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Retinal Detachment in 31 Eyes with Retinitis Pigmentosa

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Objective: To describe clinical features and treatment outcomes of retinal detachment (RD) in eyes with retinitis pigmentosa (RP).

Design: Single-center, retrospective, interventional case series.

Subjects: All RP patients with RD examined between April 2003 and December 2013 and minimum 2 months of follow-up.

Methods: Medical records of RP patients were screened and 31 eyes with RD were included. Family history of RP, duration of symptoms, age at presentation, associated ocular and systemic findings, and detailed ophthalmic evaluation including presenting visual acuity, type and amount of refractive error, fundus findings, electroretinogram details, surgical details, and postoperative complications and outcomes were evaluated. Univariate analysis was done to determine risk factors associated with RD in eyes with RP and risk factors associated with poor visual outcomes after treatment. Subset analysis was also done for comparing the functional and anatomical outcomes between patients undergoing scleral buckling or vitrectomy.

Outcome Measures: Final surgical reattachment rate, best-corrected visual acuity.

Results: Mean age at presentation was 22 years (median, 17; range, 4−63). Mean duration of symptoms was 12 months (median, 3 months: range, 3 days−60 months). Associated ocular findings included nyctalopia (n = 23), myopia (n = 21), and hyperopia (n = 10). Systemic associations included hearing loss (n = 5), deafmutism (n = 1), and Bardet-Biedel syndrome (n = 1). No association between degree of myopia and RD was noted ($P = 0.63$). Observed retinal breaks included horse-shoe−shaped tear (n = 15), lattice with hole (n = 7), atrophic retinal hole (n = 3), retinal dialysis (n = 3), and macular hole (n = 3). The most common location of breaks was superotemporal quadrant (n = 15). Younger age, male gender, and presence of posterior vitreous detachment were strongly associated with RD with odds ratio of 1.3 ($P = 0.001$), 8.3 ($P = 0.010$), and 6 ($P = 0.003$), respectively. Retinal reattachment was achieved in 13 of 13 eyes (100%) with scleral buckle and 9 of 10 eyes (90%) with vitrectomy. Vision improved from 1.63±0.52 to 0.87±0.25 logarithm of the minimum angle of resolution ($P < 0.001$) at a mean follow-up of 33 months (median, 24, range; 1−145).

Conclusion: Rhegmatogenous RD in eyes with RP is rare. Precocious vitreous degeneration and sparse pigmentation in younger male patients has a role in etiopathogenesis. Visual prognosis remains poor despite satisfactory surgical outcomes. *Ophthalmology Retina 2017;■:1−7 © 2017 by the American Academy of Ophthalmology*

Retinitis pigmentosa (RP) is a group of inherited retinal diseases characterized by early nyctalopia, progressive loss of photoreceptor function, and pigment deposition within the retina. The typical RP fundus reveals perivascular deposits of bone-spicule pigmentation which are most pronounced in the midperipheral region.[1] Pigment cells reach inner retina by migration from the retinal pigment epithelial (RPE).[2] This migration obliterates the potential space between the neurosensory retina and RPE, and seems to impart unusual adhesion between the 2 layers. Migration of the RPE is also seen after chorioretinopathy, and after trauma sufficient to cause a loss of photoreceptors.[3]

Retinal detachment (RD) is a rare event in eyes with RP, with prevalence ranging from 0.7% to 1.3%.[4,5] Rhegmatogenous, tractional, and exudative type of RD have been reported in eyes with RP.[6−8] Previous reports have suggested protective role of adhesions caused by migration of pigment epithelial cells and concluded that RD could only occur in a younger age group, given that the adhesions were not yet developed.[6−8] Patients with RP have a poor visual prognosis. However, because the disease is slowly progressive many patients maintain and live a functional life. In such eyes, RD results in sudden loss of vision and carries a grave prognosis, if left untreated. A PubMed search using the keywords <retinitis pigmentosa> and <retinal detachment> revealed only 9 relevant studies.[4−11] Herein, we report the clinical characteristics and surgical outcomes of RD in 31 eyes with RP.

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Methods

This single-center, retrospective, interventional, case series from a tertiary eye care institution in India reviewed the medical records of 31 patients diagnosed with RP and RD between April 2003 and December 2013. Of 5044 cases of RP, RD was seen in 66 eyes (1.3%). Of these 66 eyes, 31 eyes that had complete ophthalmic and investigational reports in support of RP and follow-up details available for ≥2 months were included in the study. Patients with bilateral pigmentary retinopathies due to inflammation, drug toxicity, spontaneous retinal reattachment, and with exudative Coat's–like features were excluded. Prior institution board review was obtained and all the tenets of the Declaration of Helsinki were followed.

Medical records of these 31 eyes were further analyzed for presenting history, family history of RP, presenting visual acuity, type and amount of refractive error, fundus findings like presence or absence of vitreous degeneration, presence or absence of posterior vitreous detachment (PVD), type of RP, disc and blood vessels details, amount of retinal pigment epithelium atrophy, amount of pigmentation seen, electroretinogram details, surgical details, post-operative complications, and treatment outcomes. Patients were divided in 2 groups on the basis of type of surgery performed, namely, scleral buckling or vitrectomy (Fig 1). Subset analysis was done for comparing the functional and anatomical outcomes between the 2 groups. Statistical analysis was done using paired *t*-test within the group and independent *t* test between the 2 groups. Univariate analysis was done to determine risk factors associated with RD in eyes with RP, after comparing them with 30 control eyes with RP but without RD. Pearson's correlation and chi-square tests were used to assess the strength of association between the variables with RD. Univariate and multivariate analysis using regression model was done to determine risk factors associated with poor visual outcomes after treatment using SPSS software version 20 (SPSS, Inc, Chicago, IL).

Results

Subjects included 29 males and 2 females. Mean age at presentation was 22 years (median, 17; range, 4–63); 27 patients (87%) were younger than 30 years. The mean duration of symptoms related to RD was 12 months (median, 3 months; range, 3 days–60 months). Preceding history of trauma was positive in 8 eyes; 2 had retinal dialysis. Mean follow-up was 33 months (median, 24; range, 1–145). Three patients had a history of consanguinity, one of which had bilateral RD. In the entire cohort, 2 cases had bilateral RD. The details about inheritance pattern, clinical classification, and ocular and systemic associations are listed in Table 1.

The mean logarithm of the minimum angle of resolution (logMAR) visual acuity at presentation was 1.63±0.89. Regarding refractive errors at presentation, although myopia (n = 21) was noticed more often than hyperopia (n = 10), the difference was not statistically significant ($P = 0.27$). Myopia was mild (0 to −3 diopters [D]) in 5 eyes, moderate (>−3 to −6 D) in 7 eyes and high (>−6 D) in 9 eyes. No association was seen between degree of myopia and RD ($P = 0.63$). A strong association of RD was seen with PVD with an odds ratio of 6 ($P = 0.003$). A statistically significant association was also seen with younger age and male gender (Table 2). In 13 eyes (42%) RD was total; it was partial in 18 eyes (58%). Macular involvement was noted in 30 eyes (96%) and only 1 eye presented before the macula was detached. In all, PVD was present in 17 eyes (55%). A horse shoe–shaped tear was the most common break, seen in 15 eyes (48%). Table 1 depicts the

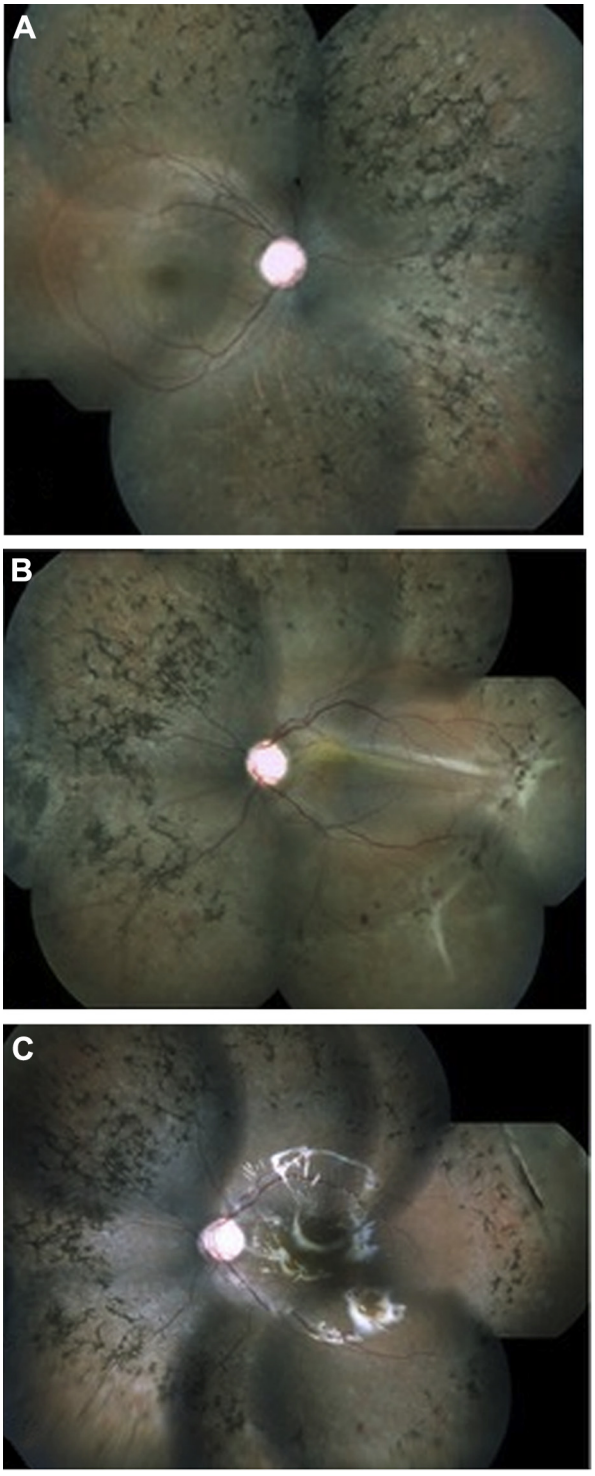

Figure 1. At presentation, color fundus photographs of the right (**A**) and left (**B**) eyes reveal waxy disc pallor, arteriolar attenuation, midperipheral retinal pigment epithelium atrophy, and bone-spicule pigmentation. The left eye also revealed partial rhegmatogenous retinal detachment with starfold at the 3-o'clock meridian (**middle**). Five months after vitrectomy, encirclage, endolaser photocoagulation, and silicone oil injection in left eye, the retina was completely attached (**C**).