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Case report: Incidental finding of adult-onset of Coats' disease

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Abstract
We report a case of adult onset of Coats' disease encountered during presentation of fellow eye pathology. Patient was referred for right eye corneal foreign body. On examination, left eye vision was poorer than right eye and patient was not aware of the severity of his eye condition. Upon further fundus examination, it shows signs of coats disease in left eye. The diagnosis was then supported by optical coherence tomography (OCT) scan and fundus fluorescein angiography (FFA) study. In order to avoid dreaded complication of untreated Coats' disease, it is of utmost importance to achieve early diagnosis through clinical and radiological evaluation. The appropriate treatment for Coats' disease is based on the severity of disease. Laser or cryotherapy has been most effective treatment along with adjuvant therapy of antivascular endothelial growth factor (VEGF).

Keywords: Coats disease, adult-onset, laser therapy, anti-VEGF, fundus fluorescein angiography (FFA)

Introduction

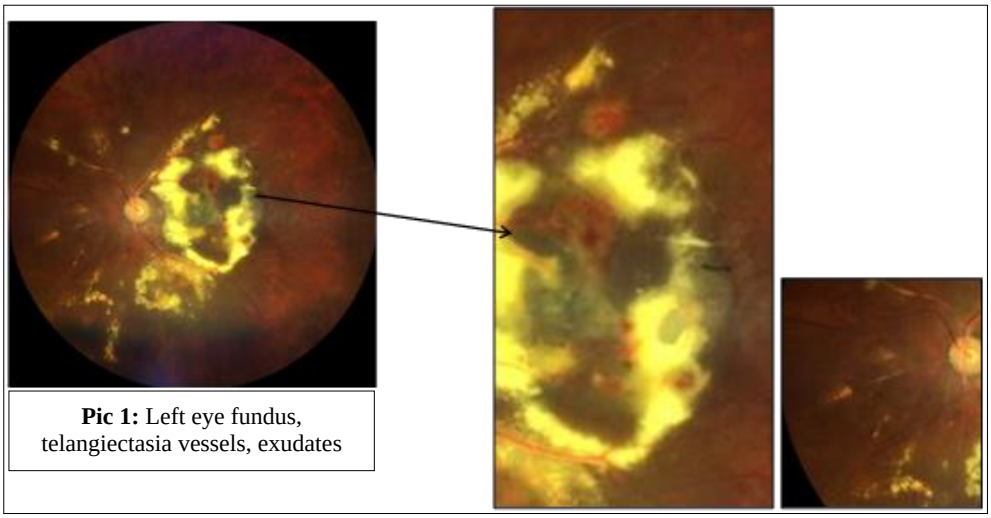
Coats disease is an idiopathic, non-hereditary, telangiectasic neovascular disease of the retina. There is typically no racial preference and the condition is 95% unilateral. It typically affects men and is ususally identified early, with an average diagnosis age of six years [2]. Although it has been seen in both infant and elderly people, it is more common in males than females with a ratio of 3:1. Coats disease is characterised by progressive growth of abnormal arteries, exudates, and hemorrhage in the affected person's retina. As the disease progresses, there is increased intraretinal and subretinal exudation, which may result in exudative retinal detachment. Patients may develop rubeosis and choroidal or retinal neovascularization, which can lead to hemorrhage, glaucoma, and blindness. Management varies from ablative therapy to surgery depends on the severity of the disease.

Case Presentation

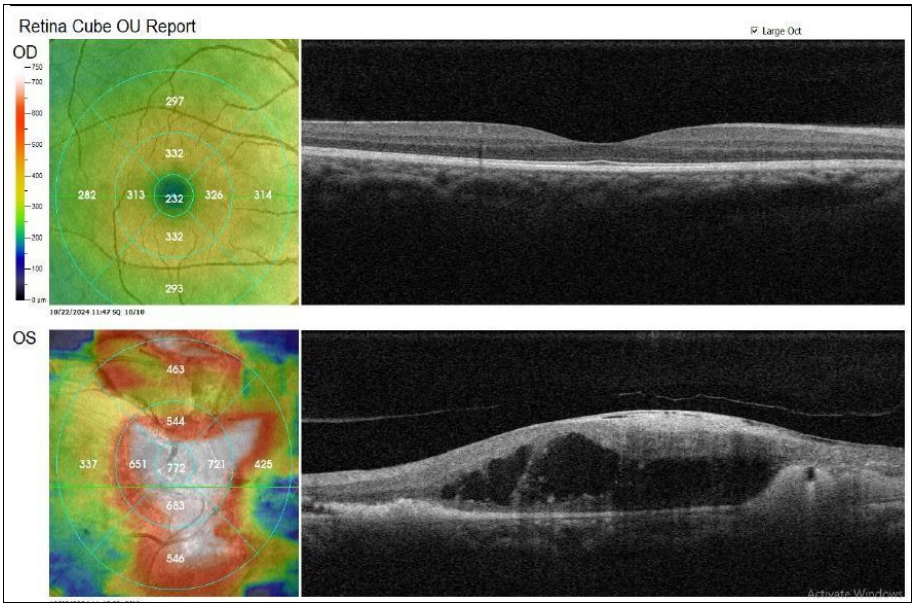
A 35-year-old Malay man with no known medical illness came to the clinic with the complaint of right eye metal dust insertion while grinding at workplace 4 days ago. He was complaining of right eye pain associated with redness, tearing and blurring of vision. Visual acuity on presentation was right eye 6/7.5 whereas left eye counting finger. Upon further questioning, patient found out of left eye blurring of vision for more than 1 year while rubbing the fellow eye after waking up from sleep. He did not seek any medical advice thinking it was normal. The patient had no previous history of systemic and ocular diseases. He denies any constitutional symptoms or other ocular symptoms like floaters, metamorphopsia and diplopia. No significant family history. He is a married man and blessed with 3 children. He was an ex-substance abuser, stopped four years ago. Previously, addicted to metamphetamine however denies sexual promiscuity.

Examination of anterior segment, presence of small corneal foreign body near limbus at 9'oclock position over right eye whereas left eye was unremarkable. No signs of anterior uveitis bilaterally. Fundus examination of right eye, optic disc was pink with 0.3 cup-disc ratio and well defined. Macula and vessels appears normal. Retina was ratio and well defined. There's presence of peripheral sclerosed vessels, subretinal exudate and multiple telangiectasia vessels (Pic 1).

Flat with no abnormality or lesion seen. Left eye optic disc was pink with0.3 cup-disc



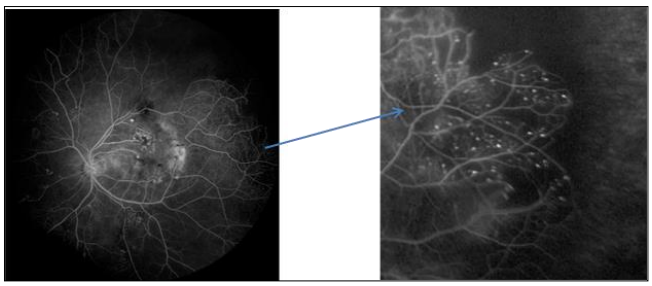
Pic 1: Left eye fundus, telangiectasia vessels, exudates



Pic 2: OCT macula, OD: Normal, OS: Subretina; intraretinal fluid, traction

Left eye optical coherence tomography (OCT) showed presence of traction, subretinal and intraretinal fluid with central foveal thickness of 772µm (Pic 2). Right eye was normal with central foveal thickness of 232µm (Pic 2). Fundus fluorescein angiography (FFA) examination of left eye revealed multiple areas of telangiectasias at posterior pole, inferotemporal and superotemporal with macular oedema and vitreomacular traction. Extensive areas of capillary non-perfusion except superonasal were also seen (Pic 3). Right eye FFA was unremarkable. Blood investigation showed no signs of infection.

The left eyen was treated with panretinal photocoagulation laser at avascular region and focal laser at telangiectasia region. Intravitreal anti-vascular endothelial growth factor (VEGF) injection was not given due to presence of vitreomacular traction. In order to avoid dreaded complication of untreated Coats' disease, it is of utmost importance to achieve early diagnosis through clinical and radiological evaluation and appropriate treatment given. Patient's right eye corneal foreign body (metal dust) was removed and treated with topical antibiotic. The visual acuity of right eye was 6/6 with no complaint afterwards.



Pic 3: Left eye FFA, area of non-perfusion, telengiectasias

Discussion

Coats disease is a telangiectatic neovascular disease of the retina that is idiopathic and frequently affects unilateral eyes of young males. However, mutations in retinal proteins encoded by Crumbs cell polarity complex component (CRB1) and Norrie disease pseudoglioma (NDP) genes is believed to be the possible pathogenesis. (3) Lebers miliary aneurysm is considered to be a milder variant of the Coats disease. it does not usually involve significant subretinal exudation, hemorrhage, or exudative retinal detachment. Type 1 idiopathic macular telegiectasia is now considered the same as Coats' disease [3].

Histopathological processes of Coats disease include the collapse of the blood-retina barrier at the vascular endothelium, as well as the loss of pericytes and endothelial cells. Leakage into the vessel wall is followed by thickening, saccular and fusiform aneurysms and vessel obliteration. Exudates in the intra- and subretinal space contain blood, lipids, cholesterol crystals, lymphocytes, and fibrin. Coats disease is characterized by high levels of vascular endothelial growth factor (VEGF), which is essential for the establishment of normal retinal capillaries and vascular permeability.

In adults, the most common major complaint is painless vision loss, although children usually present with more diverse symptoms, including as impaired vision, nystagmus, strabismus, and leukocoria. Although the illness progresses, it can be asymptomatic until it is detected using ophthalmoscopy. It has been noted that the younger the age at presentation, the more severe the disorder and the lower the visual prognosis. Patients who present later in life may have been asymptomatic in their childhood.

Shields *et al*. staging system to describe the severity of Coats disease based on the telangiectasias appearance, exudation leading to retinal detachment, and secondary complications [1, 2]. These stages help guide the clinician in determining the best management option and assessing the prognosis.

Stage 1: Retinal telangiectasias only
Stage 2a: Telangiectasias and extrafoveal exudation
Stage 2b: Telangiectasias and foveal exudation
Stage 3a: Subtotal exudative retinal detachment
Stage 3b: Total exudative retinal detachment
Stage 4: Total detachment with secondary glaucoma
Stage 5: Advanced end-stage disease

Treatment option is decided based on the severity of the disease. Mild cases can be observed, but if exudation, or

retinal or subretinal fluid, is present, laser ablation or crotherapy is the mainstay of treatment especially in areas of telangiectasia and nonperfusion [1, 2].

Laser: Thermal laser photocoagulation has s disease is to obliterate abnormal vasculature and eliminate hyperpermeability of aneurysms. Even in the presence of subretinal fluid, vascular anomalies can be directly photocoagulated using a laser.

Cryotherapy: Indicated if peripheral retinal telangiectasia with significant exudative retinal detachment impedes adequate laser photocoagulation.

Surgery: In the case of total exudative retinal detachment, persistent vitreous hemorrhage, and/or secondary complications are present, surgery may be needed for retinal reattachment or enucleation.

Adjuvant therapy: Anti- VEGF agents have been found to be effective in reducing subretinal fluid and exudation. However, vitreoretinal fibrosis and associated tractional retinal detachment may develop following anti- VEGF treatment.

In this case, patient was treated with panretinal photocoagulation laser at avascular region and focal laser at telangiectasia region. Intravitreal anti-vascular endothelial growth factor (VEGF) injection was not given due to presence of vitreomacular traction.

Conclusion

Coats disease is an idiopathic, progressive disease that predominantly affects male children, but may also present later in adolescents and adults. Adult Coats disease is an uncommon cause of vision loss beginning in childhood. It is usually gradually progressive disease**.** The clinical, radiological and angiographic findings are vital in diagnosing Coats disease and retinal telangiectasia. Treatment by laser photocoagulation in area of leakages may be beneficial in preventing visual loss even best corrected vision is not achieved.

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