Pediatric idiopathic macular hole – A case report and review of literature

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Macular hole in the pediatric age group is usually post-traumatic unlike, the adult counterpart. Herein we describe a case of idiopathic macular hole occurring in a 9-year-old male who presented to us with complaints of diminution of vision in OD (oculus dextrus) for 2 months. The child underwent 25-gauge pars plana vitrectomy (PPV) with internal limiting membrane (ILM) peeling. On follow-up his best-corrected visual acuity improved with type 1 closure of macular hole.

Key words: Macular hole, nontraumatic macular hole, pediatric idiopathic macular hole

A macular hole is a full-thickness defect in the foveal center with variable prevalence. Incidence of macular hole in adult population was 3.3 per 1000 in the Baltimore Eye study[1] Macular holes may be primary or secondary. Most macular holes are primary due to abnormal vitreomacular traction. Idiopathic macular holes commonly occur in the 6th–7th decade of life.[2]

Macular hole in a young patient is rare and is usually associated with blunt trauma.[3] Moreover, causes of nontraumatic macular hole in a young patient may be related to Coats disease,[4,5] retinal dystrophies and degenerations[6,7] like retinitis pigmentosa, X-Linked retinoschisis and Best disease, retinopathy of prematurity,[8] Bartonella neuroretinitis,[9-11] Juvenile idiopathic epiretinal membrane,[12] incompletely regressed Bergmeister papilla,[13] laser injury,[14] choroidal coloboma,[15] and Idiopathic cavitary maculopathy.[16]

Idiopathic macular hole in a young patient has only been reported infrequently in the literature. We hereby report a case of idiopathic macular hole in a 9-year-old boy, who underwent successful 25-gauge pars plana vitrectomy (PPV) with internal limiting membrane (ILM) peeling and also review the existing literature.

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Case Report

A 9-year-old boy presented with diminution of vision in OD (oculus dextrus) of 2 months duration. The child was brought to our centre by his father who was a reliable informant and further history was elicited by both the father and the child. There was no history of trauma or night blindness. Birth history, family history, and systemic history were insignificant. The child was emmetrope and his best-corrected visual acuity (BCVA) was 20/120 and 20/20 in OD and OS (oculus sinister) respectively. Slit-lamp examination revealed anterior chamber to be unremarkable in oculus uterque (OU) with no signs of trauma or inflammation. In particular, there was an absence of anisocoria, sphincteric tear, iridodialysis, angle recession, and lens subluxation. There were no other objective signs of trauma. Intraocular pressure was 14 and 16 mm of mercury in OD and OS respectively. Dilated fundus examination of OD [Fig. 1a] revealed a full thickness macular hole [FTMH, arrow Fig. 1a] with regular margins, cuff of fluid, and no posterior vitreous detachment (PVD). There were no signs of trauma, inflammation, or retinal dystrophy. Optic disc was normal and symmetric in appearance, there were no retinal pigment epithelial changes and no peripheral dialysis or tear. Fundus of OS was unremarkable [Fig. 1c].

The child was co-operative for investigations. Swept-source optical coherence tomography of OD [SSOCT, Fig. 1b] confirmed the presence of an FTMH (with minimal hole diameter of 507 microns) with cystoid changes at the edges. No vitreomacular traction or posterior vitreous detachment was noted. OS SS-OCT was normal [Fig. 1d].

Ultra-wide field fluorescein angiography revealed [Fig. 2a] hyperfluorescence (window defect) corresponding to the macular hole in OD [Fig. 2b]. No peripheral abnormalities were found on fluorescein angiography, which could have contributed towards formation of macular hole. OS was normal on ultra-wide-field imaging [Fig. 2c and d] Based on clinical picture, a diagnosis of OD idiopathic macular hole was made.

Following a written informed consent from the parents, the patient underwent 25-gauge PPV under general anesthesia. After core vitrectomy, triamcinolone assisted posterior vitreous detachment (PVD) was done. PVD induction was routine for the patient's age and was followed by Brilliant blue dye assisted ILM peeling. Fluid air exchange and tamponade with 25% sulfur hexafluoride were done. Postoperatively, the child was advised facedown position for 2 days.

At postoperative week one, BCVA was 20/60 and hole had closed [Fig. 3a]. SSOCT showed type 1 hole closure, while ellipsoid zone was discontinuous [Fig. 3b]. BCVA improved to 20/40 at 2 months and SSOCT showed small defect of ellipsoid zone [Fig. 3c and d]. In addition, dissociated nerve fibre layer

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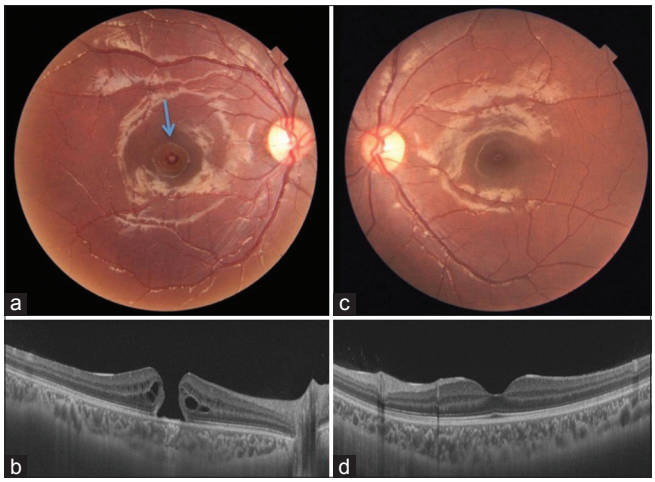

Figure 1: Colour fundus photograph of OD showing the presence of a full-thickness macular hole (FTMH) with regular margins and no posterior vitreous detachment (PVD) (a). Swept-source optical coherence tomography (SS-OCT) confirmed the FTMH with cystoid changes at the edges and no PVD (b). Colour fundus photograph of OS showing normal foveal reflex (c) and SS-OCT showing normal foveal contour (d)

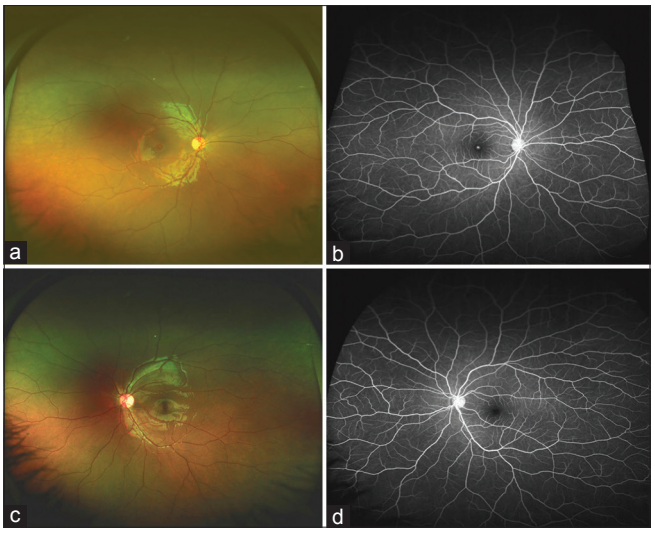

Figure 2: Ultra-wide field fundus image of OD showing no peripheral lesion (a). Ultra-wide field fluorescein angiography showing central hyperfluorescence (window defect) corresponding to the macular hole in OD (b). Ultra-Wide field fundus image (c) and Fluorescein angiography of OS (d) was unremarkable

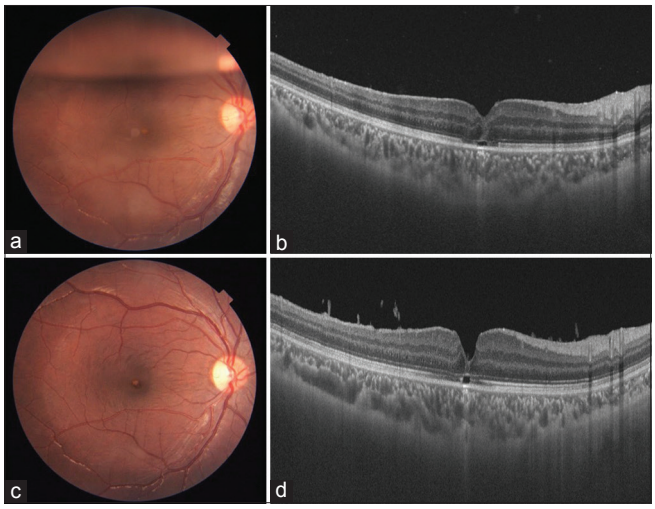

Figure 3: Postoperative week 1 colour photo and SS-OCT of OD showing type 1 closure of the macular hole with the discontinuous ellipsoid zone (a and b). Postoperative month 2 colour photo and SS-OCT showing a small residual defect in the ellipsoid zone and dissociated nerve fibre layer (c and d)

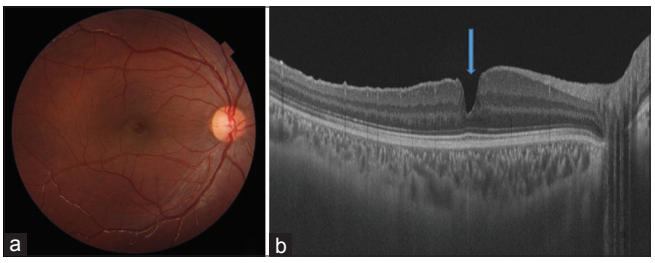

Figure 4: Postoperative year 2 colour photo and SS-OCT showing type 1 macular hole closure with the restoration of the ellipsoid zone with a deep foveal contour (a and b)

was seen. The clinical picture was maintained at 2 years follow up [Fig. 4] with the fellow eye showing no FTMH either. The patient was advised regular follow up.

Discussion

Macular holes in the pediatric age group are usually secondary to blunt trauma.[3] Truly idiopathic macular holes in the pediatric age group are extremely rare.

On extensive research using the keywords Idiopathic macular hole or pediatric macular hole in PubMed, we came up with 18 pediatric eyes with a nontraumatic macular hole of which 15 had a secondary nontraumatic cause. Only three pediatric eyes satisfied the diagnosis of idiopathic macular hole. In each case, a surgical closure was attempted with the outcome of the macular hole being closed in the postoperative period.

A summary of all such cases with relevant findings and final outcomes are listed in Table 1.

The review of literature reveals that only one truly idiopathic case of a pediatric macular hole has been previously reported. Our case adds to the existing literature on an idiopathic macular hole in the pediatric age group and allows us the advantage of having a long-term follow-up for the patient. Proposed mechanism of hole formation in this child remains truly an enigma with our proposed hypothesis being a congenital abnormality in the development of the fovea In cases of traumatic macular hole, spontaneous closure[19] may occur. The child in our case had presented at the cusp of amblyogenic age. Hence an early intervention in the form of surgery was offered that resulted in good outcome.

Early surgery although addresses the problem of amblyopia, has its own pitfalls as PVD induction in the young may be challenging. Inadvertent iatrogenic retinal tears, vitreous

Table 1: Review of literature on cases of Idiopathic macular hole in pediatric age group

Author, Year	Nakano *et al.*[13] 2005	Manayath *et al.*[17] 2018	Lim *et al.*[18] 2018	Our case
Age at presentation (in years)/Gender	10/F	8/M	10/F	9/M
Duration of symptoms	3 days	2 weeks	Not mentioned	2 months
Observation period	1 month	2 months	None	None
Initial vision at presentation and Fall in vision on observation	20/30 to 20/120	20/80 to 20/200	No Observation At presentation 20/60	No observation At Presentation 20/120
Fundus findings	FTMH Fibrous membrane on superior peripapillary retina No peripheral lesion	FTMH No PVD/VMA/ERM, No RPE changes No peripheral lesion	FTMH No PVD/VMA/ERM, No RPE changes No peripheral lesion	FTMH No PVD/VMA/ERM, No RPE changes No peripheral lesion
Hole size	Not reported	Not reported	365 microns	507 microns
Surgery	PPV with ICG assisted ILM peeling with 20% SF6 Tamponade	PPV with BBG assisted ILM peeling with 20% C3F8 tamponade	PPV with BBG assisted ILM peeling with 26% SF6 tamponade	PPV with BBG assisted ILM peeling with 25% SF6 tamponade
Intra-op	Fibrous membrane removed - Tightly adhered	Abnormally tight vitreoretinal adherence PVD just beyond vascular arcades	Induction of PVD easy	Induction of PVD routine
Outcome	Macular hole closed Mild atrophic changes of RPE	Macular hole closed Mild Foveal thinning with ellipsoid zone discontinuity	Macular hole closed Small defect at photoreceptor level	Macular hole closed Complete restoration of ellipsoid zone
Follow-up period	15 months	6 months	4 months	2 years
Final visual outcome	20/60	20/60	20/20	20/40
Proposed pathogenesis	Contraction of peripapillary fibrous membrane due to incomplete regression of Bergmeister papillae	Abnormally tight vitreoretinal adhesion in mid peripheral region causing tangential traction over the fovea resulting in development of FTMH	Truly idiopathic	Truly idiopathic

haemorrhage, and cataract formation are the other major adverse effects of surgery.

Conclusion

To conclude, idiopathic macular holes in the pediatric age group are extremely rare. Observation of up to 2 months showed no benefit in the previously published reports. An option of early surgery could be considered following adequate discussion with the parents regarding the pitfalls of both observation and surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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