Cyclophotocoagulation-induced sympathetic ophthalmia in a Coats' disease patient supported by histopathology and immunohistochemistry

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We describe a case of a 13-year-old male patient of Coats' disease who developed sympathetic ophthalmia (SO) following contact diode laser cyclophotocoagulation. There was no history of invasive surgery or any perforating injuries preceding cyclodestructive therapy. The eye had neovascular glaucoma secondary to Coats' disease, which was treated once with contact cyclophotocoagulation. Subsequently, the intraocular pressure slowly decreased, and the eye became phthisical. Intraocular inflammation developed in the fellow eye and SO was suspected, which was confirmed by characteristic findings seen on fluorescein angiography. The case was successfully managed with the help of topical and systemic immunosuppression. Enucleation with silicone ball implantation was performed in the right phthisical eye and specimen was sent for histopathological examination. Histopathology and immunostaining supported the diagnosis of SO.

Key words: Coats' disease, diode laser cyclophotocoagulation, fluorescein angiography, neovascular glaucoma, sympathetic ophthalmia

Sympathetic ophthalmia (SO) is an uncommon bilateral panuveitis probably related to an autoimmune response to uveal antigen and usually occurs 4–8 weeks after the precipitating traumatic event.[1]

Diode laser cyclophotocoagulation (DLCP) is considered to be a safe, simple, and effective modality in the management of refractory glaucoma, neovascular glaucoma, and glaucoma in a blind painful eye.[2] A number of complications have been reported following diode cyclodestruction, including SO.[3]

We describe a case of neovascular glaucoma secondary to Coats' disease in which DLCP was performed to obtain control of intraocular pressure (IOP), and SO occurred 6 weeks after treatment.

Case Report

A 13-year-old boy presented with severe loss of vision in the right eye (RE) over the last month, not associated with penetrating injury or any ocular surgery. Best-corrected visual acuity (BCVA) was light perception in RE and 20/20 in the left eye (LE). RE showed relative afferent pupillary defect and vitreous cells. IOP of both eyes was 20 mmHg. The patient was diagnosed as Coats' disease with exudative retinal detachment (RD) in RE. There were no pathologic findings in LE. Over a period of 18 months, he developed neovascular glaucoma in RE with an IOP of 45 mmHg. His BCVA in LE was 20/20 and an IOP of 20 mmHg. The patient was treated with DLCP to superior and inferior half (120° each) of RE ciliary body. Twenty-five diode laser burns of 2000 ms duration and using a power of 2000 mW were applied.

Six weeks after cyclophotocoagulation, he experienced photophobia and sudden deterioration of vision in his LE and mild pain in RE. BCVA in LE was 20/63 for distance and near vision was N18. IOP of RE was 21 and in LE 15 mmHg. Slit lamp examination revealed anterior chamber cells, flare and keratic precipitates in both eyes, and a dense cataract precluding fundal view in RE. Fundus examination of LE revealed disc hyperemia and yellow-white subretinal infiltrates that partly obscured choroidal vasculature [Fig. 1a]. Ultrasound B-scan of RE showed total RD with increased choroidal thickness, and LE showed shallow RD with increased retinochoroidal thickness. Ocular coherence tomography showed retinal elevation due to the accumulation of subretinal fluid in LE [Fig. 1c]. Fluorescein angiography of LE revealed multifocal punctate hyperfluorescent spots in the early venous phase which then continued to late leakage [Fig. 1b].

A diagnosis of SO was made, and the patient was treated with intravenous methylprednisolone (500 mg/day for 3 consecutive days) followed by oral prednisone. Over the next 20 days, intraocular inflammation in both the eyes reduced and visual acuity returned to 20/20 in the LE. Fundus examination revealed resolution of subretinal infiltrate and exudation. Although RE became phthisical, the patient had complaints of repeated pain and discomfort. The patient preferred removal of the phthisical eyeball for cosmetic reasons as well as the pain and discomfort. Enucleation with silicone ball implant was

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done after obtaining informed consent, and specimen was sent for histopathological examination. LE remained unremarkable with BCVA of 20/20.

Gross examination of vertical section of RE ball showed small whitish calcified areas with detached retina, vitreous hemorrhage, and thickened sclera posteriorly [Fig. 2]. Microscopic examination under hematoxylin and eosin stain revealed corneal edema with neovascularization. The limbal area showed congestion with inflammatory cell infiltrate [Fig. 3]. The anterior chamber was deep with angle closure at one end. Iris neovascularization with ectropion uvea was noted. Lens was partially cataractous with membranous structure in front of it. Ciliary body showed atrophic changes. There was exudative RD with fibrotic retinal structure. Retinal telangiectatic vessels with numerous cholesterol clefts in the exudation with focal osseous changes noted [Fig. 4a-d]. There was diffuse choroidal involvement with lymphocytic infiltration sparing the choriocapillaris [Fig. 5]. Immunohistochemistry showed CD 20 (B-cell) and CD 3 (T-cell) positivity [Fig. 6a and b].

Discussion

SO is a clinical diagnosis; there are no specific tests. The diagnosis is suggested by the appearance of a bilateral uveitis occurring after penetrating, accidental or surgical trauma or after nonpenetrating cyclodestructive procedures. SO should be considered in patients who develop bilateral uveitis[3] following cyclodestructive procedures.

DLCP has been shown to be effective in lowering IOP in end-stage glaucoma. Due to its more targeted destruction, it produces less inflammation[4] and therefore fewer side effects than those produced by cyclocryotherapy and Nd:YAG cyclophotocoagulation.[5] Albahlal *et al*.[6] suggested that the incidence of SO after DLCP was computed as 0.001%, which

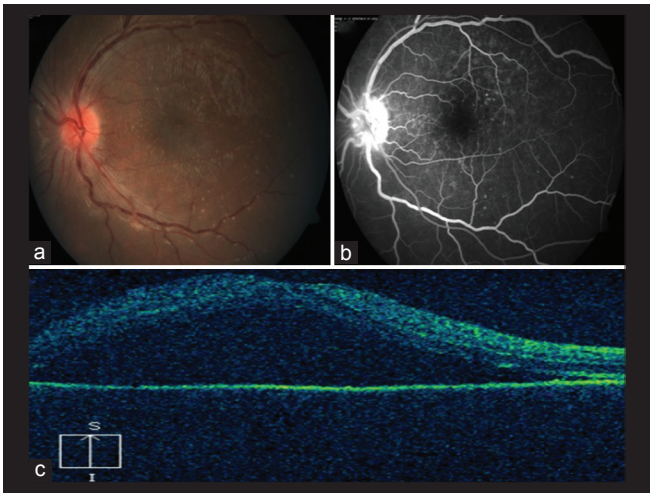

Figure 1: (a) Color fundus photograph of the left eye showing multiple yellow-white lesions at the level of the pigment epithelium, multifocal exudative retinal detachment, and hyperemic optic disc. (b) Fluorescein angiogram showing multifocal leaks in the posterior pole. (c) Optical coherence tomography demonstrating serous retinal detachment

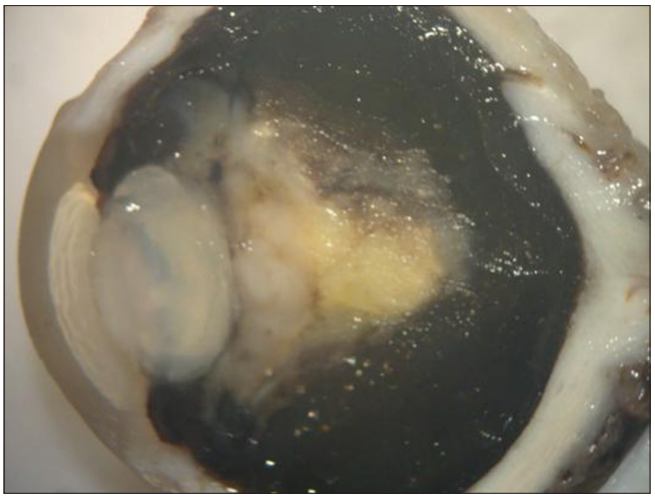

Figure 2: Gross section of enucleated eyeball showing massive thickness of the choroid and with retrolental grayish-white condensed exudates

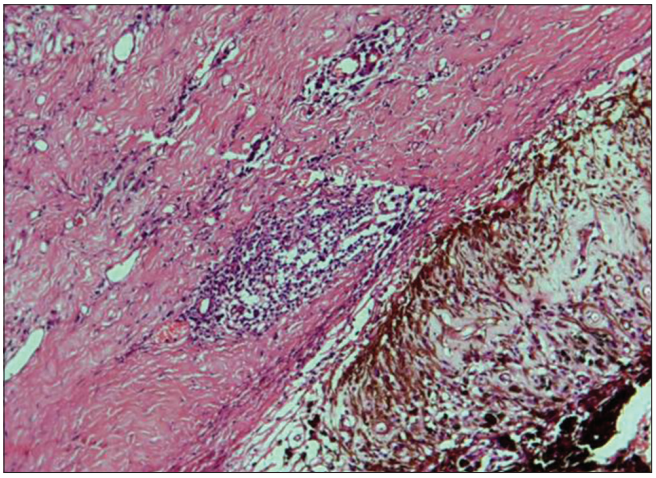

Figure 3: Inflammation of the limbus with adhesion of the iris to the peripheral part of cornea, which was the site of previous diode laser cyclophotocoagulation

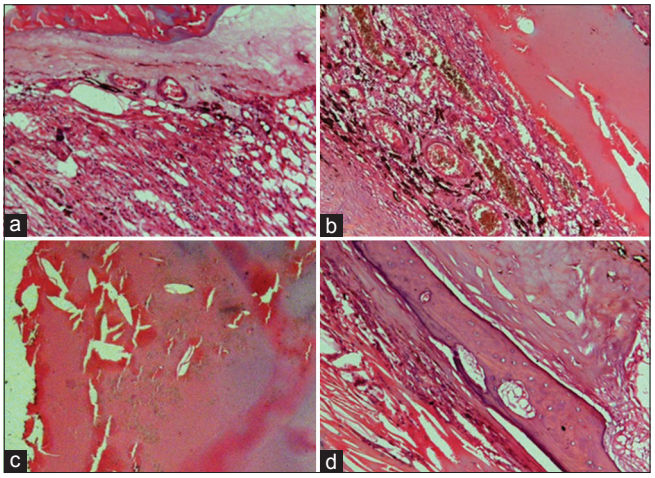

Figure 4: (a) Telangiectatic vessels, (b) telangiectatic vessels with exudation, (c) cholesterol clefts within the exudation, and (d) bone formation

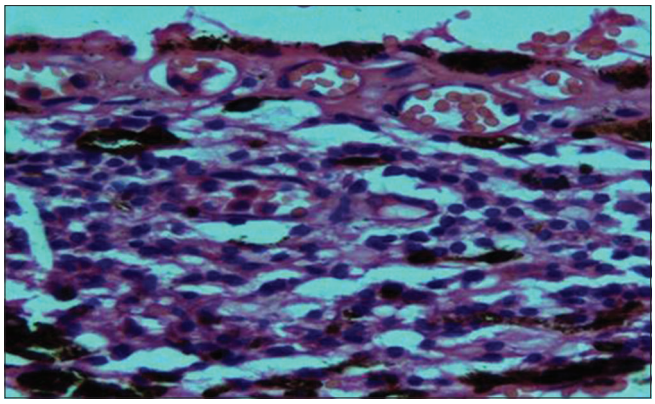

Figure 5: Lymphocytic infiltration involving the choroid and sparing choriocapillaris suggestive of sympathetic ophthalmia

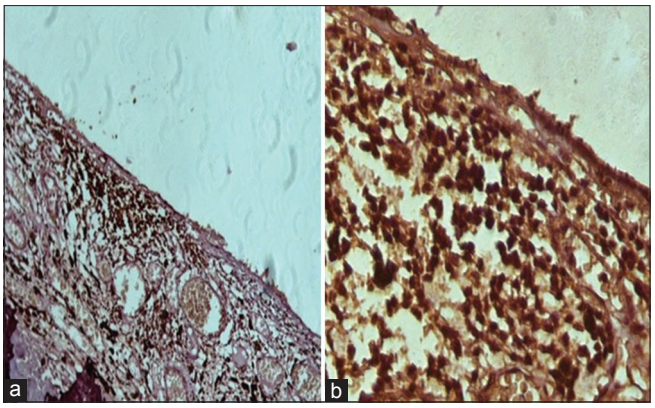

Figure 6: Showing immunohistochemistry with (a) CD 20 (B-cell) + positivity (×10) and (b) showing CD 3 (T-cell) +++ positivity (×40) (BioGenex, 49026 Milmont Drive, Fremont, CA 94538, USA)

is quite low compared with that reported after noncontact and contact Nd:YAG laser (5.8% and 0.67%, respectively)-related cyclodestruction. However, all these cases of SO had a history of multiple ocular surgeries which made investigators question the role of laser procedures as an inciting event and considered laser treatment as a triggering factor.[6] Taking these observations into consideration and the low rate of SO, the role of the diode laser as a trigger should be interpreted with caution.

We describe a case of neovascular glaucoma secondary to Coats' disease in which DLCP was performed and SO occurred 4 months after treatment. Pathological examination revealed limbitis with congestion which could be the sequelae of previous DLCP. Exudative RD, retinal telangiectasia, and cholesterol clefts were suggestive of previous Coats' disease. Gross thickening with chronic inflammation of the choroid with sparing of superficial choriocapillaris[7] was consistent with SO. Characteristic Dalen-Fuchs nodules could not be appreciated on histopathology examination. The predominance of T-lymphocytes on immunostaining expressing the suppressor/cytotoxic phenotype mediated delayed hypersensitivity reaction presumably directed at antigens in SO.[8]

The influence of Coats' disease in the pathogenesis of SO is not known. The increased vascular permeability and the high number of inflammatory cells present in eyes with Coats' disease could theoretically promote the presentation of uveal autoantigen to lymphocytes, which is believed to be a crucial step in the development of SO.[9]

We found only two cases of SO in Coats' disease after Nd:YAG cyclotherapy and one case of SO after cyclocryotherapy in the literature.[9,10] To the best of our knowledge, this is the first reported case of SO following transscleral DLCP in Coats' disease.

To conclude, SO following diode laser CPC is an uncommon entity. Sympathizing and inciting eyes followed a fairly typical clinical course with prominent posterior segment findings in the LE. Aggressive topical and systemic immunosuppression yielded adequate control of inflammation and good visual outcomes.

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Conflicts of interest

There are no conflicts of interest.

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