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Acute bilateral Irvine-Gass Syndrome following uneventful cataract surgery in a patient without systemic risk factors

Síndrome de Irvine-Gass aguda bilateral após cirurgia de catarata sem complicações em paciente sem fatores de risco sistêmicos

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Abstract

We herein report a patient without risk factors who presented acute bilateral Irvine-Gass syndrome after uneventful phacoemulsification. The novelty of our case lies on the fact that the patient presented acute bilateral Irvine-Gass syndrome without a predisposing systemic disease. Even though Cystoid Macular Edema (CME) was somehow expected in the first eye because of the ocular history of trauma, prophylactic measures were not strong enough to avoid its development. Furthermore, those measures could not avoid developing CME in the second eye. A 44-years-old male who underwent cataract surgery in both eyes presented bilateral Irvine-Gass syndrome. Despite prophylactic measures, both eyes developed CME after uneventful cataract surgery. Regular treatment options could not solve the situation and intravitreal Anti-VEGF injections were needed. Bilateral cases of Irvine-Gass Syndrome are rare and generally associated with systemic risk factors. Patients who developed CME following their first cataract surgery should be counseled about the risks of developing the condition following surgery on the contralateral eye. On top of that, aggressive prophylactic measures should be encouraged to prevent CME in these cases.

Keywords: *Cystoid macular edema; Irvine-Gass syndrome; Cataract extraction; Phacoemulsification*

Resumo

Relatamos aqui um paciente sem fatores de risco que apresentou síndrome de Irvine-Gass bilateral aguda após facoemulsificação sem intercorrências. A novidade do nosso caso reside no fato de o paciente apresentar síndrome de Irvine-Gass bilateral aguda sem doença sistêmica predisponente. Embora o Edema Macular Cistoide (EMC) fosse de alguma forma esperado no primeiro olho por causa do histórico ocular de trauma, as medidas profiláticas não foram suficientemente fortes para evitar seu desenvolvimento. Além disso, essas medidas não puderam evitar o desenvolvimento de EMC no segundo olho. Homem de 44 anos submetido a cirurgia de catarata em ambos os olhos apresentou síndrome de Irvine-Gass bilateral. Apesar das medidas profiláticas, ambos os olhos desenvolveram EMC após a cirurgia de catarata sem intercorrências. As opções de tratamento regular não conseguiram resolver a situação e foram necessárias injeções intravítreas de Anti-VEGF. Casos de Síndrome de Irvine-Gass bilateral são raros e geralmente associados a fatores de risco sistêmicos. Os pacientes que desenvolveram EMC após a primeira cirurgia de catarata devem ser avisados sobre os riscos de desenvolver a doença após a cirurgia no olho contralateral. Além disso, medidas profiláticas agressivas devem ser incentivadas para evitar a EMC nesses casos.

Palavras-chave: Edema macular cistoide; Síndrome de Irvine-Gass; Extração de catarata; Facoemulsificação

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Introduction

Pseudophakic Cystoid Macular Edema (CME), also known as Irvine-Gass Syndrome, represents one of the most important causes of painless decreased visual acuity after cataract surgery. Even in uneventful cataract surgeries and while considering the modern surgical techniques of small incisional phacoemulsification, the incidence of clinically significant CME is about 0.1 to 3%.[1] It has been also demonstrated that asymptomatic CME is present in almost 10% of patients after cataract extraction when measured by Angiography or Optical Coherence Tomography (OCT), even in uneventful cataract extractions.[1,2]

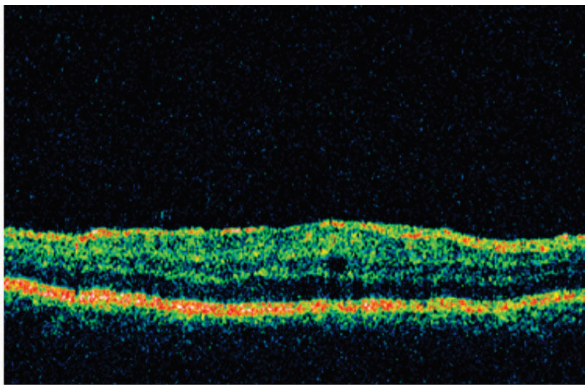
Irvine-Gass Syndrome is multifactorial; however, its exact pathogenesis is far to be fully elucidated. The consensus in the literature is that CME represents the end of the pathway of the inflammatory cascade triggered by an intraocular surgery, leading to the blood aqueous barrier breakdown and cystic accumulation of extracellular intraretinal fluid. Complicated cataract surgery, vitreomacular traction, posterior capsular rupture with vitreous loss and retained lens material are well known risk factors for the development of CME.[3] Also, some ocular comorbidities with high vasoactive profile like diabetic retinopathy or uveitis may increase the incidence of Irvine-Gass syndrome up to 20%.[2]

Although pseudophakic CME is usually self-limited with spontaneous resolution within 3-6 months after its presentation, some cases last more than 6 months and can lead to permanent vision loss.[4] Thus, the prevention and treatment of Irvine-Gass Syndrome represent a challenging situation for ophthalmologists around the world due to its ability to affect patients with no predisposing factors.

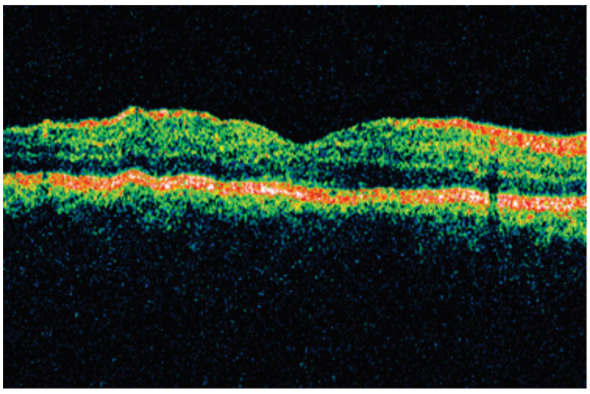
Case Report

A 44-years-old male was referred to our clinic for evaluation and treatment of progressive blurred vision in both eyes secondary to cataract. A complete ophthalmological evaluation was performed. Best Corrected Visual Acuity (BCVA) was 1/10 in the right eye (RE) and 8/10 in the left eye (LE) based on the Snellen chart, with no evidence of any other clinical ocular disease. Slip-lamp examination revealed a 4+ cortical, 2+ nuclear sclerotic cataract in the RE and 2+ cortical, 2+ nuclear sclerotic cataract in the LE. The only positive ocular history event was a direct ball strike in the RE eight years earlier with no visual affection. Systemic hypertension treated with oral enalapril 5 mg. Preoperative Stratus OCT showed an alteration of normal macular architecture with no signs of edema in the RE, while macular Stratus OCT in the LE was normal (Figure 1).

Prophylactic Bromfenac 0.09% drop was started one day prior to surgery. A standard phacoemulsification with intraocular lens implantation (single-piece acrylic, CT ASPHINA 509MP, Carl Zeiss) was performed in the RE. Postoperative medications included topical Difluprednate 0.05% and Gatifloxacin 0.5% four times a day for two weeks and Bromfenac 0.09% four times a day for one month. After one week, BCVA was 10/10, but the visual acuity decreased to 9/10 in postoperative week 6. No signs of inflammation were detected in both anterior chamber and vitreous. Dilated fundus examination revealed loss of foveal depression in RE and Stratus OCT evaluation revealed macular edema with hyporeflective cystic intraretinal spaces in the RE, while LE was normal (Figure 2). Topical Prednisolone 1% and Nepafenac 0.1% four times a day were started, but the patient showed no signs of improvements and at postoperative month 3 the BCVA decreased to 6/10. Intravitreal Anti-VEGF injection was prescribed and the CME resolved within 1 month, with final BCVA of 10/10 in the RE and normal Stratus OCT scans.

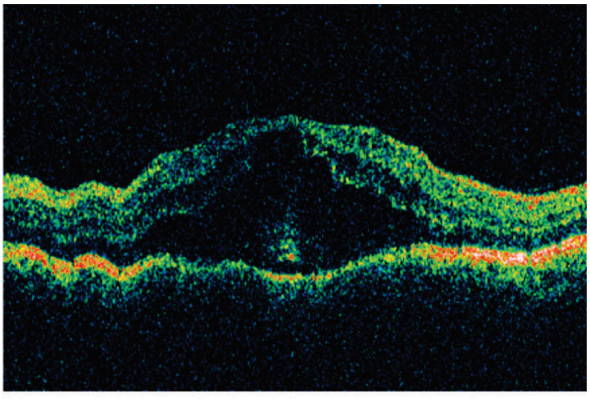


Preoperative OCT scan in the right eye



Preoperative OCT scan in the left eye

Figure 1: Preoperative macular OCT scans in both eyes.



Macular OCT scan in the right eye 3 months after surgery

Figure 2: Postoperative macular Stratus OCT scan in right eye

After that, the patient underwent standard phacoemulsification with intraocular lens implantation (single-piece acrylic, CT ASPHINA 509MP, Carl Zeiss) in the LE. Surgery was completed without any intraoperative complications. Preoperative Nonsteroidal Anti-Inflammatory Drugs (NSAIDs) and postoperative topical medications were indicated with the

same regime as for RE. Postoperative BCVA was 10/10 at day 7, but worsening was demonstrated 90 days after surgery. BCVA decreased to 8/10 and a Stratus OCT revealed cystoid macular edema in the LE (Figure 3). Treatment for CME was started with topical Prednisolone 1% and Nepafenac 0.1% four times a day, plus intravitreal injection of Anti-VEG in the LE. Complete resolution of cystic intraretinal fluid was achieved one month after treatment, with final BCVA of 10/10 in the LE.

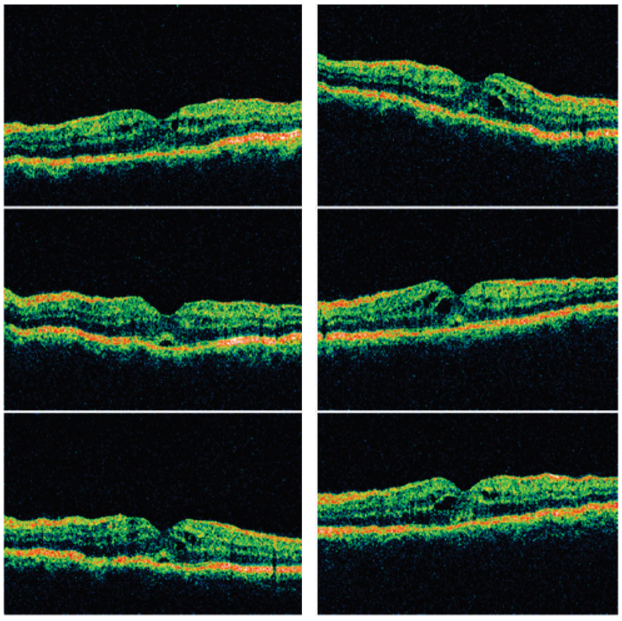


Figure 3: Postoperative macular Stratus OCT scan in left eye

Discussion and Conclusion

Bilateral cases of CME after cataract surgery have been barely described in the literature.[5,6] Most of the time, bilateral CME were reported in association with systemic diseases including Crohn's disease and infections.[7]

The differential diagnosis of pseudophakic CME is broad, and patients with underlying ocular and systemic conditions that predispose them to other causes of CME may confer diagnostic challenges. CME can also be seen in diabetic macular edema, retinal vein occlusion, radiation retinopathy, hypertensive retinopathy, uveitis, topical prostaglandins, retinal dystrophies, choroidal tumors, leukemia and chronic renal failure, among others.

Irvine-Gass syndrome following uncomplicated phacoemulsification on the patient's first eye is not uncommon. The patient has a history of ocular trauma in the right eye 8 years before surgery and preoperatory Stratus OCT scans showed images of altered macular architecture, both situations that can be expected to increase the risk of developing CME after cataract surgery. This was discussed with the patient and consulted with retina specialists, who recommended prophylactic NSAIDs treatment to be started before surgery. The use of NSAIDs as a routine prophylactic scheme in uncomplicated cataract surgery and even in patients without risk factors for CME has been widely described in the literature. It has been demonstrated that the use of prophylactic NSAIDs in combination with topical steroids in the perioperative period can reduce the incidence of CME after routine cataract surgery.[8-10]

The development of Irvine-Gass syndrome in the second eye following uneventful cataract surgery, despite topical prophylactic measures, was unexpected. The patient's medical history contained no predisposing factors for the development of CME, and had no history of ocular trauma and macular Stratus OCT scans were completely normal. Although it is probable that those patients who develop CME after cataract surgery in one eye are at increased risk of developing it following surgery in the second eye, there is no strong evidence that can anticipate the exact level of risk of these individuals. Thus, specific prophylactic measures in these particular patients are still under debate. As was formerly said, several clinical studies recommend a combination of NSAIDs and steroids for the reduction of CME after phacoemulsification. Further, oral Indomethacin has also been indicated for the prevention of pseudophakic CME.[11]

There is no general consensus about the best therapeutic options for treating CME. While the vast majorities of cases are self-limited and show spontaneous resolution within 3-6 months, rapid visual recovery after cataract surgery is almost mandatory nowadays considering that patients have high social activities and/or vision-depending occupations. In a systematic review, Kessel et al.9 found that topical NSAIDs are more effective than steroids in controlling postoperative inflammation and preventing CME, while Wittpenn et al.[10] found a synergistic effect when combining NSAIDs and steroids for this purpose.

Periocular and intravitreal corticosteroids have been also used for the treatment of CME, especially in refractory cases. Thach et al.[12] demonstrated in a series of 48 patients with refractory CME, that sub-Tenon's injections improved visual acuity from 20/92 to 20/50 ($P = 0.0001$), and retrobulbar injections improved visual acuity from 20/97 to 20/58 ($P = 0.035$), and there was no statistical outcome difference between the two techniques, while Benhamou et al.[13] reported using 8 mg of intravitreal triamcinolone to treat 3 cases of refractory chronic pseudophakic CME, and showed that macular thickness and visual acuity improved, but the effects were transient.

Recently, intravitreal Anti-VEGF have been used for treating refractory CME after cataract surgery due to its anti-inflammatory effects. Several reports and case series showed interesting results of Anti-VEGF for reducing macular edema and improving visual acuity in patients with pseudophakic CME.[14-17] Nevertheless, further prospective controlled studies are needed to compare the long-term safety and efficacy between intravitreal Anti-VEGFs and other treatment options in cases of refractory Irvine-Gass syndrome.

The novelty of our case lies on the fact that the patient presented acute bilateral Irvine-Gass syndrome without a predisposing systemic disease. Besides CME was somehow expected in the first eye because of the ocular history of trauma, prophylactic measures were not strong enough to avoid it development. Further, those measures could not avoid developing CME in the second eye.

In conclusion, CME remains an important cause of decrease visual acuity after cataract surgery. Bilateral cases of Irvinge-Gass Syndrome are rare and generally associated with systemic risk factors. Patients who develop CME following their first cataract surgery should be counseled about the risks of developing the condition following surgery on the contralateral eye. Even more, aggressive prophylactic measures should be encouraged to prevent CME in these cases.

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