Spontaneous scleral rupture associated with retinochoroidal coloboma

Ruptura espontânea de esclera associada a coloboma coriorretiniano

Jefferson A. S. Ribeiro 1,2  https://orcid.org/0000-0001-5995-9193
Deborah Conrado 2 https://orcid.org/0000-0002-3630-7528
Ricardo Carvalho 2 https://orcid.org/0000-0002-3429-1817
Daniel S. Ribeiro 2 https://orcid.org/0000-0002-8405-182X

ABSTRACT

We reported a case of ocular hypotony due to spontaneous scleral rupture in retinochoroidal coloboma and the treatment which was performed. This is a prospective case report in which a 21-year-old woman complained of abrupt vision loss in her left eye. Ocular hypotony due to spontaneous scleral rupture in retinochoroidal coloboma was identified through tests. An ultrasonography confirmed the scleral lesion but magnetic resonance imaging and fluorescein angiography have also proven to be useful. Due to the posterior location of the perforation, we opted for a sub-tenon injection of autologous blood and intraocular gas tamponade, which were successful, resulting in improved visual acuity and intraocular pressure. In this case, an alternative to invasive surgical procedure in the treatment of spontaneous scleral perforation and retinochoroidal coloboma was presented.

Keywords: Eye coloboma; Ocular hypotension; Scleral diseases; Rupture; Ultrasound

RESUMO

Relatamos um caso de hipotensão ocular secundária a ruptura escleral espontânea em coloboma coriorretiniano e o tratamento realizado. Trata-se de estudo de caso prospectivo de uma mulher de 21 anos de idade que se apresentou queixando perda súbita de visão no olho esquerdo. Ao exame, foi evidenciado hipotensão ocular em virtude de ruptura escleral espontânea de coloboma coriorretiniano. Ultrassonografia confirmou a lesão da esclera embora ressonância magnética e angiofluoresceinografia também tenham sido úteis. Em virtude da localização posterior da perfuração, optamos por injeção sub-tenoniana de sangue autólogo e tamponamento intraocular com gás, que demonstraram ser úteis, resultando em melhoria da acuidade visual e da pressão intraocular. Neste caso, apresentamos uma alternativa a procedimento cirúrgico invasivo para tratamento de perfuração espontânea de esclera em coloboma coriorretiniano.

Descritores: Coloboma ocular; Hipotensão ocular; Ruptura da esclera; Ultrasom

1 Superior School of Health Sciences, Universidade do Estado do Amazonas, Manaus, AM, Brazil.
2 Instituto de Oftalmologia de Manaus, Manaus, AM, Brazil.

Institution where the work was completed: Instituto de Oftalmologia de Manaus

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**INTRODUCTION**

Retinochoroidal colobomas are developmental disorders often associated to other ocular conditions, such as retinal detachment.\(^1\) They are rarely related to spontaneous scleral rupture.\(^2\)

Herein we describe a case of ocular hypotonia due to spontaneous scleral rupture in retinochoroidal coloboma.

**Case report**

A 21 year-old female was admitted to the emergency unit complaining of abrupt visual loss in her left eye for less than 24 hours. She had reported no previous history of visual impairment and no systemic diseases. Corrected visual acuity was 20/20 OD and 20/100 OS, tonometry was 12 mmHg OD and 6 mmHg OS and fundoscopy revealed a swollen optic disc (Figure 1A), choroidal folds in the macula and a nasal retinochoroidal coloboma (Figure 1B) in her left eye, OD fundoscopy was unremarkable. A systemic investigation was carried out in order to exclude inflammatory diseases of the optic nerve and uveal tract, and no other manifestations were present. Ultrasonography and magnetic resonance imaging evidenced the coloboma in OS with suggestion of mild collection of fluid posterior to the eye wall in MRI. Ultrasonography evidenced the scleral rupture in the nasal border of the coloboma (Figure 1C). Fluorescein angiography showed staining of the optic disc and border of the colobomatous lesion with an oval area stain in the nasal aspect of the coloboma, at the supposed site of the rupture (Figure 1D). Due to the location of coloboma and scleral rupture, our treatment option was a sub-tenon injection of 3 ml of autologous blood intravitreal 100% C3F8 0,3 ml followed by the immediate postoperative period by prophylactic laser photocoagulation around the colobomatous lesion. BCVA had improved one week after the procedure to 20/40 OS as well as the ocular pressure to 10 mmHg. Three months after the treatment, visual acuity was 20/20 and tonometry was 12 mmHg OS with improvement of macular folds and optic disc swelling (Figure 2).

**DISCUSSION**

Spontaneous scleral rupture associated with retinochoroidal colobomas is a rare event, as there have only been a few cases reported in literature.\(^2\) - \(^6\) The clinical picture is often of macular hypotony ensuing complaints of decreased vision, as it was evident in the present case, in which the patient complained of abrupt vision loss unassociated with trauma or strain. Due to the rarity of disease, a systemic clinical work up has been performed in order to exclude optic neuritis or other inflammatory conditions of uvea and sclera that may be presented with optic disc edema and macular folds.

Imaging is useful in identifying the site of scleral rupture although it may sometimes be difficult to clearly point out the lesion on an MRI or US, and exploratory surgery may be necessary.\(^2\) MRI and US may also show fluid behind the scleral lesion.\(^4\)\(^,\)\(^7\) In the present case, US identified the scleral rupture at the nasal border of the colobomatous lesion, next to the optic nerve, but MRI only suggested a fluid collection behind the globe. An interesting point was the finding of an oval area of staining in the nasal aspect of coloboma that may correspond to the site of the scleral rupture. Treatment options frequently involve scleral patching to the damaged area.\(^2\)\(^,\)\(^5\)\(^,\)\(^6\) Scleral buckle, scleral patch, dural patch and polytetrafluoroethylene patch were the treatments employed in similar cases in literature,\(^2\)\(^,\)\(^6\) although an autologous blood injection was the choice in other cases for spontaneous scleral rupture related to posterior staphyloma which is difficult to access by surgery.\(^8\)\(^,\)\(^9\) Although a spontaneous closure of the scleral lesion may occur,\(^9\) treatment options aim to increase ocular pressure by inducing a fibrotic scar formation in the scleral defect.\(^4\)\(^,\)\(^5\) In the present case, due to the posterior location of the scleral rupture next to the optic nerve, we intended to avoid surgical exploration and instead we chose a less invasive method that consisted of a subtenon autologous blood injection. In order to tamponade the lesion and improve ocular pressure, C3F8 was also injected in the vitreal cavity, and prophylactic laser was performed around the colobomatous lesion.

So, ocular hypotony may be the manifestation of spontaneous scleral rupture associated with retinochoroidal coloboma.
and treatment often leads to successful vision restoration. US, MRI and also fluorescein angiography may be useful in identifying the site of the rupture and treatment plan. Surgery is effective in most cases but a less invasive approach consisting of a sub-tenon injection of autologous blood may also be successful.

REFERENCES


Corresponding author:
Jefferson A. S. Ribeiro
Av. Maneca Marque, 55, apto 402
Parque 10 de Novembro, Manaus, Amazonas, Brazil.
Zipe code 69055-021
Phone: 55 92 98233-4371
E-mail: jef_ribeiro@yahoo.com.br