

Surgical treatment of macular detachment secondary to congenital pit of the optic disc: case report

Tratamento cirúrgico do descolamento macular secundário à fosseta congênita do disco óptico: relato de caso

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ABSTRACT

The authors describe a patient with macular detachment in the right eye, secondary to congenital pit of the optic disc, submitted to vitrectomy, drainage of the subretinal fluid, perfluorocarbon, endolaser and perfluoropropane gas (C₃F₈). A sample of the vitreous humor was taken for comparative analysis with the subretinal fluid. Results of retinography, optical coherence tomography, and illustrations of the surgical procedure are presented. Optical coherence tomography revealed the detachment resolved 6 months after surgery. Visual acuity improved from 20/400 to 20/30. This surgical technique can lead to successful reattachment of the macula and improvement of central vision. Additionally, a qualitative and comparative analysis showed a similar biochemical composition of the subretinal fluid and the vitreous. Despite technical limitations, this analysis can confirm the pathophysiology of the disease, suggesting that the subretinal fluid may originate from the vitreous cavity in macular detachment secondary to congenital pit of the optic disc.

Keywords: Optic nerve/abnormalities; Vitreous body/surgery; Vitrectomy/methods; Retinal detachment/etiology; Retinal detachment/therapy; Tomography, optical coherence/methods; Case reports [Publication type]

INTRODUCTION

Congenital pit of the optic nerve head is a rare anomaly first described by Wiethe in 1882⁽¹⁾. Serous macular retinal detachment is associated with 25% to 75% of optic nerve head pits⁽²⁻⁴⁾. Using the cross-sectional retinal imaging provided by optical coherence tomography (OCT), several authors have confirmed the concept of a bilaminar structure with schisis-like retinal changes overlying a more central neurosensory retinal detachment⁽⁵⁻⁷⁾.

The pathogenesis of the optic disc pit maculopathy is unknown. It has been proposed that the vitreous could be the origin of the subretinal fluid, considering the higher incidence of posterior vitreous detachment in eyes with optic disc pits and retinal detachment⁽⁸⁾. The transfer of the dye from the vitreous cavity to the subretinal space has been demonstrated in animal models. However, these studies failed to identify, in the subretinal space, any glycosaminoglycan, an important component of the vitreous⁽⁹⁾. Others believe the cerebrospinal fluid should be considered as the source of subretinal fluid in eyes with pits⁽¹⁰⁻¹¹⁾, although no animal or human model has demonstrated a connection between the pit and the subarachnoid space. Other possibilities would be leakage from the choroid or from vessels within the pit⁽¹²⁾.

Study carried out at the Retina and Vitreous Service, Universidade de São Paulo - USP, São Paulo (SP) - Brazil, and São Rafael Hospital, Monte Tabor Foundation, Salvador (BA) - Brazil.

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On the other hand, the management of this entity has also been controversial, since no treatment has clearly proved more advantageous than the others. However, the low incidence of this entity makes it difficult to obtain series large enough to determine the efficacy of the vitrectomy-laser-gas procedure, and other treatment modalities and be able to suggest a procedure of choice.

The objective of this report is to describe an alternative surgical technique for the management of retinal detachment with active internal drainage of subretinal fluid, and to analyze the subretinal fluid in one case of macular detachment secondary to congenital pit of the optic disc, comparing it with the vitreous composition of the patient himself, in order to help understand the pathogenesis of retinal changes.

CASE REPORT

MSC, 23 years old, male, Caucasian, university student, born and raised in São Paulo, was referred to the Emergency Room of the Hospital das Clínicas, University of São Paulo Medical School, for evaluation, due to a decrease in visual acuity of the right eye first noticed 3 months ago. The patient denied other ocular changes. Medical and family history was not significant. Best corrected visual acuity was 20/400 in the right eye and 20/20 in the left eye. Biomicroscopy and intraocular pressure were normal in both eyes. Fundoscopy showed neurosensory detachment in the macular area and inner retinal schisis associated with optic disc pit was found on OCT, showing that the retinal elevation consisted of separation of both the inner and outer layers in the right eye (Figure 1). No changes were observed in the left eye.

The patient underwent a standard vitrectomy, removal of the posterior hyaloid, with perfluorocarbon, laser photocoagulation, fluid-gas exchange and 12% perfluoropropane (C_3F_8) injection. The whole procedure was explained to the patient beforehand. The drainage of subretinal fluid was performed under direct visualization (Landers flat lens) through a 39-gauge cannula. In addition, a sample of the vitreous humor was taken for comparative analysis. There were no intercurrent events during the surgical procedure (Figure 2).

The qualitative and comparative analysis of subretinal fluid and vitreous was carried out by the Biochemical and Cerebrospinal Fluid Laboratory of the Hospital das Clínicas. Similar results (pH, hyaluronic acid, glycoproteins, sodium, potassium, bicarbonate, glucose and ascorbic acid) were obtained from both samples.

Postoperatively, the patient was asked to remain in a face-down position for three weeks. After 2 months, the macular elevation gradually resolved, and the OCT showed the flattening of the outer layer detachment in the fovea. At the last ophthalmologic evaluation (6 months), an OCT revealed complete resolution of the detachment (Figure 3), and visual acuity was 20/30.

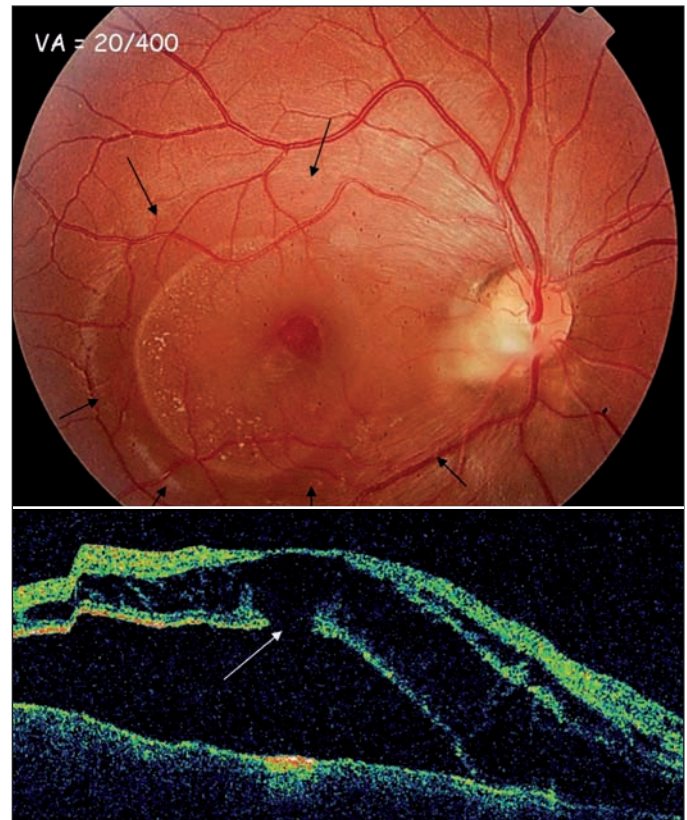


Figure 1 - Preoperative: 23-year-old male patient with macular detachment secondary to congenital pit of the optic disc. A fundus photograph of the right eye shows infratemporal optic disc pit associated with a large oval-shaped area of macular detachment (black arrows). Stratus OCT shows retinal schisis and rupture of the external leaflet in the macular area (white arrow).

DISCUSSION

There is controversy about the treatment of retinal detachment associated with congenital pit of the optic disc. Treatment modalities such as external laser therapy alone⁽¹³⁾, intravitreal gas injection alone⁽¹⁴⁾, laser therapy combined with gas⁽¹⁵⁾, vitrectomy associated with gas tamponade, with or without additional laser treatment⁽¹⁶⁻¹⁷⁾ and scleral buckling⁽¹⁸⁾ have been described in the literature. In this particular case, the best choice was posterior vitrectomy via *pars plana*, with perfluorocarbon, endophotocoagulation and gas tamponade (C_3F_8); however, active internal drainage of the subretinal fluid can lead to successful reattachment of the macula and improvement of central vision. Subretinal fluid is eventually absorbed by the retinal pigment epithelium (RPE) and eventually complete apposition of the neurosensory retina with the RPE is achieved. Some retinal detachments are associated with a high volume of subretinal fluid, and its removal can be an important step for the successful surgical procedure. Several factors related to retinal detachment associated with congenital pit of the optic disc are noteworthy. First, good visual recovery is possible even months after the onset of maculopathy⁽¹⁹⁾, and serous macular retinal detachment responds well to early vitrectomy⁽²⁰⁾.

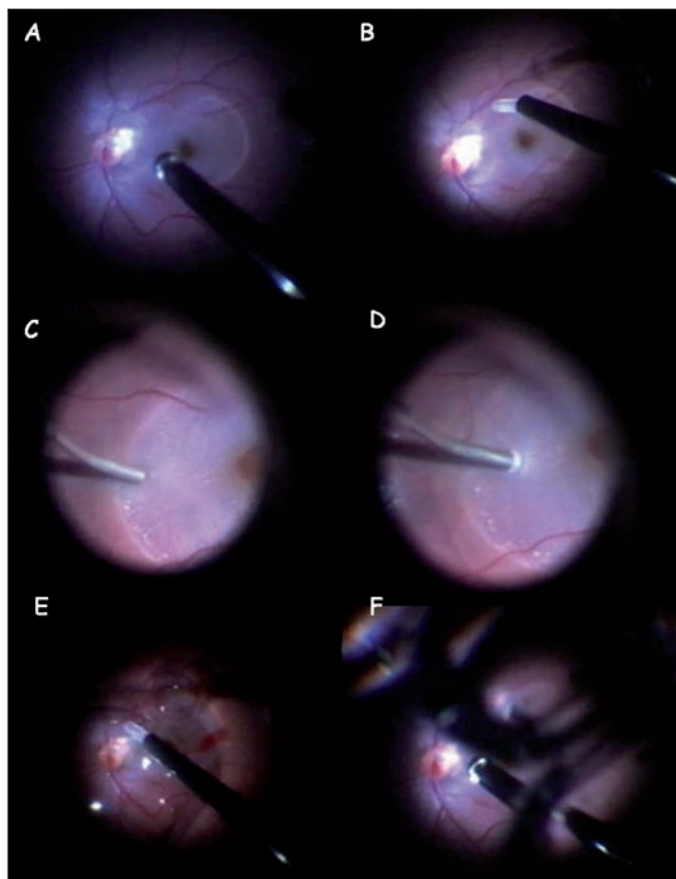


Figure 2 - Intraoperative: (A) Conventional vitrectomy of the macular detachment secondary to the congenital pit of the optic disc; (B) Cautious removal of the posterior hyaloid; (C, D) Drainage of the subretinal fluid conducted under direct visualization (Landers flat lens) through a 39-gauge cannula; (E) Perfluorocarbon injection and application of endolaser; (F) Fluid-air exchange and subsequent perfluoropropane injection (C_3F_8)

Macular detachments associated with pits of optic disc occur in young patients, who usually do not have significant vitreous liquefaction, which, in turn, makes less likely the hypothesis that the liquid comes from the vitreous⁽²¹⁾. The absence of separation of the posterior vitreous in patients with macular detachment suggests that vitreous traction in this area could play a part in the genesis of the passive movement of fluid to the subretinal space⁽²²⁻²³⁾. The fluid from the pit elevates the layer of nerve fibers, causing internal retinal layers to separate and evolving into serous retinal detachment⁽²⁴⁾. Spaide et al. published a case that had a rapid improvement in acuity and fundus appearance with the simple introduction of a partial thickness fenestration near the optic disc pit, to redirect the flow of fluid from the retina into the vitreous cavity⁽²⁵⁾. Yet, there is still no consensus on the pathogenesis of the retinal alterations.

During retinal detachment, subretinal fluid is present, whose composition and pathophysiology are still little known. Subretinal fluid composition varies according to detachment duration; total protein concentration in subretinal fluid increases with time. In addition, all proteins are qualitatively modified. The detached retina loses its oxygen supply, and it then uses the anaerobic pathway to degrade glucose. Thus, long-duration retinal detachments feature increased lactic acid and dextrose concentrations. Phospholipids are also increased in subretinal fluid, reflecting retinal degradation⁽²⁶⁾. On the other hand, the presence of components derived from hyaluronic acid depolarization is a valid argument in favor of the predominantly vitreous origin of subretinal fluid⁽²⁶⁾. So, the composition of the subretinal fluid was similar to that of the vitreous humor, suggesting that the origin of the fluid may be the vitreous cavity. The limitation of this result was that, during surgery, the drainage of the subretinal fluid was carried out

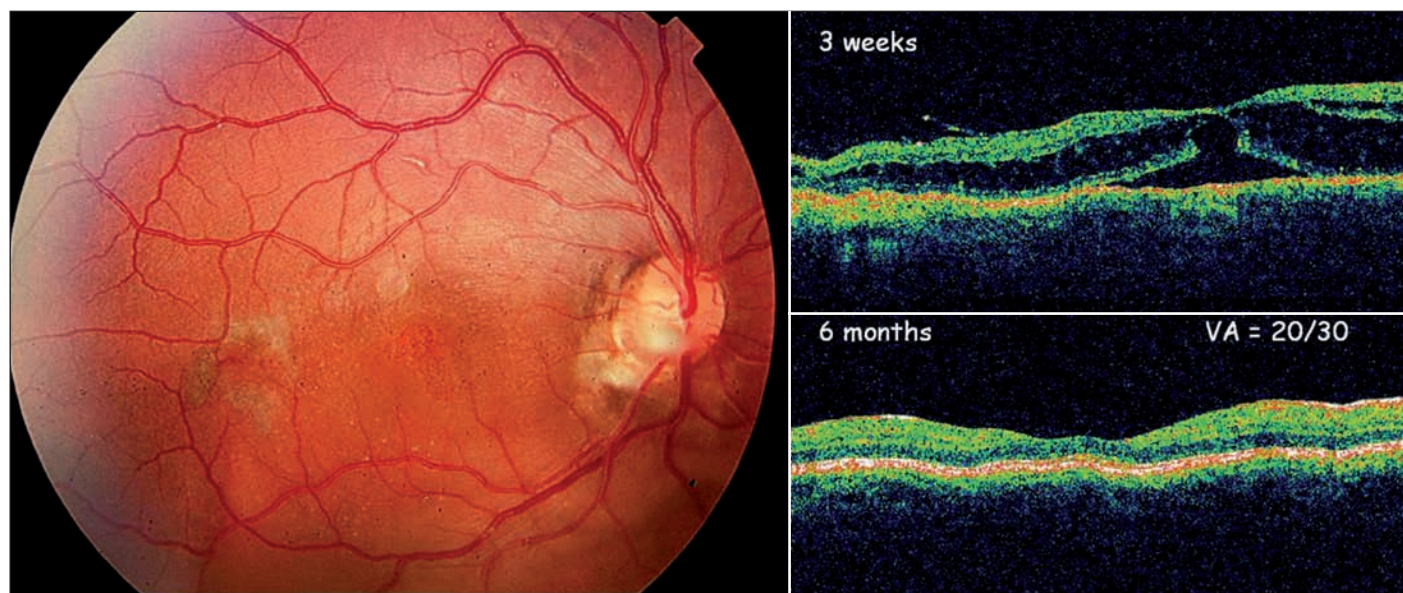


Figure 3 - Postoperative: Three weeks after surgery, the OCT showed a decrease in the inner layer separation and outer layer detachment. At 6 months, the OCT showed complete resolution of the inner layer separation and reestablishment of the fovea structure. Fundus photograph revealed attached macula with changes in pigmentation due to the endophotocoagulation. Best corrected visual acuity was 20/30.

under direct visualization, drainage being technically impossible after fluid-air exchange that would isolate the subretinal fluid from potential contamination by the vitreous cavity liquid. The amount of subretinal fluid and vitreous was enough to conduct the analysis, and results were similar. However, in our hospital, the test for β -trace protein is not available. This protein is produced by the epithelial cells of the choroid plexus and the leptomeninges, and a highly accurate cerebrospinal fluid marker⁽²⁷⁾. Despite technical limitations, this analysis can corroborate the pathophysiological mechanism of this disease, suggesting that the subretinal fluid may originate from the vitreous cavity.

The present case showed reestablishment of the anatomic macular structure and significant functional improvement after 6 months of the surgery. OCT helped in monitoring the macular elevation that resolved gradually, with a progressive increase in visual acuity coinciding with the reabsorption of subretinal fluid. The limited recovery of visual function is probably related to its longstanding nature, and to the deposition of exudate on the posterior retinal surface and marked alteration in macular function characteristic of these detachments.

In conclusion, this case report documents the successful reattachment of a serous macular detachment associated with optic disc pit by vitrectomy, active internal drainage of the subretinal fluid, perfluorocarbon, endolaser, and fluid-gas exchange. OCT permitted anatomic assessment of the retina before and after treatment and was useful in the diagnosis and follow-up of this case. Further reports of the management of this uncommon condition are required so that firmer guidelines on treatment can be established.

RESUMO

Os autores descrevem um paciente portador de descolamento macular secundário à fosseta congênita do nervo óptico no olho direito, submetido à vitrectomia, drenagem do fluido sub-retiniano, perfluorocarbono, endolaser e gás perfluoropropano (C_3F_8). Foi retirada amostra do humor vítreo para análise comparativa com o fluido sub-retiniano. São apresentadas retinografia, tomografia de coerência óptica e ilustrações do procedimento cirúrgico. Após 6 meses da cirurgia, houve resolução do descolamento evidenciada por meio da tomografia de coerência óptica. A acuidade visual melhorou de 20/400 para 20/30. A técnica cirúrgica utilizada pode ter melhorado os resultados obtidos. Adicionalmente, a análise bioquímica qualitativa e comparativa do fluido sub-retiniano e do vítreo mostrou composição semelhante. Apesar das limitações técnicas, esta análise pode corroborar na fisiopatogênese da doença, sugerindo que o fluido sub-retiniano pode ser originado da cavidade vítrea na fosseta congênita de papila.

Descritores: Nervo óptico/anormalidades; Corpo vítreo/cirurgia; Vitrectomia/métodos; Descolamento retiniano/etiologia; Descolamento retiniano/ terapia; Tomografia de coerência óptica/métodos; Relatos de casos [Tipo de publicação]

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