A Combined Treatment Approach for Stage 3A Coats’ Disease

Abstract
A seemingly healthy young male presents with unilateral floaters. Fundus examination reveals subretinal exudate, hemorrhages, and retinal detachment identified as stage 3A Coats’ disease. This case demonstrates a combined treatment with cryotherapy and triamcinolone injection.

I. Case History
- An 18 year-old Caucasian male patient presents for his first eye exam with the chief complaint of floaters OD.
- The floaters occurred 4-5 months ago and appear grey in color. In the past he had noticed multiple floaters at times, however presently sees only one reoccurring in his peripheral vision.
- Patient reports good distance and near vision, and denies flashes, loss of vision, double vision, headache, browache, ocular pain or discomfort.
- The patient has no remarkable ocular or medical history.

II. Pertinent Findings

Clinical

Entering VA: OD: 20/50, OS: 20/20
BCVA: OD: 20/20-1, OS: 20/20
Pupils: PERRL (-) APD OU
(-)Leukocoria
IOP GAT: OD 17mmHg, OS 17mmHg
Slit lamp exam of the anterior segment: Unremarkable
DFE:

-OD:
Lens: Clear
Vitreous: Large central floater
Optic Nerve Head: .5/.5 NRR intact
Vessels: 2/3, H0, A0
Macula: ERM and suspected edema with trace thickening
Periphery: Significant peripheral subretinal exudate from 4-11 o’clock positioning, scattered dot/blot hemorrhages, temporal exudative retinal detachment (macula on)

-OS:
Lens: Clear
Vitreous: Clear
Optic Nerve Head: .5/.5 NRR intact
Vessels: 2/3, H0, A0
Macula: Flat and intact
Periphery: Flat and intact, no breaks or detachments

Additional Testing

Fundus Photography: To document a baseline image of the retina to compare with in the future to determine progression or resolution with time and/or treatment.

- Fundus OD: Displays normal ONH, possible macula edema, peripheral exudative retinopathy with scattered lipid predominantly Inf/Temp, peripheral temporal telangiectasia and aneurysms, shallow temporal exudative detachment from 7-10 o’clock.
- Fundus OS: Displays normal ONH, normal macular, normal periphery.

Macula OCT: An essential tool to aid in determining if there is truly macula edema and subsequent macula thickening as suspected on dilated fundus examination.

HD 5 Line Raster:

- OD: Presents a macular area with a preserved foveal contour and thickened retinal tissue consistent with macula edema.
- OS: Not Imaged

Macular Cube: More appropriate to quantify thickening.

- OD: General macular thickening, center thickness 308um.
- OS: Not Imaged

Fluorescein Angiography:

- OD: Normal AV transit, late hyperfluoresence consistent with macula edema, hyperfluoresence from telangiectasis.

B-Scan:

- OD: Echographic findings consistent with limited serous retinal detachment, peripheral hyperechoic signals within retina, not choroid. No choroidal mass detected.

III. Differential Diagnosis

Primary Diagnosis:

- Coats’ Disease
Differential Diagnosis:
- Retinopathy of Prematurity - Norrie's Disease
- Retinoblastoma, - Toxocariasis
- Retinal Astrocytoma - Eales Disease
- Choroidal/ Intracranial mass

IV. Diagnosis and Discussion

Definitive diagnosis: Coats’ Disease OD Stage 3A.

- The patient's retinal appearance of significant exudate, telangiectasia, and aneurysms with the consequent exudative detachment establish the patient as Stage 3A.
- The presence of macula thickening depicts disease progression to the macular area and a direct danger of vision loss with further progression. Likely to occur without treatment intervention in a present 20/20-1 eye.
- Coats’ disease is a predominantly unilateral disorder affecting only one eye in over 75% of cases and occurs most commonly in young males, with a ratio of 3:1 (male/female), occurring in the first decade of life in 60-70% of cases.
- The initial presentation is often blurred vision, leukocoria, or strabismus.
- Coats' disease is a rare idiopathic retinal vascular disease where abnormal vessel permeability results in an exudative retinopathy and exudative detachments. Also pathognomonic of the disease is the presence of telangiectatic retinal vessels and retinal vascular aneurysms.
- George Coats first described the disease, which he termed an exudative retinopathy, as a unique idiopathic ocular disorder manifesting as retinal telangiectasis with massive intraretinal and subretinal exudation that can cause severe vision loss. Coats classified his cases of exudative retinopathy into three groups, based off of their morphologic presentation.
- Shields classified Coats’ into five stages based off of its clinical manifestations, making it useful for clinical prognosis and treatment.

V. Treatment and Management

- The patient was sent urgently to a retinal specialist for confirmation of diagnosis and treatment.
- Treatment options were discussed including laser, cryotherapy, intravitreal triamcinolone, intravitreal anti-VEGF injection, and surgery. Cryotherapy with or without intravitreal triamcinolone was recommended as first line treatment.
- Cryotherapy and intravitreal Kenolog injection was performed 19 days after initial diagnosis.
- The retina showed regressed telangiectasis, a few persistent retinal hemorrhages, well-treated serous RD with cryotherapy, and improvement in inf/temp subretinal exudates after initial treatment.
- Two further laser photocoagulation procedures to aid with vessel regression were performed on subsequent examinations.
- Presently patient presents with moderate regression of the aneurysmal vessels, slight reduction of the subretinal exudate, a flat macula, and a remaining shallow temporal exudative detachment post treatment.
- Patient is undergoing regularly scheduled follow-ups with tentative additional treatment.

VI. Conclusion

- The treatment objective of Coats’ disease is the destruction of the abnormal retinal vessels with the goal of eliminating the retinal exudation.
- The management plan is entirely dependent on the severity of the disease with the goal of restoration of the retina.
- Laser photocoagulation and cryotherapy are often primary management options to ablate the anomalous vessels.
- Triamcinolone and anti-VEGF drugs are also used as adjunct treatments to help reduce the subretinal fluid and ensure recovery.
- In advanced cases with severe retinal detachments surgical means can be utilized.
- In the presence of severe exudative retinopathy and retinal detachment such as in our patient, the efficacy of laser ablative therapy is diminished and cryotherapy is a more efficacious option.
- In a study by Bergstrum combination cryotherapy and triamcinolone injection for exudative detachment from severe Coats’ disease, reduced subretinal fluid in all patients. However there was an observed risk for patients to develop an inoperable retinal detachment with severe proliferative vitreoretinopathy.
- A study by Lin from 2005-2011 reviewed reported Coats’ cases and the outcomes of the varied treatment modalities. The retrospective study discovered that eyes treated with combination cryotherapy and intravitreal Bevacizumab showed a similar risk for vitreous fibrosis that evolved into a tractional retinal detachment. Ramasubramanian and Shields proposed the vitreoretinal fibrosis was caused by the rapid absorption of subretinal exudates and upregulation of transforming growth factor beta.
- The results of combination triamcinolone and cryotherapy treatment can be successful, however holds inherent risk. When deciding on treatment, this risk must be weighed against the potential benefit of treatment.
Our patient shows early macula involvement; hence the utmost importance lies on the preservation of vision by halting the disease's progression towards the posterior pole and macula.

The perplexities of Coats' disease have plagued researchers since 1908. Though one thing remains clear, due to the extreme variability of Coats’ disease, treatment must be determined by the presenting severity.

Coats' disease presents a great risk to a patient's vision, but with a prompt diagnosis and an understanding of the intricacies of the disease and treatment, vision can be preserved and even improved.

References


