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Adult Onset Coats' Disease

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Abstract

Coats' disease is a condition involving retinal telangiectasias and retinal vessel aneurysms causing subretinal and intraretinal exudation and lipid accumulation in healthy individuals. It is found to occur most frequently unilaterally in young males. In the case of two Latino male patients diagnosed with adult onset Coats' disease, non-macular peripheral exudation and telangectasias were evident. Fortunately, due to lesion location, both maintain good central vision. Current treatment options for Coats' include laser photocoagulation, steroid injection and intravitreal anti-vascular endothelial growth factor (VEGF) injections. Careful evaluation of location and severity of lesions must be assessed prior to determination of treatment modality.

Introduction

Coats' disease, first described in 1908 by ophthalmologist George Coats',[1] involves idiopathic retinal telangiectasias and aneurysms of retinal blood vessels that result in vascular leakage, hemorrhaging, capillary dropout, and exudation into surrounding tissues in the absence of vitreoretinal traction.[2-5] In Coats' disease, retinal vessels exhibit hyperpermeability and a breakdown of the blood retinal barrier.[6] This is thought to be caused by high levels of vascular endothelial growth factor (VEGF) consistent with a deficiency in the protein product norrin in the eye, suggesting its association as a causative factor in the disease.[7-10] VEGF is necessary for normal vascular development and angiogenesis because of its high selectivity for vascular endothelial cells, but in diseased states it is secreted by hypoxic retinal pigment epithelial (RPE) cells. As a result, aberrant neovascularization develops in eyes with elevated levels of VEGF.[11] Often the progression of exudation and lipid deposition[7] can lead to a total exudative retinal detachment.[12] Patients may be asymptomatic or present with decreased visual acuities, blindness, leukocoria, strabismus, pain, and/or nystagmus.[4,12,14]

In the case of the two Latino male patients diagnosed with adult onset Coats' disease, non-macular peripheral exudation and telangectasias were discovered on a routine comprehensive exam.

Case 1

A 49-year-old male Latino patient presented to clinic for a comprehensive eye exam with no entering complaints. His past medical history was significant for elevated liver function tests, depression and osteoarthritis. On examination his best-corrected visual acuity was 6/6 (20/20) OD and OS with a plano DS and +0.25 DS correction, respectively. Pupils were normal, round and equally reactive to light, extraocular muscle movements were full OU, confrontation visual fields were full to finger count both eyes, and he was orthophoric in primary gaze. Anterior segment evaluation of both eyes was found to be within normal limits. Intraocular pressures were 19 mmHg OD and 18 mmHg OS with Goldmann applanation tonometry. Blood Pressure was found to be 127/83 mmHg

The dilated fundus examination revealed clear media, mild arteriovenus nicking OS, and a cup to disc ratio of 0.30 round with distinct margins and healthy rim tissue OU. A drance hemorrhage was noted OS (Fig. 1A,B). Both maculas were found to be flat and avascular. The left eye was notable for extensive exudative material temporally from 1 to 5 o'clock, denser inferotemporally bordering a large intraretinal hemorrhage. The right eye was notable for inferotemporal telangectasias and micro aneurysms without exudation. The left eye was also significant for 2+ arterial sheathing and large areas of temporal lipid exudates with subretinal fluid outside the macula.

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The authors have no financial or proprietary interest in any material mentioned in this article.

This article was peer-reviewed.

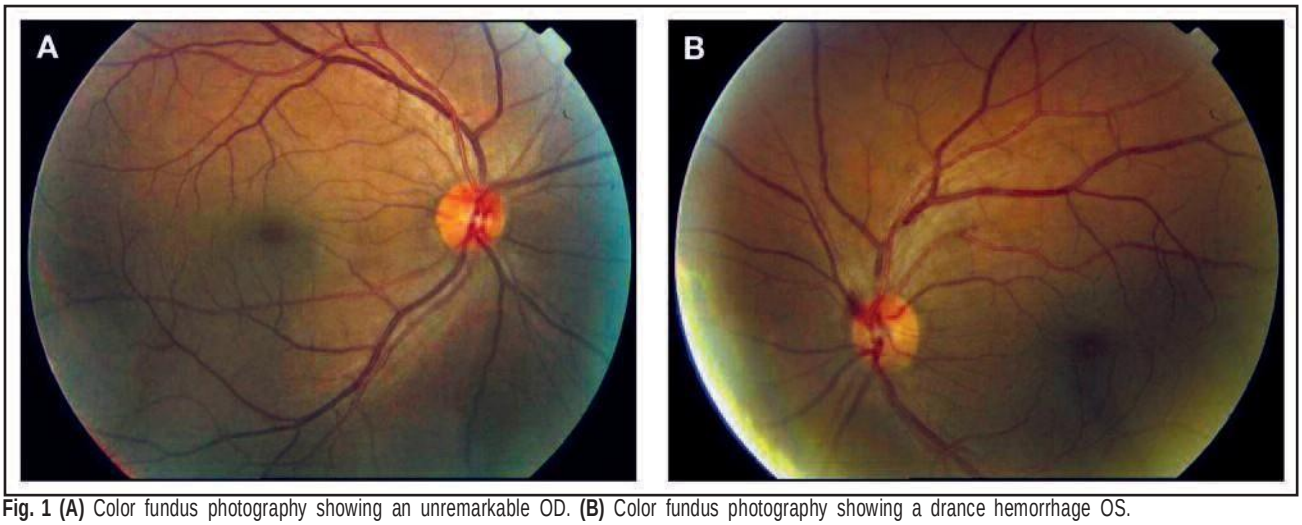

Fig. 1 (A) Color fundus photography showing an unremarkable OD. **(B)** Color fundus photography showing a drance hemorrhage OS.

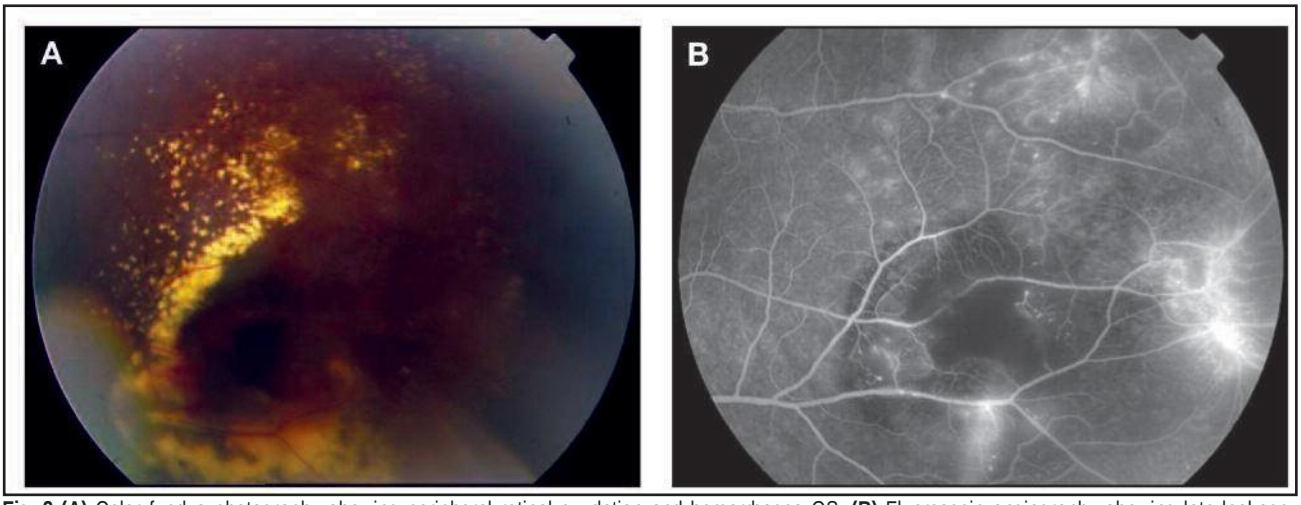

Fig. 2 (A) Color fundus photography showing peripheral retinal exudation and hemorrhages OS. **(B)** Fluorescein angiography showing late leakage oftelangiectasias OS.

Findings were confirmed on follow-up by fluorescein angiography showing focal areas of capillary dropout, telangectasias and capillary changes with mild hyperflourescence in the affected area suggestive of a slow mild leakage with no obvious neovascularization along with an area of hyperflourescence continuous with a macroaneurysm and an area of blocked fluorescence from the subretinal heme. (Fig. 2A,B) Optical coherence tomography (OCT) was not attempted given the lesion's peripheral location. A preliminary diagnosis of Coats' disease was made.

The patient was referred to his primary medical provider to rule out diabetes, thromboembolic disease, blood pressure control, renal function tests, carotid ultrasounds, echocardiogram, and electrocardiogram given the unlikely hemorrhage finding along with exudation. All exams returned normal making idiopathic Coats' disease the primary working diagnosis. A consultation with our retinal specialist confirmed the diagnosis, and recommended quadrantic panretinal photocoagulation (PRP) in the left eye. The patient underwent a single session, 714 spots of PRP OS in the temporal retina, with no complications. The patient did not return for the second session of PRP and presented again 3 years later with no evidence of exudation or hemorrhage. Vision remained stable at 6/6 (20/20) OD, 6/4.8 (20/16) OS, and he continues to be followed on an annual basis.

Case 2

A 47-year-old Latino male presented to clinic with a complaint of decreased vision at near through his current

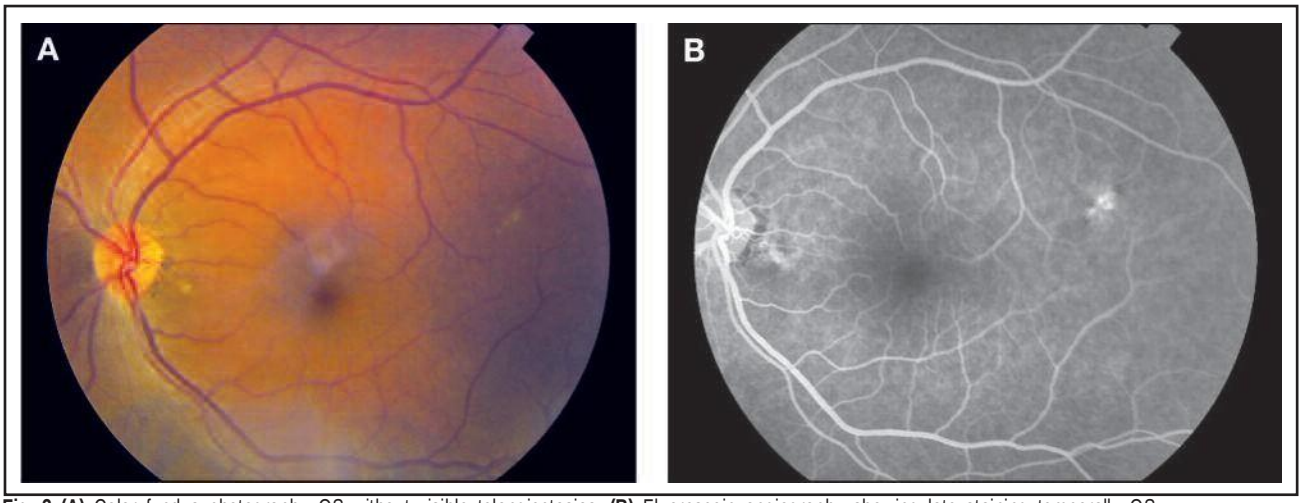

Fig. 3 (A) Color fundus photography OS without visible telangiectasias. **(B)** Fluorescein angiography showing late staining temporally OS.

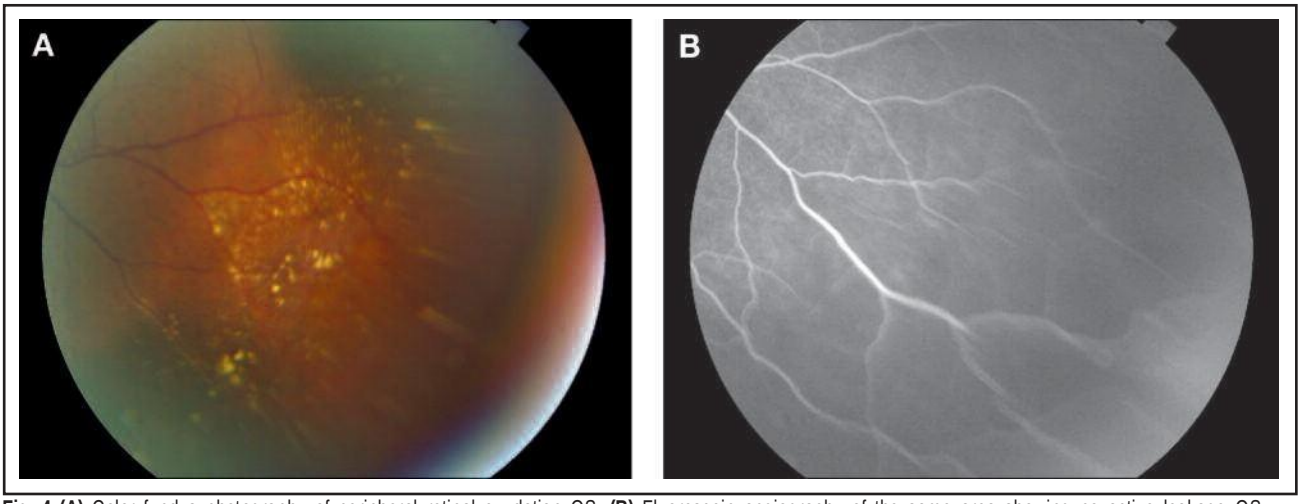

Fig. 4 (A) Color fundus photography of peripheral retinal exudation OS. **(B)** Fluorescein angiography of the same area showing no active leakage OS.

glasses. His previous medical history was significant for hypertension, hyperlipidemia, hepatitis B carrier, and positive PPD. The patient's best corrected visual acuity was 6/6 (20/20) in the right eye with a -0.25 DS correction and 6/6 (20/20) in the left eye with a plano -0.50 x 120 correction. Pupils were normal, extraocular muscles were full OU, and confrontation fields were full to finger count each eye. Anterior segment evaluation revealed pingueculae OU with all other findings within normal limits. Intraocular pressures were 14 mmHg OU with Goldmann applanation tonometry. Blood pressure was found to be 127/83 mmHg.

Dilated posterior segment evaluation revealed clear media, cup to disc ratios of 0.20 round OD and OS with disc margins distinct and rims tissue pink and healthy. Maculas were flat and avascular both eyes. The peripheral retinal exam was significant for possible peripheral vasculitis with exudates inferior and inferior temporally in the left eye without hemorrhages as well as temporally along superior arcades.

Findings were confirmed on follow-up with fluorescein angiography (Figs. 3A,B; 4A,B) exhibiting an area of late staining superiotemporally with a window defect and no vascular leakage or vasculitis. Recent blood work ruled out diabetes. A diagnosis of adult onset idiopathic Coats' disease OS was made with a recommendation for a wide field fluorescein angiography to be completed to further assess the peripheral aspects of the lesion to make appropriate recommendations for treatment. The patient did not return for the fluorescein angiography and has since been lost to follow-up.

Discussion

Coats' is a non-hereditary disease of mosaic phenotype[8,10] with no racial predilection.[6] It manifests predominantly

Table I Five stages of Coats' disease

Stage 1: Retinal telangiectasias
Stage 2: Telangiectasias and exudation
A: Extra foveal
B: Foveal
Stage 3: Exudative retinal detachment
A: Subtotal detachment
1. Extra foveal
2. Foveal
B: Total retinal detachment
Stage 4: Total retinal detachment and glaucoma
Stage 5: Advanced end stage disease

unilaterally in young males in the first decade of life,[6] although a small percentage of patients do not present with symptoms until adulthood.[6] A 2010 study by Morris et al found the estimated incidence to be at least 0.09 per 100,000.[13] According to some authors, the disease has an association with some systemic conditions such as Turners syndrome and familial renal retinal dystrophy,[3] while others argue there is no direct correlation to any systemic disease, stating that abnormal retinal telangiectasias with systemic associations do not technically qualify as Coats' disease which is strictly idiopathic.[3]

Pathogenesis of vessel leakage begins with weakened artery walls and fewer pericytes, leading to telangiectasias, microaneurysm, and sausaging of vessels.[3] Resulting poor blood flow can lead to macular ischemia and neovascularization.[3] Retinal and vitreous hemorrhages are typically uncommon in Coats' disease although they may be found at a higher rate in adult onset Coats'.[1,4] In adult onset Coats' vascular abnormalities generally occur in peripheral and equatorial regions consistent with both cases presented.[14]

George Coats' originally described variations of these retinal vascular anomalies as three different diseases but later went on to merge the first two as variations of the same disease[3] and the last was later defined as Von Hippel-Landau angiomas.[5] More recently, Shields et al classified the disease into five stages that are useful clinically for documentation (Table I).[4,6]

Differential diagnoses are many and include: retinopathy of prematurity,[4] familial exudative vitreoretinopathy and idiopathic juxtafoveal telangiectasias, as well as age-related macular degeneration, diabetes, hypertension, and hyperlipidemia.[4] As late stages of Coats' disease may sometimes mimic retinoblastoma, it is important to rule out this out as a diagnosis given its potential for morbidity.[6] Clinical diagnosis is based on location and appearance. A careful fundus exam with condensing lenses, funds photography, OCT, and fluorescein angiograph are often necessary to accurately classify a lesion with active leakage requiring treatment. Telangectasias show early hyperflourescence along with late leakage while exudates hypoflouresce.[1] Fluorescein angiography may also help identify areas of capillary non-perfusion and macular edema. An extra-foveal inactive lesion can be followed regularly without immediate treatment when indicated.

Complications resulting from Coats' disease include capillary non-perfusion, neovascular glaucoma, cystoid macular edema, and retinal detachment from exudative accumulation in the subretinal space. Therefore, individualized considerations to determine the treatment modality best fit for the patient are necessary.[3,4,15] Historically Coats' was treated with enucleation when presenting in children due to its similarity to retinoblastoma in appearance and to prevent progression into neovascular glaucoma.[5,6,16] In such cases computed tomography (CT) scan, magnetic resonance imaging (MRI), B scan ultrasonography to identify a mass or a fine needle biopsy confirming the absence of cancer cells should be performed to rule out a tumour and save the eye whenever possible.[5,16] The high levels of vascular endothelial growth factor in the eye associated with vascular permeability are decreased by laser photocoagulation; more recently, severe cases have been treated with cryopexy and panretinal photocoagulation.[17] In cases with resultant retinal detachment, scleral buckle is employed to prevent or repair the detachment.[16] Lastly, if salvage is not possible, enucleation of the eye remains an option. Prognostic visual outcome has been shown to depend on macular involvement[9] and level of vision at presentation, and not specifically on treatment modality.[16]

Most recently, in addition to surgical and laser treatments, abnormal telangiectasias can be treated with the help of anti-VEGF intravitreal injections if diagnosed early.[9] Medications such as bevacizumab,[18,19] (pegaptanib sodium)[8] and ranibizumab which bind and neutralize active forms of VEGF in the eye,[11] may reduce the angiogenic factors, resulting in stability or resolution of retinal exudation.[20] In addition, the corticosteroid triamcinolone acetate has been shown in studies to increase the reabsorbtion of subretinal fluid when used in conjunction with other treatment modalities.[20] Patients who exhibit limited responsiveness to laser treatment may be good candidates for these alternatives to reduce exudates, macular edema and sub-retinal fluid due to their effectiveness against microangiopathy.[20] As with any treatment option, complications are always a factor that must be taken into consideration. With the use of fluorescein angiography and more recently OCT, more detailed views of vessel leakage can be defined and localized, and are useful tools for follow-up and documentation of treatment outcomes.[9]

Conclusion

Coats' disease can commonly progress or reoccur; therefore, follow-up is necessary throughout life.[3,21]

Luckily for the patients with adult onset Coats' presented here, the disease appeared to be generally less aggressive with a limited area of involvement.[21] With ever-changing research and advancements in technology, we continue to gain better understanding of Coats' disease with new discoveries aiding in diagnosis and in treatment modalities that will provide the best quality of care for patients. 0

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