• Letter to the Editor •

Recurrent suprachoroidal hemorrhages in a patient with classical Ehlers-Danlos syndrome: a case report

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Received: 2026-01-21 Accepted: 2026-04-17

DOI:10.18240/ijo.2026.12.24

Citation: Li ZY, Deng WL, Zhang YL. Recurrent suprachoroidal hemorrhages in a patient with classical Ehlers-Danlos syndrome: a case report. *Int J Ophthalmol* 2026;19(12):2514-2516

Dear Editor,

Classical Ehlers-Danlos syndrome (cEDS), one of the subtypes of Ehlers-Danlos syndrome (EDS), is a rare genetic disorder of collagen and extracellular matrix proteins[1], characterized by common manifestations of collagenopathies, including joint hypermobility, skin hyperelasticity and hyperextensibility, abnormal wound healing, and tissue fragility[2]. Little is known regarding the ocular manifestations of cEDS, reporting ophthalmic complications include blue sclera, dry eye, keratoconus, myopia, and retinal detachment (RD)[3].

Currently, there are no documented reports establishing a link between cEDS and suprachoroidal hemorrhage (SCH). Herein, we present a unique case of recurrent SCH during vitrectomy in a patient with cEDS.

CASE REPORT

Ethical Approval All procedures adhered to the tenets of Declaration of Helsinki. Written informed consent was obtained from the patient.

A 45-year-old woman underwent standard cataract surgery under local anesthesia at another hospital 20d prior. Preoperatively, an ocular examination revealed a keratoleukoma with no light perception in the right eye and a brunescent cataract with a visual acuity of counting fingers at 10 cm in the left eye. Additionally, a B-ultrasound of the left eye showed posterior staphyloma (Figure 1). She denied having any other significant past medical history, including atopic, allergic, skin, and cardiovascular diseases.

During the initial cataract surgery, a posterior capsule rupture happened, causing the nucleus to drop into the vitreous cavity and leaving the patient aphakic. A vitrectomy was promptly initiated; however, persistent and significant bleeding occurred at the scleral tunnel incision, necessitating an immediate cessation of the procedure to ensure the patient's safety. On postoperative day 1, B-ultrasound revealed obvious SCH and vitreous hemorrhage (VH; Figure 2A).

After 11d of symptomatic treatment, the SCH and VH had largely resolved (Figure 2A). At that time, serial B-scan ultrasonography demonstrated marked reduction in the height of the SCH without evidence of appositional ("kissing") choroidal detachment, and intraocular pressure (IOP) had stabilized. Based on these findings, the referral surgeon considered the hemorrhage to be in a resolving phase and proceeded with vitrectomy and lensectomy to remove the dropped nucleus. Suprachoroidal drainage was not performed concurrently, as there were no clinical or ultrasonographic signs of tense or non-resolving SCH, and premature drainage before complete liquefaction was considered to carry a high risk of rebleeding, particularly in the absence of confirmed coagulopathy at that time. The second vitrectomy and lensectomy were successfully performed under general anesthesia. Unexpectedly, on postoperative day 3, the patient's visual acuity dropped to no light perception, and IOP decreased to 8 mm Hg, with approximately 3 mm of inferior hyphema. An urgent B-ultrasound examination and color Doppler ultrasonography showed the recurrence of SCH, VH, and even possible RD (Figure 2B). Consequently, the patient was transferred to our hospital for further management. Upon referral, the management strategy followed classical principles for treating SCH, as the patient presented with a recurrent but

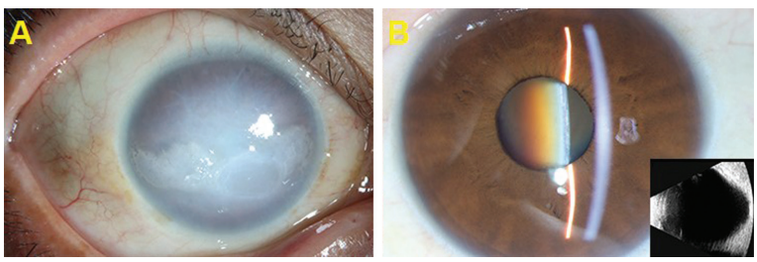


Figure 1 The anterior segment of the right eye (A), the left eye (B) prior to the first surgery, with corresponding B-scan ultrasonography (insert).

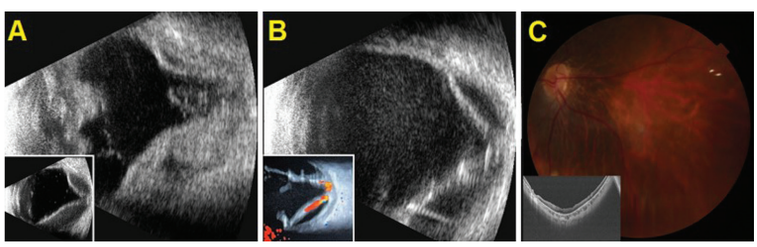


Figure 2 Postoperative examinations and follow-up findings A: Ultrasound images of post-first surgery. SCH and VH visible on post-operative day 1; gradual resolution of SCH documented on postoperative day 11 (insert). B: Ultrasound images of post-second surgery: SCH, VH, and possible RD observed on post-operative day 3; color ultrasound confirmed RD presence (insert). C: Fundus photograph with optical coherence tomography angiography (insert) obtained at the 3-month follow-up. RD: Retinal detachment; SCH: Suprachoroidal hemorrhage; VH: Vitreous hemorrhage.

non-appositional SCH and clinically low IOP. In accordance with established practice, we first aimed to stabilize the globe and then determine the cause of the recurrent hemorrhage before attempting any further intraocular manipulation.

Upon review of history, it was noted that the patient presented with axial high myopia (31.59 mm), moderate corneal astigmatism (1.42 D), finger joint deformities, and cleft palate. Systemic findings were discussed with the hematology, medical genetics, and rheumatology teams, which prompted a genetic evaluation. Whole-exome gene sequencing identified a heterozygous missense variant in collagen type V alpha 1 chain (*COL5A1*; c.685G>A, p.Asp229Asn), which causes cEDS.

Given that the patient had a blind right eye, salvaging vision in the left eye posed a significant challenge for the surgeons. To minimize the surgical scope while managing vascular risks, we first performed suprachoroidal drainage under general anesthesia to relieve serous fluid retention. Subsequently, an intraoperative fundus examination was carried out. Dramatically, during the surgery, no RD was found, contrary to the imaging results suggesting a funnel-shaped RD; instead, choroidal detachment that mimicked RD was observed. The following day, a B-ultrasound examination did not reveal any signs of SCH or RD in the left eye. At the three-month follow-up, the patient's best-corrected visual acuity improved to 20/200, with retinal flattening confirmed by fundus photography and optical coherence tomography angiography (Figure 2C).

DISCUSSION

EDS, which was named in 1949, is likely underdiagnosed, with an overall incidence of approximately 1 in 5000 to 1 in 100 000 individuals globally[1]. The diagnosis of EDS is based on family history, clinical examination, and clinical findings, and is ultimately confirmed *via* molecular genetic analysis. However, diagnosis is frequently delayed since the clinical manifestations of the disorder are subtle, and the diagnostic criteria are continually evolving, making it tough for ophthalmic clinicians to diagnose cEDS solely by eye appearance.

cEDS is primarily caused by mutations in either the *COL5A1* gene or the collagen type V alpha 2 chain (*COL5A2*) gene[1]. Aberrant coding in these genes results in the production of structurally compromised and functionally impaired type V collagen. Due to the collagen-rich nature of the eyes, collagen malformations are highly prevalent in this organ. This might thus offer an explanation for the abnormal ocular features observed in the patient: corneal astigmatism, posterior capsule weakness, and a long axial length. It is important to clarify, however, that while abnormal collagen metabolism in cEDS may contribute to ocular structural alterations, such as increased axial length, high myopia is an independent risk factor for SCH due to scleral thinning and choroidal vascular stretching. Therefore, the patient's cEDS-related connective-tissue weakness and her extreme axial myopia should be regarded as parallel, compounding vulnerabilities rather than as a linear causal sequence. Both conditions likely acted synergistically to increase the susceptibility to SCH in this case.

SCH is traditionally considered an uncommon yet devastating intraocular complication, and its spontaneous occurrence

is exceptional. However, patients with cEDS may face an increased risk of aneurysm and have also been associated with spontaneous arterial rupture. These manifestations are secondary to inadequate connective tissue support of the blood vessels and direct involvement of the vessel wall, rather than to abnormal platelet function or coagulation defects[1]. Moreover, blood vessel fragility is a hallmark of multiple EDS subtypes, including cEDS, which may also be the root cause of recurrent spontaneous SCH in our patient.

Surgery may be associated with increased complications in patients with EDS compared to the general population. Currently, there are no comprehensive and detailed guidelines available for performing ophthalmic surgeries on patients with cEDS. Several published case reports have focused on patients with other subtypes of EDS undergoing ocular surgery, describing various challenges encountered during these procedures[4-5].

To our knowledge, this is the first case that describes repeat SCH in a patient with cEDS. Our experience underscores the importance of careful preoperative thorough physical examination, even if it seems like a routine ocular surgery. Moreover, it highlights the necessity of prioritizing the reduction of surgical scope whenever no viable alternative treatments are at hand. This case also offers an important lesson from the initial management phase: in situations where significant SCH occurs following intraocular surgery, earlier consideration of timely suprachoroidal drainage may help prevent persistent choroidal elevation and reduce the risk of recurrent bleeding. Although the referring surgeon's decisions were reasonable based on the clinical information available at that time, our experience suggests that targeted evaluation for SCH drainage should be incorporated into the early management strategy, especially in eyes suspected structural or vascular fragility.

ACKNOWLEDGEMENTS

Authors' Contributions: Li ZY: data curation, writing-original draft. Deng WL: methodology; writing-review & editing. Zhang YL: conceptualization; writing-review & editing.

AI-Generated Content Disclosure: This work was prepared without artificial intelligence.

Foundation: Supported by the Science and Technology Research Program of Chongqing Municipal Education Commission (No.KJZD-K202400402).

Conflicts of Interest: Li ZY, None; **Deng WL,** None; **Zhang YL,** None.

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