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SHORT COMMUNICATION

A case of retinal detachment in retinitis pigmentosa

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PURPOSE. *To report case of retinitis pigmentosa in association with rhegmatogenous retinal detachment.*

METHODS. *An eight year old boy complained of a sudden visual loss. The patient had night blindness, bone spicule-like hyperpigmentation, pale optic disc in both eyes, and the retina was totally detached in the right eye.*

RESULTS. *He was initially treated with conventional scleral buckling surgery, then pars plana vitrectomy with silicone tamponade was performed and retinal reattachment was established. After the phacoemulsification combined with silicone oil removal the final visual acuity of counting fingers was obtained.*

CONCLUSIONS. *The association of retinitis pigmentosa and rhegmatogenous retinal detachment is uncommon in young patients. (Eur J Ophthalmol 2007; 17: 677-9)*

KEY WORDS. *Retinitis pigmentosa, Rhegmatogenous retinal detachment*

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INTRODUCTION

Retinitis pigmentosa (RP) is a dystrophy of photoreceptors caused by mutations of different genes. Association of RP with rhegmatogenous retinal detachment (RD) is rare, but a few cases have been reported (1-6). The infrequency of rhegmatogenous RD in cases with RP is accounted for by the histologic changes of this disease (7-9). Due to the migration of the retinal pigment epithelium to the neurosensorial retina; the potential subretinal space is obliterated, causing firm adhesions between the neurosensory retina and retinal pigment epithelium (2). The vitreous changes in RP have been described previously (10).

In this case report, an 8-year-old boy with RP associated with rhegmatogenous RD is presented. The characteristics of RD and the course of surgical treatment are reported and compared with previous reports. In the current English literature, this case has the youngest age at occurrence.

Case report

An 8-year-old boy with a complaint of acute vision loss for 10 days in his right eye presented to the Department of Ophthalmology of Ankara Training and Research Hospital. The parents were first-degree relatives and they had been aware of the worsening visual acuity of their child for several years. There was neither history of trauma nor vision loss in the other family members. There was no evidence of systemic disease.

On examination visual acuity in the right eye was hand motions and best-corrected visual acuity in the left eye was 20/200. Biomicroscopic examination of the right eye revealed mild posterior subcapsular cataract, while the anterior segment of the left eye was normal, and bilateral vitreous degeneration was present. Intraocular pressures of both eyes were normal.

Funduscopic examination of the right eye revealed bone spicule-like hyperpigmentation, pale optic disc, and the retina was totally detached, with two holes located in up-

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per and inferotemporal peripheral retina. In the left eye pale optic disc, macular atrophy, vessel attenuation, peripheral atrophic regions, and hyperpigmentation were detected. Electroretinography amplitudes were severely reduced.

The eye was treated by cryotherapy and an encircling band, and drainage with intravitreal air injection. The retina was attached and visual acuity was counting fingers from 1 meter. One week after surgery visual acuity was hand motions and retina was redetached and pars plana vitrectomy, membrane pealing, posterior retinotomy, fluid-air exchange, endolaser, and air-silicone oil exchange was performed. Five months after vitrectomy, visual acuity was counting fingers from 1 meter, intraocular pressure was 18 mmHg, and significant posterior subcapsular cataract was present.

Six months after the vitrectomy phacoemulsification and silicone oil removal was performed. At the last examination after 12 months, the visual acuity was counting fingers and retina remained attached, with optic atrophy.

DISCUSSION

RP is the common name of a group of hereditary transmitted diseases characterized by progressive loss of photoreceptor and pigment epithelial function. Today it is known that the basic pathology is genetic and whatever the genetic defect is, the loss of photoreceptors is due to apoptosis (11).

The association of RP and RD is not common. Previous reports suggested that adhesions caused by the migration of pigment epithelial cells protect retina from detachment (8, 9). Most of the previously reported patients have been young, leading to the conclusion that in young patients with RP the adhesions are not yet well developed (1). Arakawa et al and Pruett reported 0.7% detachments among 572 RP cases, and 1.2% among 192 RP cases, respectively (4, 6). Comparing these detachment rates in RP cases with normal subjects causes contradiction. Are the histologic changes in RP protective against detachment? In addition, the high association of myopia with RP brings more difficulty.

The case presented here has the youngest age among the previously reported cases. The fact that most reported cases are under 20 years of age supports the idea that RP becomes protective against detachment in older patients.

Genetic penetration in RP is common. In addition to autosomal dominant, autosomal recessive, and X-linked transition, isolated cases with no family history are present. Systemic associations with RP are common. In this case, with related parents, no family history or systemic association was detected. Glaucoma, keratoconus, and refractive errors were absent. On the first examination in the right eye mild posterior subcapsular cataract was present. Electroretinography was typical.

In previous reported cases, RP-associated RDs have been noted along with hole, dialyses, horseshoe tears, giant tears, and no tears. In our case two holes was the cause of the RD. After unsuccessful conventional RD surgery, pars plana vitrectomy with silicone tamponade was performed and retinal reattachment was established. After the phacoemulsification combined with silicone oil removal the final visual acuity was obtained.

Even if low visual acuity is common in RP cases, sudden decrease in visual acuity or visual field loss from RD must be considered. Because the adhesions between pigment epithelium and neurosensorial retina are not strong enough, the protective effect is not established. The success rates of detachment surgery are not different from those in typical cases.

Proprietary interest: None.

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