. The zonules fail to form in the anterior secondary vitreous or the marginal bundle of Druault during the 3rd -4th month of gestation, due either to the failure of condensation and differentiation of the vitreous substance or to the failure of the lens to induce such changes.[2]

Lens coloboma usually associated with iris coloboma, choroid coloboma, disc coloboma, and cataract in unilateral or bilateral cases.

crystalline lens coloboma most of the time reported with marfan syndrome, congenital ptosis, cardiac pathology, retinal detachment, superior rectus palsy, and optic nerve hypoplasia with orbital hemangioma. In our case apart from a refractive error and low visual acuity, and bergmeister papilla, we did not found any other ocular abnormalities.

CONCLUSION

The case report is unique as there is bilateral nasal lens coloboma with bergmeister papilla without any systemic illness.

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