INTRODUCTION

Several macular complications related to abnormalities of the vitreoretinal interface have been classically attributed to retinitis pigmentosa (RP) of which cystoid macular edema (CME) is the most common. Other less frequent complications are as follows: epiretinal membranes (ERM), vitreomacular traction (VMT) syndrome and macular holes (MHs). The detection of these abnormalities is crucial because they can significantly reduce the visual acuity in patients with RP whose central vision is usually well preserved until the late stages of the disease. The exact prevalence of these alterations is unknown, but we can say that the presence of a MH in patients affected by RP varies from 0.4% to 10%. The possibility of spontaneous resolution of a full-thickness MH in a patient with RP was once described in the literature but, generally, a therapeutic approach based on vitrectomy is necessary.

We report a case of MH closure after pars plana vitrectomy (PPV) and late reopening, in a patient with RP.

METHODS

A 64-year-old woman, with the previous diagnosis of RP, was referred to our department with a complaint of central visual loss in her left eye (LE) for 12 months. A complete ocular exploration, including determination of Best Corrected Visual Acuity (BCVA) (decimal notation), funduscopy and Optical Coherence Tomography (OCT) examination was performed.

RESULTS

AT FIRST EXAMINATION. her BCVA was 0.1 in her LE, and 0.8 in her right eye (RE).

An examination of the anterior segment was unremarkable. A funduscopy examination of her LE revealed the presence of a stage-4 macular hole (MH) with a cuff of subretinal fluid surrounding it. A posterior vitreous detachment was observed. Narrowing of the retinal arterioles, waxy yellow appearance of the disk and hyperpigmentation in a bone-spicule configuration in the midperipheral retina were also observed in both eyes. However, the macular appearance of her RE was normal.

An electroneudetogram showed reduced scotopic and photopic responses in both eyes. Perimetry revealed the presence of an annular scotoma in both eyes, associated to central scotoma in her LE. OCT (Stratus OCT™, Carl Zeiss) confirmed the presence of a full-thickness MH in her LE.

Therefore, we decided to perform 20-gauge pars plana vitrectomy (PPV). Peeling of the internal limiting membrane (assisted by the dye Brilliant Blue G, Brilliant Puff) in the area around the macular hole and 25% sulfur hexafluoride intravitreal gas fill were performed. Face down positioning for 5 days was also advised.

AT 2 MONTHS AFTER THE VITRECTOMY. her BCVA was 0.4 (decimal notation). Funduscopy and OCT confirmed the hole closure, but severe macular atrophy was observed. After 6 months, her BCVA was 0.2. Biomicroscopy showed the presence of a posterior subcapsular cataract.

Funduscopy and OCT examination, confirmed the closure of the MH. Therefore, crystalline extraction with phacoemulsification was performed, and no complications occurred. However, BCVA after cataract surgery did not improve. The patient came to our center 2 YEARS AFTER PPV complaining of mild episcopal visual acuity loss in her LE. Her BCVA at this time was 0.1 in the LE and 0.8 in the RE. A funduscopy and OCT examination demonstrated the reopening of the previously closed full-thickness MH and the presence of a ring of subretinal fluid and intense retinal atrophy surrounding the hole. Because of the guarded prognosis, the high likelihood that a reopening had occurred again due to the severe retinal atrophy around the hole, we decided not to perform a new surgery.

CONCLUSION

The pathogenesis of macular hole formation in patients with retinitis pigmentosa is unclear. Surgical outcomes may not always be favorable, and the possibility of reopening must be taken into account, even after a long time.

REFERENCES