

Excessive Myopia and Anisometropia Associated with Familial Exudative Vitreoretinopathy

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Background: To describe associations between familial exudative vitreoretinopathy (FEVR) and refractive status.

Methods: We conducted retrospective studies of patients with clinical diagnoses of familial exudative vitreoretinopathy from June 1986 through September 2000. All patients had cycloplegic refraction, visual acuity with correction, and underwent fundus examination. Ocular history since early childhood was described.

Results: Nine patients were recruited. All had excessive myopia, which was noted since early childhood. Amblyopia was noted in seven of the nine patients. Asymmetric FEVR was found in four of the nine patients. In all four patients with asymmetric FEVR, anisometropia equal to or greater than 2.5D was noted. Myopia was higher and amblyopia was more severe in the eye more severely affected with FEVR.

Conclusion: Excessive myopia, anisometropia and amblyopia were more likely to occur in eyes with familial exudative vitreoretinopathy. Patients with asymmetric disease tended to have higher myopia in the eye with more severe degree of FEVR as compared with the paired eye and thus correlated with anisometropia.

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Key words: familial exudative vitreoretinopathy (FEVR), peripheral retinal avascularization, excessive myopia, anisometropia.

Familial exudative vitreoretinopathy (FEVR) is a disease of peripheral retinal avascularization present in full-term infants. Fundus abnormalities of FEVR are similar to those of retinopathy of prematurity (ROP). It has been well recognized that myopia and anisometropia are more likely to occur in eyes after regression of ROP. In 1999, Cheeseman et al. reported the first patient with asymmetric FEVR who had pronounced axial myopia in the more severely affected eye.⁽¹⁾

Because of the frequently asymptomatic course

and diverse signs, the disease is often unrecognized or incorrectly diagnosed. The purpose of this study was to examine the refractive status of patients with FEVR and to check whether FEVR was associated with myopia and anisometropia as observed in patients with ROP.

METHODS

Nine patients with clinical diagnoses of FEVR were evaluated from June 1986 through September

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2000. A diagnosis of FEVR was established based on characteristic findings for the fundus. None of the subjects had history of premature birth.

Findings for the fundus of patients with FEVR were classified into one of the following three stages according to the classification by Pendergast.⁽²⁾ Stage 1 denoted the presence of a retinal avascular zone particularly involving the temporal periphery. Stage 2 denoted a peripheral avascular zone, vitreous bands, and dragging of the vessel or neovascularization. Stage 3 indicated phenomena described in stage 2 along with tractional and/or rhegmatogenous retinal detachment.

The best corrected visual acuity testing was performed according to the Snellen chart. Excessive myopia was defined as myopia greater than -6.0D. Anisometropia was defined as a difference in refractive status of equal or greater than 2.5D between the two eyes. Treatment strategies were determined based on disease severity. Eyes showing only peripheral avascularity were not treated. Patients with moderate disease were treated with peripheral laser ablation. Patients with retinal detachment were treated with vitrectomy and scleral buckling.

RESULTS

Eighteen eyes of nine patients (7 male and 2 female patients) were noted to have FEVR in our clinic from June 1986 to September 2000. Five of them had the disease noted during routine fundus checks. Three patients were referred to our clinic

with the diagnosis of retinal detachment. Tracing back their history, all patients had developed myopia in early childhood (myopia greater than -5.0D in at least one eye before the age of 8 years). None had previous diagnosis of FEVR. Eight eyes were in stage 1 of the disease, seven eyes were in stage 2, and three eyes were in stage 3. Amblyopia was noted in seven of the nine patients. Anisometropia was noted in six of the nine patients. Four of the nine patients had asymmetric FEVR, while all of them had anisometropia greater than 2.5D and higher degree of myopia and amblyopia in the eye with more severe degree of FEVR (cases 1-4). Two (cases 6 and 9) of the five patients with symmetric FEVR had anisometropia.

In cases 1, 2 and 3, excessive myopia and amblyopia had been noted since early childhood. Eyes with the more severe form of FEVR demonstrated amblyopia and higher degree of myopia than the paired eyes. Fundoscopic examination showed peripheral avascular retina with cicatricial changes. Combined rhegmatogenous and tractional retinal detachment developed in cases 1 and 2, both in the more severely affected eye. In case 3, a localized retinal detachment developed later in the more severely affected eye which was treated with laser photocoagulation.

Case 4 was a 4-year-old boy. Poor visual attention was noted during infancy. Fundus examination showed dragging of vessel and macula in the left eye and peripheral whitened areas in both eyes. Cycloplegic refraction, as shown in Table 1, also

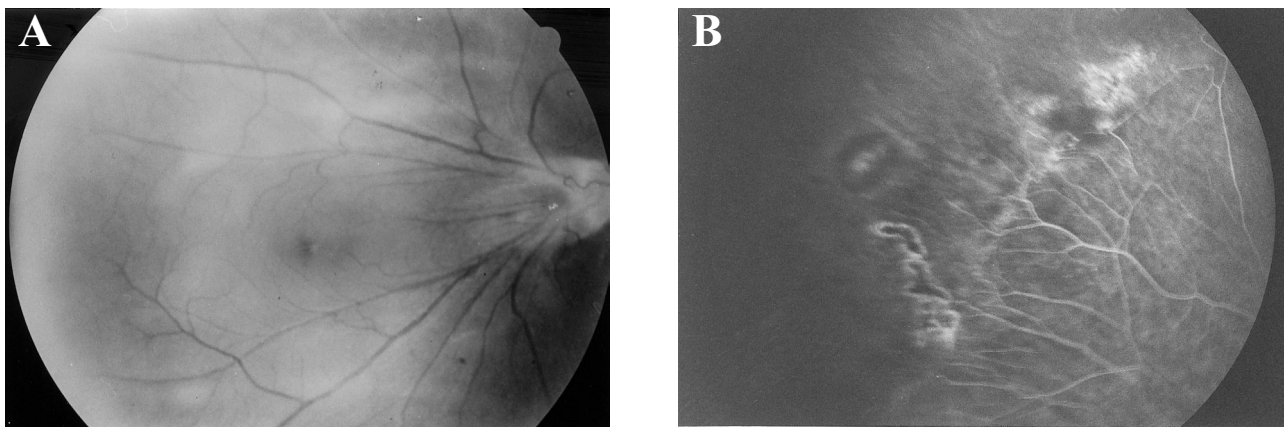


Fig. 1 (A) Fundus photograph of case 6. Right eye. Temporal dragging of optic disc and straightening of retinal vessels; (B) Fluorescein angiography showed peripheral vascular nonperfusion of both eyes compatible with FEVR.

Table 1. Clinical Data for the 9 Patients with Familial Exudative Vitreoretinopathy

No.		Age (years)	Gender	FEVR stage	BCVA	Spherical equivalent	RD
1	OD	15	M	3	20/200	-15.50	+
	OS			2	20/100	-10.50	-
2	OD	12	M	1	20/ 60	-11.50	-
	OS			3	20/200	-14.0	+
3	OD	6	M	*1 → 2	20/ 40	- 8.0	-
	OS			2 → 3	20/200	-13.0	+
4	OD	4	M	1	20/100	- 3.50	-
	OS			2	CF/50cm	-10.0	-
5	OD	15	F	1	20/ 20	- 8.75	-
	OS			1	20/ 20	- 8.75	-
6	OD	26	F	2	20/400	-16.75	-
	OS			2	20/ 40	-11.0	-
7	OD	14	M	2	20/ 20	- 7.0	-
	OS			2	20/ 20	- 8.0	-
8	OD	3	M	1	20/ 25	-11.0	-
	OS			1	20/ 40	-11.0	-
9	OD	6	M	1	20/100	-13.0	-
	OS			1	20/100	-10.0	-

Abbreviations: OD: right eye; OS: left eye; FVER: familial exudative vitreoretinopathy; BCVA: best-corrected visual acuity; RD: retinal detachment.

Stage 1: mild form, 2: moderate form, 3: severe form.

*progression in the follow-up period

demonstrated that myopia was higher in the more severely affected eye.

In cases 5 to 9, high myopia occurred during early childhood. These patients came to our clinic for routine fundus check. Fundus examination showed peripheral avascular retinas. Disc and retinal dragging were noted in both eyes in cases 6 and 7 (Fig. 1A). Fluorescein angiography showed peripheral vascular nonperfusion of both eyes which was compatible with FEVR (Fig. 1B).

Clinical data of the patients are summarized in Table 1.

DISCUSSION

Familial exudative vitreoretinopathy is usually bilateral and sometimes asymmetric. Van Nouhuys reported that 32% of FEVR patients have more than one diopter of anisometropia and 13% have anisometropic amblyopia.⁽³⁾

In our study, all patients had excessive myopia at the time of examination and had relatively high myopia since early childhood. We concluded that

FEVR was associated with the development of myopia. However, there might be selection bias in our study since patients with excessive myopia are prone to ask for fundus examination and thus have higher chance to be found to have FEVR. However, we have never found emmetropic patients with vitreous floater or any other retinal anomalies have peripheral vascular anomalies simulating FEVR.

In our study, six of the nine patients (case 1-4,6,9) had anisometropia equal or greater than 2.5 diopter (from 2.5 D to 6.5D). The incidence was much higher than that of the general population. Among the four patients with asymmetric FEVR, all had anisometropia. The eyes more severely affected with FEVR were always the ones with the higher degree of myopia, thus implying that the severity of FEVR regulated the growth of the eye ball and therefore influenced the degree of myopia. Amblyopia was noted in seven of the nine patients. Amblyopia might have been due to the disease of FEVR itself or excessive myopia induced by FEVR which was not diagnosed correctly during early childhood.

The basic pathology of FEVR is incomplete

vascularization of peripheral retina, similar to that of ROP. Since myopia and anisometropia are also more prevalent in patients with ROP, it is not surprising that patients with FEVR would have the same condition. Quinn et al.⁽⁴⁾ reported that among patients with ROP, the incidence of myopia increased in direct relation to the severity of ROP. According to the report of Krushner et al.⁽⁵⁾ asymmetric ROP may well contribute to anisometropia. In our study, similar conditions were noted in that patients with asymmetric FEVR always had higher myopia in eyes that were more severely affected.

The exact mechanism of myopia in patients with FEVR remains obscure. Feltcher and Brandon⁽⁶⁾ suggested that myopia in ROP might be due to an elongation of the globe, alteration of the lens, changes of the corneal curvature, or a combination of these factors. Cheeseman et al. reported that it was asymmetric elongation of the eyeball in children with asymmetric FEVR.⁽¹⁾ In our study, we did not perform studies on axial length, lens thickness, or corneal curvature. Since we observed tessellated fundus in all our patients, we believe that axial length elongation plays an important role in the pathogenesis of myopia in patients with FEVR.

Unlike what is usually seen in myopia patients without FEVR or ROP, all of our patients developed high myopia and amblyopia during early childhood; however, none of them had been informed of this diagnosis before they visit our clinic. This is probably because deliberate examination of fundus peripheral is more difficult in children and mild degrees of FEVR are frequently missed without the help of fluorescein angiography. Since patients with FEVR have much higher likelihood of developing retinal

detachment, routine fundus checks are necessary. We recommend careful fundus examination for children with high degrees of myopia. Such exercise would allow for early detection of the disease and regular follow up.

To the best of our knowledge, this study is the first case series discussing the association of FEVR with excessive myopia, and anisometropia with asymmetric disease severity. The association of myopia with retinopathy of prematurity is well known and the fundus abnormalities of FEVR are similar to those of ROP. These two disease entities may have the same pathophysiology in inducing myopia. Further investigations about axial length, lens thickness and anterior chamber depth associated with myopia are needed.

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