DJO

Vol. 30, No. 4, April-June 2020

Case Report

Idiopathic Epiretinal Membrane in Young: A Rare Entity

Aeshvarya Dhawan, Ashish Kakkar

Department of Ophthalmology, Shri Guru Ram Rai Institute of Medical and Health Sciences and Shri Mahant Indiresh Hospital, Dehradun, Uttarakhand, India

Abstract

Epiretinal membrane is usually seen commonly in older age group individuals secondary to intra ocular surgery, trauma, retinal vascular diseases and diabetes mellitus. Here in this case report we describe an unusual case of idiopathic Epiretinal membrane in young individual and importance of fundus examination in patients of microtropia to rule out any macular pathology.

Delhi J Ophthalmol 2020;30;57-60; Doi http://dx.doi.org/10.7869/djo.548

Keywords: *Epiretinal membrane, amblyopia, microtropia.*

Introduction

Epiretinal membrane (ERM) is a disorder of the vitreomacular interface which involves both the perimacular and macular regions. It can cause visual impairment, metamorphopsia, micropsia and occasionally monocular diplopia.[1] ERMs are known to cause wrinkling and distortion of macular surface which is caused by retinal cell proliferation.[2] ERM progression is generally slow but some patients need a treatment for visual complains due to tangential tractions on macula caused by fibrotic membranes.[3] In adults, ERMs are usually associated with ocular disease, idiopathic, or after retinal reattachment surgery.[2] Epiretinal membranes in young age group are rare. Common causes of ERMs in younger age group are trauma, retinal vascular disease, uveitis or tumors.[2]

The prevalence of ERMs according to Beaver dams eye study [BDES] was 28% for age 63 – 74 years and 53% for those aged 85 years and more.[4] The prevalence of ERM in subjects less than 20 years of age is around 1 in 20,000. Xiao, et al. had reported the risk factors of ERM in a meta-analysis and they had found only female gender and ageing as ERM associated risk factors.[5] Several other population based studies investigated epidemiology of ERMs in various countries like japan, Australia, china, Singapore. They concluded variable consideration among different races and regions of the world.[5]

Almost all the studies dedicated to ERMs have shown incidence more in the patients aged 50 years or more.[6,7] Very few reports document ERM in children.

Here we report a rare case of idiopathic ERM in a young male child which is contrary to usual age of presentation.

Case Report

We describe a case of 9 year old male who came to our eye OPD with complaints of decreased visual acuity in right eye since 3 years, and outward deviation of right eye since 2 to 3 years. There is no history of any ocular trauma, surgery or any ocular illness. Visual acuity was 5/60 and N36 OD and 6/6 and N6 OS on Snellen's visual acuity chart. On evaluation, there was exotropia of 10 PD in right eye. Right eye fixation was central, unsteady and not maintained. Ocular movements were free and in full range. Anterior segment evaluation of right eye revealed relative afferent pupillary defect. On posterior segment evaluation of right side, vertical cup disc ratio was 0.5:1, neuro retinal rim was pink and thinning in temporal quadrant was noted. Foveal reflex was dull and there was greyish coloured membrane in macular area with internal limiting membrane striations with straightening of vessels of temporal arcade, suggestive of right ERM (pre macular fibrosis with vascular distortion involving macula with macular oedema). On evaluation left eye was within normal limits. Fundus photography and spectral domain optical coherence tomography (SDOCT) was done. The image quality was suboptimal as the child was having no fixation from right eye (Figure 1). Also acquiring high quality SDOCT was difficult because the optics used are based on assumptions about the dynamic axial length, refractive error, corneal curvature, and astigmatism of particular age group. SDOCT showed a membrane covering the macular area. There was increased thickness of macular area with normal integrity of photoreceptor complex (Figure 2).

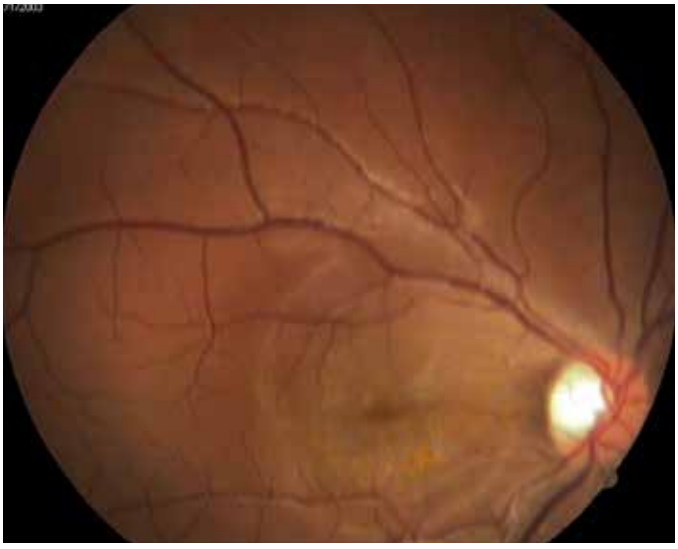

Figure 1: *Right eye fundus photograph showing grade 1 ERM.*

Discussion

ERMs that occur without any antecedent ocular conditions or surgical procedures are termed idiopathic or primary ERMs. Those associated with other eye diseases (e.g., retinal vascular occlusion and diabetic retinopathy), trauma or surgery are referred to as secondary ERM.[8]

A clinical grading system was proposed by Gass to describe the different stages of the disease. In Grade 0 (also termed cellophane maculopathy), a translucent membrane with no underlying retinal distortion is observed. These membranes are asymptomatic, and the diagnosis is often an incidental finding during routine examination. An ERM associated with irregular wrinkling of the inner retina is classified as Grade 1. When this involves the fovea, patients often complain of distorted or blurred vision. Eccentric Grade 1 ERMs that do not involve the fovea may be asymptomatic. A Grade 2 ERM is characterized by an opaque membrane causing marked full thickness retinal distortion and obscuration of underlying vessels.[9]

Various epidemiologic studies have shown the prevalence of ERM varies from 2.2% to 28.9% depending on the population being studied. According to different studies results it has been estimated that 30 million people of advanced age in the US have an ERM in at least one of the eye.[10]

In our case the child had RAPD in right eye, the possible cause for which cannot be explained as optic nerve head was healthy on examination.

It is now accepted that there is a much lower incidence of ERM in children than in adults, as paediatric ERMs typically develop secondary to another pathology such as trauma or inflammation rather than the idiopathic form associated with PVD common in adults[2], but contrarily here in this case child had ERM of idiopathic origin.

The opaque PMF form of ERM is more prevalent in children while the translucent CMR ERM is observed more frequently in adults.[2,3,11] In our case, the child had Grade 1 ERM.

Clinical evaluation also suggests a higher incidence of ERM adherence to Vessels, and infiltration into the posterior vitreous with traction and subsequent vessel distortion in the paediatric population.[12,13] In our case also, there was ERM adherence to vessels and straightening of temporal vessels in right sided fundus.

The major symptoms of ERM are reduced visual acuity, metamorphopsia, and aniseikonia. Metamorphopsia, which at present is commonly assessed with the M-CHARTS[14,15], is measured in the horizontal and vertical directions. Fundus photography and optical coherence tomography, both are used in establishing the diagnosis of ERMs.

One of the differential diagnosis of Idiopathic ERM is Combined Hamartoma of Retina and Retinal Pigment Epithelium (CHR-RPE). But due presence of only ERM and no papillary component or greyish pigmented mass, Idiopathic ERM was kept as first differential diagnosis.

Suh et al found a statistically significant correlation between poor visual outcome and inner segment (IS)/ outer segment (OS) junction disruption. Other predictors of post-operative functional improvements include cone outer segment tip line disruption and photoreceptor outer segment length. It is believed that the tractional force from the ERM may cause irreversible damage to the rods and cones that can be diagnosed preoperatively on SDOCT.[16]

In our case the inner segment (IS)/outer segment (OS) junction was maintained.

The indications for ERM surgery are patient-reported symptoms of decreased VA with or without metamorphopsia. Although surgery is usually reserved for patients with a reduced acuity to ≤20/60, improvements in surgical techniques have led to patients undergoing surgery with better VA if disturbed by symptoms of metamorphopsia or diplopia and also for Occupational reasons.[17,18,19,20]

The surgical management includes a standard pars plana vitrectomy with ERM peeling facilitated by use of vital dyes viz, brilliant blue stain, indocyanin green (ICG) and trypan blue.[21,22,23]

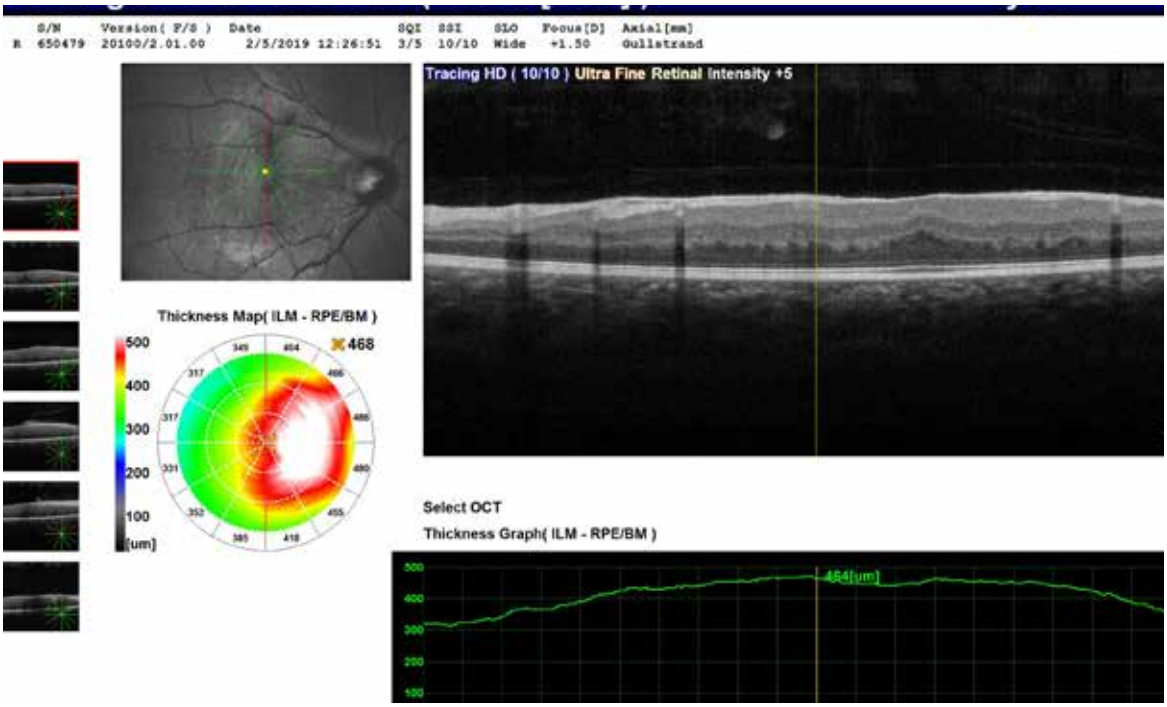

Figure 2: *Right eye SDOCT showing ERM with increased macular thickness.*

Internal limiting membrane peeling can also be done for complete myo- fibroblast removal which helps in decreasing reccurrence.[24,25,26,27] Few reports suggest that ILM peeling leads to damage to Muller cell foot plates, without changing rates of reccurrence.[28,29]

Banach et al also examined the surgical and nonsurgical visual outcomes of young subjects with idiopathic ERMs. They concluded that young subjects with idiopathic macular ERMs and a presenting visual acuity of 20/50 or better had a favourable visual outcome with observation. Subjects with an initial vision of 20/60 or worse, or those who had a visual decrease to < or =20/60 had significantly improved visual acuity after vitrectomy. But keeping the fact in mind that recurrence of ERMs is relatively high after surgery.[30]

Multiple regression analysis has shown that a higher preoperative VA is associated with a higher final acuity at 6 and 12 months, even though the percentage improvement may be less.[31,32]

Visual acuity in our case was 5/60 and also the long duration of symptoms (3 yrs.) indicate a guarded visual prognosis.

The child's attendants were not willing to undergo surgical intervention due to chances of amblyopia and no assured post-operative visual prognosis.

Conclusion

ERM in a young individual is a rare presentation and should be evaluated for all the associated causes and managed timely so as to prevent sensory amblyopia. This case report also highlights the importance of fundus examination in patients having microtropia which may be due to any macular pathology as in our case.

References

1. Ayse Gul Kocak Altintas, Cagri Ilhan. "Epiretinal Membrane". Acta Scientific Ophthalmology 1.3 (2018): 18-23.
2. Khaja HA, McCannel CA, Diehl NN, Mohney BG. Incidence and clinical characteristics of epiretinal membranes in children. *Arch Ophthalmol,* 2008; 126:632–636.
3. Mitchell P, Smith W, Chey T, et al. Prevalence and associations of epiretinal membranes. The Blue Mountains Eye Study, Australia. *Ophthalmology,* 1997; 104:1033–1040.
4. Meuer SM, et al. The Epidemiology of Epiretinal Membranes and Posterior Vitreoretinal Traction as Detected by SD-OCT: The Beaver Dam Eye Study. *Investigative Ophthalmology & Visual Science,* 2014; 55:688.
5. Xiao W, Chen X, Yan W, et al. Prevalence and risk factors of epiretinal membranes: a systematic review and meta-analysis of population based studies. *BMJ Open,* 2017; 7:e014644
6. Jacobsen CH. Epiretinal membranes. *Optom Clin,* 1996; 5(1):77-94.
7. SR Dawson, M Shunmugam and TH Williamson.
8. Bu SC, Kuijer R, Li XR, et al. Idiopathic epiretinal membrane. *Retina,* 2014; 34:2317–35.
9. Gass JDM. Stereoscopic atlas of macular diseases: diagnosis and treatment. 3rd ed. *St. Louis: Mosby,* 1987.
10. Klein R, et al. "The epidemiology of epiretinal membranes". *Transactions of the American Ophthalmological Society,* 1994; 92:403-425.
11. Fraser-Bell S, Ying-Lai M, Klein R, Varma R. Prevalence and associations of epiretinal membranes in latinos: the Los Angeles Latino Eye Study. *Invest Ophthalmol Vis Sci,* 2004; 45:1732–1736.
12. Joshi MM, Ciaccia S, Trese MT, Capone A Jr. Posterior hyaloid contracture in pediatric vitreoretinopathies. *Retina,* 2006; 26:38-41.
13. Benhamou N, Massin P, Spolaore R, et al. Surgical management of epiretinal membrane formation. *Graefes Arch Clin Exp Ophthalmol,* 2009; 247(7):865-83.
14. Matsumoto C, Arimura E, Okuyama S, Takada S, Hashimoto S, Shimomura Y. Quantification of metamorphopsia in patients with epiretinal membranes. *Invest Ophthalmol Vis Sci,* 2003; 44:4012–6.
15. Kinoshita T, Imaizumi H, Okushiba U, Miyamoto H, Ogino T, Mitamura Y. Time course of changes in metamorphopsia, visual acuity, and OCT parametersafter successful epiretinal membrane surgery. *Invest Ophthalmo Vis Sci,* 2012; 53:3592-7.
16. Rothman LA, Folgar FA,Tong AY,Toth CA. Spectral Domain Optical Coherence Tomography Characterization of Pediatric Epiretinal Membranes. *Retina. Author manuscript,* 2014; 34(7):1323–1334.
17. Pesin SR, Olk RJ, Grand MG, et al. Vitrectomy for premacular fibroplasia. Prognostic factors, long-term follow-up, and time course of visual improvement. *Ophthalmology,* 1991; 98(7):1109–14.
18. Massin P, Allouch C, Haouchine B, et al. Optical coherence tomography of idiopathic macular epiretinal membranes before and after surgery. *Am J Ophthalmol,* 2000; 130(6):732–9.
19. Wong JG, Sachdev N, Beaumont PE, Chang AA. Visual outcomes following vitrectomy and peeling of epiretinal membrane. *Clin Exp Ophthalmol,* 2005; 33(4):373–8.
20. Scheerlinck LM, van der Valk R, van Leeuwen R. Predictive factors for postoperative visual acuity in idiopathic epiretinal membrane: a systematic review. *Acta Ophthalmol,* 2015; 93(3):203–12.
21. Haritoglou C, Eibl K, Schaumberger M, et al. Functional outcome after trypan blue-assisted vitrectomy for macular pucker: a prospective, randomized, comparative trial. *Am J Ophthalmol,* 2004; 138(1):1–5.
22. Haritoglou C, Gandorfer A, Schaumberger M, et al. Trypan blue in macular pucker surgery: an evaluation of histology and functional outcome. *Retina,* 2004; 24(4):582–90.
23. Kwok AK, Lai TY, Li WW, Yew DT, Wong VW. Trypan blue- and indocyanine green-assisted epiretinal membrane surgery: clinical and histopathological studies. *Eye,* 2004; 18(9):882–8.
24. Bovey EH, Uffer S, Achache F. Surgery for epimacular membrane:impact of retinal internal limiting membrane removal on functional outcome. *Retina,* 2004; 24(5):728–35.
25. Park DW, Dugel PU, Garda J, et al. Macular pucker removal with and without internal limiting membrane peeling: pilot study. *Ophthalmology,* 2003; 110(1):62–4.
26. Sandali O, El Sanharawi M, Basli E, et al. Epiretinal membrane recurrence: incidence, characteristics, evolution, and preventive and risk factors. *Retina,* 2013; 33(10):2032–8.
27. Ahn SJ, Ahn J, Woo SJ, Park KH. Photoreceptor change and visual outcome after idiopathic epiretinal membrane removal with or without additional internal limiting membrane peeling. Retina 2014;34(1):172–81.
28. Lim JW, Cho JH, Kim HK. Assessment of macular function by multifocal electroretinography following epiretinal membrane surgery with internal limiting membrane peeling. *Clin Ophthalmol,* 2010; 4:689–94.
29. Kifuku K, Hata Y, Kohno RI, et al. Residual internal limiting membrane in epiretinal membrane surgery. *Br J Ophthalmol,* 2009; 93(8):1016–19.
30. Banach MJ, Hassan TS, Cox MS, et al. Clinical course and surgical treatment of macular epiretinal membranes in young subjects. *Ophthalmology,* 2001; 108:23–26.
31. Inoue M, Morita S, Watanabe Y, et al. Preoperative inner segment/outer segment junction in spectral-domain optical coherence tomography as a prognostic factor in Epiretinal membrane surgery. *Retina,* 2011; 31(7):1366–72.
32. Shiono A, Kogo J, Klose G, et al. Photoreceptor outer segment length: a prognostic factor for idiopathic epiretinal membrane surgery. *Ophthalmology,* 2013; 120(4):788–94.

Cite This Article as: *Dhawan A, Kakkar A. Idiopathic Epiretinal Membrane in Young: A Rare Entity.*

Acknowledgments: Nil

Conflict of interest: None declared

Source of Funding: None

Date of Submission: 12 July 2019
Date of Acceptance: 11 December 2019

Address for correspondence

Ashish Kakkar **MD Ophthalmology**
Department of Ophthalmology,
Shri Guru Ram Rai Institute of Medical and Health Sciences and Shri Mahant Indiresh Hospital, Dehradun - 248001, Uttarakhand, India
Email id: ashishkakkar2007@gmail.com

Quick Response Code