ORIGINAL RESEARCH PAPER

Ophthalmology

KEY WORDS: Adult onset coats disease, exudative retinal detachment, telangiectatic, methyl prednisolone,

COATS' DISEASE OF ADULT ONSET-A RARE ENTITY

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ABSTRACT

Typically Coats disease appears in childhood and rarely has been diagnosed for first time in adults.This article describes patient affected by Coats disease diagnosed in fifth decade of life.A 46 year old male presented to Ophthalmic OPD with painless diminution of vision in left eye since 6 days.On ophthalmological examination,best corrected visual acuity in left eye was 6/60 and 6/6 in right eye.Right eye fundus was normal.Left eye fundus exhibited massive subretinal exudates with exudative retinal detachment in superonasal quadrant and hard exudates along inferotemporal arcade and telengiectatic vessels.Fundal fluorescein angiogram in left eye revealed early hyperfluorescence of telangiectatic vessels and hypofluorescence consistent with sub retinal exudates.Optical coherence tomography showed increased macular thickness.He was diagnosed to have adult Coats' disease.Patient was treated with pulse therapy methyl prednisolone,posterior subtenon and subconjunctival steroid.Two weeks post laser,vision improved to 6/18 in left eye, reduction of the hard exudates and resolution of angiomatous changes

INTRODUCTION-

Coats' disease is an idiopathic disease characterized by retinal telangiectasia, intraretinal, subretinal exudation and exudative retinal detachment.It was first described in 1908 by George Coats, a Scottish medical student,as a unilateral condition with retinal exudation and telangiectasia in male children.Four years later, Leber defined a similar condition albeit more severe, in which there were multiple retinal aneurysms,as well as retinal detachment rather than telangiectasias[1].For the first half of 20th century, these clinical entities were considered as separate conditions. Subsequently,Shields et al.defined Coats' disease as 'Idiopathic retinal teleangiectasia associated with intraretinal exudation and frequent exudative retinal detachment without signs of appreciable retinal or vitreal traction'.It is often described as 'light bulb telangiectasia' due to large amounts of yellow exudates accompanying the condition[2].In addition, there may be capillary nonperfusion, aneurysmal formation, and massive lipid deposition[3].Degeneration of the endothelial and mural cells of the retinal arteries and veins has been suggested to be the primary pathologic defect of this disease [4]

Coats' disease is mostly unilateral, progressive condition affecting mainly males during childhood,with average age of diagnosis being between 8 and 16 years.It typically presents in early childhood with vision loss,strabismus or leukocoria, and must often be differentiated from retinoblastoma.Coats' disease if untreated, can lead to total retinal detachment and secondary glaucoma, sometimes requiring enucleation [5].Several studies have revealed that the earlier the age of presentation,more severe the disease progression and the greater the likelihood of enucleation[6]. Specifically,it is hypothesized that Coats' disease could be a consequence of a mutation in the NDP gene,which results in a deficiency of norrin,a protein thought to be important for normal retinal vasculogenesis [7].Of note, Black et al [8],reported a case of a mother with unilateral Coats' disease, who gave birth to a son with Norrie disease, in which both had mutations in the NDP gene.

Coats' retinopathy only on rare occasions, has been diagnosed for the first time in adults[9]. This article describes a patient affected by Coats disease diagnosed in the fifth decade of life. Systemic conditions such as hypertension, diabetes mellitus, and hypercholesterolemia have been reported with adult onset Coats' disease, whereas no such association has been reported with the childhood onset disease.Coats' disease diagnosed in adulthood is an idiopathic,retinal exudative vascular disease without an inciting factor and has retinal features different from the childhood disease [10]. Although various methods have been employed to treat Coats' disease, including diathermy, retinal photocoagulation, cryotherapy, and vitreous surgery to elucidate the abnormal vessels, thereby minimizing exudation, the prognosis of visual function remains unsatisfactory.

CASE REPORT

A 46 year old male patient presented to Ophthalmic OPD with painless progressive diminution of vision in his left eye since 6 days. On ophthalmological examination, best corrected visual acuity(BCVA) in left eye was 6/60 and in right eye was 6/6.Goldman applanation tonometry readings in both eyes were 18 mm Hg. Reaction to light was normal in both eyes and there was no relative afferent papillary defect.Anterior segment examination was normal in both eyes.The right eye fundus was also normal. The left eye fundus exhibited around 3-4 disc diameter size massive subretinal exudates with exudative retinal detachment in superonasal quadrant 2 disc diameters away from disc and hard exudates along inferotemporal arcade and telengiectatic vessels.(Figure-1)

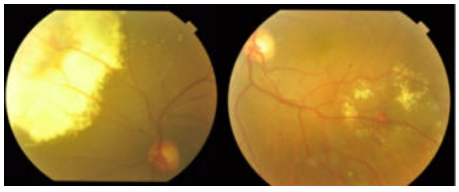

FIGURE-1

Fundal fluorescein angiogram (FFA) revealed early hyper fluorescence of telangiectatic, leaking vessels in the superonasal and inferotemporal regions and hypofluorescence consistent with sub retinal exudates in left eye(Figure-2)

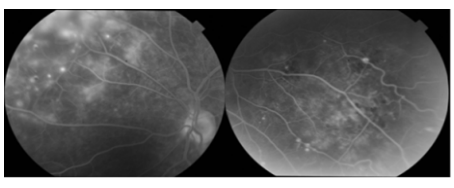

FIGURE-2

Macula looked edematous on slit lamp biomicroscopy with 90D lens. Optical coherence tomography (OCT) showed increased macular thickness in left eye(465µm)(Figure 3).

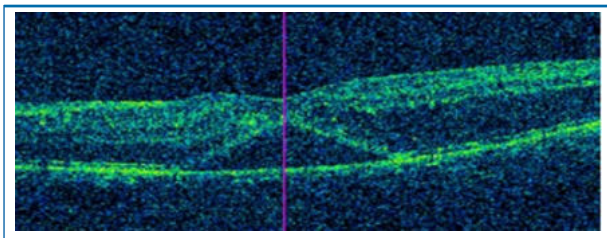

FIGURE-3

Based on clinical and fundus angiogram findings' he was diagnosed to have idiopathic adult retinal telangiectasia or adult Coats' disease.Patient was treated with pulse therapy of methyl prednisolone, posterior subtenon (0.5 ml of 40mg/ml triamcinolone acetonide) and subconjunctival injection of steroid.The telangiectatic vessels in the inferior temporal and superior nasal retina in left eye were treated with direct argon laser photocoagulation and scatter retinal photocoagulation was applied to rest of the superior retina.

Two weeks post laser, his best corrected visual acuity improved to 6/18 in left eye. There were small reduction of hard exudates, resolution of macular edema and angiomatous changes(Figure 4)

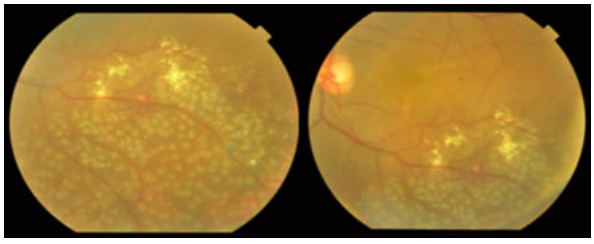

FIGURE-4

DISCUSSION-

Coats diseases is an idiopathic, unilateral exudative retinal vasculopathy characterized by telangiectasia, aneurysms and irregular dilatations of retinal vessels which lead to intra and sub-retinal exudation and generalized lipid deposits.Adult onset form is manifested by many findings typical of coats disease which include unilateral nature of the disease, male predominance, vascular telangiectasia, lipid exudation, macular edema, and areas of capillary non perfusion with in contrast to childhood onset, many adult patients have no symptoms or exhibit good visual acuity.In these cases, vascular anomalies generally appear in the equatorial and peripheral retina and lipid exudation (which appears in a massive but diffuse manner in children) is more localized[9]. These patients usually maintain good visual acuity and disease evolves at a slower pace. Also in contrast to childhood onset, Coats disease in adults is frequently associated to retinal hemorrhages caused by bleeding through retinal aneurysms. The diagnosis of Coats disease is established mainly due to the ophthalmoscopic appearance of the retina.Fluorescein angiography shows early hyper-fluorescence of telangiectasiae, hypo-fluorescence in exudates and slight late hyper-fluorescence of the sub-retinal liquid. It also frequently shows areas of non-capillary perfusion or macular edema. Idiopathic adult retinal telangiectasia with exudation is spectrum of disease which is synonymous with Coats' disease. The main entity for establishing the differential diagnosis for Coats disease in children is retinoblastoma.In adult patients it must be differentiated from Leber's miliary aneurysms, with the main differential characteristic being a small amount of retinal exudation exhibited by the latter process. In addition although with less frequency it can be confused with retinal detachment, toxocariasis, choroidal melanoma, retinitis due to cytomegalovirus or toxoplasmosis[1] . In what concerns treatment, the main goal in this disease is to obliterate the telangiectasia to facilitate the reabsorption of exudates and maintain as much visual acuity as possible13. Possible therapeutic options described in literature include observation, laser photocoagulation, cryotherapy, surgical treatment of exudative retinal detachment and enucleation. Even though our patient had good visual acuity, it was considered that the risk of progression of exudation towards the macular area was high and therefore it was decided to establish preventive treatment with laser.Fluorescein angiography can help in defining the structural and permeability alteration in the affected vessels and demonstrate the extent of the extravasular leakage of serous exudates into and beneath the retina. In Coats' disease, the fundus angiography showed eharacteristic premature hyperfluorescence of telangiectasias and presence of microaneurysms with leakage at late phase12. In previous angiographic studies of less advanced Coats' disease demonstrated that the arterial system seemed to be more damaged than the venous side. Most of the affected arteries ended in macroaneurysm-like dilatations surrounded by avascular areas or complete vascular closure in more advanced cases. In this patient, early phase of angiography showed dilatation of capillaries with hyperfluorescence localized in the area of exudates. There was leakage of fluorescein from the telangiectatic vessels noted during the late phase.

Laser photocoagulation may be taken into consideration at the beginning if telangiectasias are located in the periphery and presence of large exudation. Cryotherapy is generally reserved for more advanced cases and those patients with lesions confined to the far periphery. The purpose of these treatment modalities is to 'minimize vascular leakage and promote reabsorption of exudates. If the exudation is limited to a single quadrant or is located in the nasal sector, visual prognosis is more favourable .

CONCLUSION

Even though the majority of Coats disease cases occur in childhood, we must consider its diagnosis in adult patients with retinal vasculopathies with major exudative components. Even when in adults Coats disease courses in a more benign manner and comprises a good prognosis in what concerns visual acuity, we must assess establishing treatment when there is a risk of progression of the exudates towards the macular area. Adult Coats' disease is an uncommon cause of vision loss. It usually affects the visual acuity gradually and later may progress and become symptomatic later in life as a result of hemorrhage, edema or lipid exudation. The clinical and angiographic findings are important in diagnosing the retinal telangiectasias. Treatment by laser photocoagulation into areas of leakage may be beneficial in preventing visual loss

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