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Case Report

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Vitrectomy in a Patient with Ehlers-Danlos Syndrome: Surgical Challenges and Visual Results

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ABSTRACT

Ehlers-Danlos Syndrome (EDS) comprises a heterogeneous group of heritable connective-tissue disorders characterized by tissue fragility, skin hyperextensibility, and joint hypermobility. Ophthalmic manifestations including high myopia, corneal fragility, and a predisposition to retinal detachment have been reported across different EDS subtypes. We describe a 31-year-old male with EDS who presented with bilateral rhegmatogenous retinal detachment associated with multiple peripheral tears and early cortical opacities in both lenses. Posterior vitrectomy with scleral indentation, perfluorocarbon liquid, and silicone-oil tamponade was performed using tailored surgical adaptations to minimize trauma to fragile ocular tissues. Post-operatively, visual acuity improved from counting fingers to 20/80 in the initially treated eye, with no significant intra- or post-operative complications. This report highlights the technical challenges and need for meticulous planning in retinal surgery for EDS patients, and underscores the importance of continuous ophthalmic follow-up for early detection of complications and optimization of visual outcomes.

Keywords:

Ehlers-Danlos Syndrome (EDS), Retinal detachment, Vitrectomy, Connective tissue, Internal tamponade.

Objectives

To describe the surgical management and visual outcomes of vitrectomy in a patient with Ehlers-Danlos syndrome and bilateral retinal detachment.

Introduction

Ehlers-Danlos syndrome (EDS) comprises a heterogeneous group of inherited connective tissue disorders characterized by mutations affecting collagen processing, structure, or regulation [1]. Classical, hypermobile, and vascular subtypes are among the most prevalent, each presenting distinct systemic manifestations [2]. Collagen abnormalities not only compromise skin and joint integrity but also ocular structures, given collagen's fundamental role in maintaining corneal transparency, scleral integrity, and vitreous architecture [3]. Cohort studies by Gonçalves et al. [4] and Pereira et al. [5] indicate that up to 30% of EDS patients experience ophthalmic manifestations including keratoconus, scleral thinning, and spontaneous ocular perforations during their lifetime. A predisposition to rhegmatogenous retinal detachment, often bilateral and associated with multiple peripheral tears, stems from alterations at the vitreoretinal interface and intrinsic retinal fragility exacerbated by high axial myopia common in these patients [6].

Surgical management of retinal detachment in EDS presents unique challenges. Fragility of conjunctival and scleral tissues necessitates minimally traumatic incision and suturing techniques, the use of delicate instrumentation, and careful globe manipulation [7]. Internal tamponades such as perfluorocarbon liquids facilitate intraoperative retinal reattachment but require vigilant intraocular pressure control and precise application to avoid additional tissue injury [8]. Silicone oil tamponade, although providing prolonged support in complex cases, carries risks of hemorrhage, inflammatory debris accumulation, and excessive fibrotic scarring if insertion and removal are not meticulously timed [9]. Despite favorable outcomes reported by Brown et al. [10], standardized surgical protocols and medium- to long-term outcome data are scarce. Detailed case reports and series are therefore crucial for disseminating surgical adaptations and promoting multidisciplinary planning including genetic counseling, anesthetic management tailored to vascular fragility, and specialized perioperative nursing care—to optimize safety and visual outcomes [11].

Case Report

A 31-year-old male reported reduced visual acuity in the right eye for three months and in the left eye for one month. Ophthalmic

examination revealed light-perception vision in the right eye and 20/400 in the left. Biomicroscopy showed early cortical lens opacities bilaterally. Fundoscopy demonstrated high myopic changes (axial length >26 mm) and bilateral retinal detachment with multiple temporal- and inferior-peripheral tears. Posterior vitrectomy with scleral indentation, perfluorocarbon liquid, and silicone-oil tamponade was performed, incorporating adaptations for fragile ocular tissue. The left eye was addressed first owing to the shorter duration of symptoms; surgery on the right eye is scheduled. Immediate post-operative visual acuity in the left eye improved to 20/80 with correction. No intra- or post-operative complications were observed, apart from the planned retention of silicone oil.

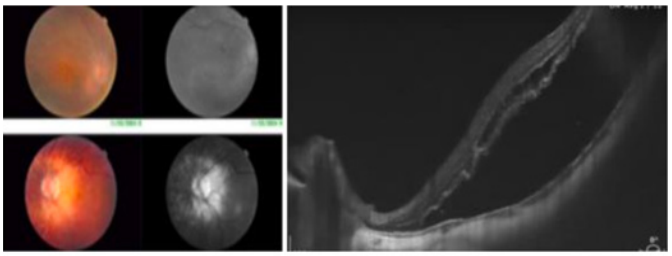

Figure 1: Fundus photograph and OE Oct.

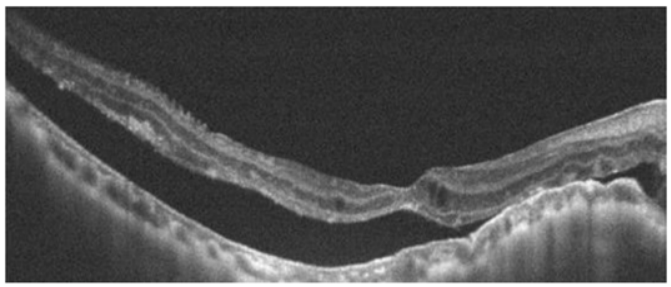

Figure 2: OD OCT.

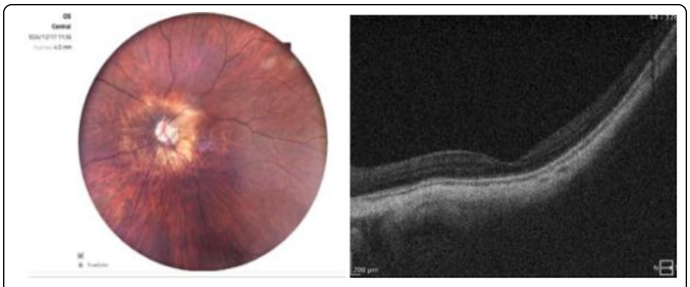

Figure 3: Retinography of the LE and Oct after the surgical procedure.

Discussion

Vitrectomy in EDS patients poses significant technical challenges due to connective tissue fragility and abnormal wound healing [6]. Instrumentation with smaller-gauge cannulas and low-tension suturing techniques can reduce scleral trauma and postoperative complications [7]. The use of perfluorocarbon liquids allows for controlled reattachment of the retina, though inadvertent subretinal migration must be avoided by careful fluid–air exchange and fundus visualization [8]. Silicone oil tamponade offers sustained support for complex detachments but necessitates monitoring for silicone-induced glaucoma and emulsification [3,12]. Our case demonstrates that adapting standard vitreoretinal techniques can yield satisfactory anatomical and functional results in EDS patients.

A tailored approach mapping tear locations, assessing scleral thickness preoperatively, and coordinating with anesthesiology for stable hemodynamics facilitates safe surgical execution [11]. Postoperative visual acuity improved from counting fingers to 20/80 in the treated eye, comparable to outcomes reported by Williams et al. [13], where success rates range from 60% to 85%. Long-term monitoring is essential to detect residual or recurrent detachments and manage complications such as silicone oil–induced ocular hypertension and fibrous proliferation [3]. Future research should focus on prospective series to establish evidence-based guidelines for tamponade selection, timing of oil removal, and rehabilitation protocols to preserve visual function and quality of life in this high-risk population [10].

Conclusion

In patients with Ehlers-Danlos syndrome, vitrectomy combined with scleral indentation, perfluorocarbon liquids, and silicone oil tamponade can achieve anatomical reattachment and functional improvement when conducted with meticulous surgical planning and tissue-sparing techniques [6,7]. Multidisciplinary collaboration involving geneticists, anesthesiologists, and specialized nursing staff is paramount for optimizing perioperative safety [11]. Regular long-term follow-up enables early detection of complications, such as silicone oil–related glaucoma, and timely management to sustain visual gains [3]. While individual case reports provide valuable insights, systematic data collection and comparative studies are needed to develop standardized surgical algorithms and improve outcomes for EDS patients undergoing retinal surgery [10].

Conflict of Interest

None.

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