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Research Article

COATS' DISEASE

Bharati Gogoi and Madhura M

Regional institute of Ophthalmology ,GMCH, Guwahati, Assam

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A B S T R A C T

Coats' disease is an idiopathic condition due to abnormal telangiectatic or aneurysmal vessels associated with intraretinal and subretinal exudation. The majority of cases can be diagnosed by age 20 with a peak incidence at the end of the first decade. Males are four times more commonly affected than females. In this review we highlight the pathogenesis, diagnosis and treatment of the disease.

INTRODUCTION

Coats' disease is an idiopathic condition due to abnormal telangiectatic or aneurysmal vessels associated with intraretinal and subretinal exudation. Subsequently, similar ocular manifestations were described in several other diseases including Leber'smiliary aneurysm. With few exceptions, most authorities now believe that Leber's disease is an early or non progressive form of Coats' disease.[1-3]

Classification[4]

George Coats1 first described this disease in 1908, and classified it into three groups.

Group I: includes eyes with massive subretinal exudates and choroidal mononuclear infiltrate but no vascular abnormalities.

Group II: includes eyes with both massive subretinal exudates and multiple retinal vascular abnormalities with intraretinal hemorrhage but no choroidalinflammatory infiltrate.

Group III: eyes have massive subretinal exudate and definite retinal arteriovenous malformations

Pathophysiology[4]

This exudative vasculopathy is neither inherited nor associated with systemic vascularabnormalities. But the growing list of genetic diseases such as retinitis pigmentosa associated with Coats' disease supports the implication that Coats' disease may be a genetic abnormality.[5-7] A few researchers believe that Coats' disease may have a primary vascular etiology. A defect in cholesterol transport has been proposed. The association between hypercholesterolemia and the adult form of Coats' disease is suggestive of a serum lipid abnormality but an association does not appear to occur in the juvenile form[8]

Epidemiology

It is predominately a disease of childhood though it can affect adults. It is unilateral in 90% or more of the cases.[9] The majority of cases can be diagnosed by age 20with a peak incidence at the end of the first decade.[9] Males are affected four times as frequently as females.[9]

Clinical Features

Predominantly a unilateral retinal developmentalvasculopathy although 10-15 percent of cases are bilateral.[10,11]The disease is seen in two forms, the adolescent form (< 20 years of age) and a less common adult form (>20 years of age).[12]

In children due to failure of recognition of poorvision, the initial presentation may be leukocoria or strabismus, commonly esotropia.[4]In adults, the most common presentation is reduced visual acuity.[4]

Anterior segment examination is usuallynormal except in more advanced cases with rubeosis iridis, angle closure, neovascular glaucoma and complicated cataract.[4]

Fundus examination reveals typical vascular abnormalities in the form of 'light bulb' or 'grape-like' aneurysmal dilatations, looping, kinking and extensivesubretinal exudates. Theextent of the retinal

Corresponding author:* **Bharati Gogoi
Regional institute of Ophthalmology ,GMCH, Guwahati, Assam

involvement is variable. The supero-temporal retina is more commonly involved followed by the macular and paramacular area.

The aggressive form of the disease is seen in children and infants.[13] Occasionally acute orbital cellulitis may occur secondary to trans scleral movement of toxic products.[14]

Clinical Course

The clinical course is variable, although most cases are progressive, if left untreated. Spontaneous resolution is an exception.[15] Acute exacerbations of the disease are usually separated by periods of disease inactivity. The end stage of the exudative process in eyes with severe long-standing disease includes total retinal detachment, intraretinalhemorrhagic retinalmacro-cysts,[16] vitreous hemorrhage, complicated cataract, iridocyclitis, rubeosis iridis, secondary neovascular glaucoma and even phthisis bulbi[17]

Diagnosis

The diagnosis is confirmed ophthalmoscopically when the typical vascular abnormalities are seen in association with lipid deposition and subretinalexudate.[4]

Investigations

Fluorescein angiography

It is useful to delineate the nature, extent and severity of vascular abnormalities. In retinoblastoma the dilated vessels are continuous with large vascular trunks extending into the tumor mass, whereas the telangiectatic vessels in Coats' disease do not extend into the subretinal exudative mass. The characteristic angiographic features of Coats'disease are temporal quadrant capillary non-perfusion with leaking microaneurysms.

Ultrasonography and computerized tomography (CT)[18] can demonstrate calcium in retinoblastoma

Magnetic resonance imaging (MRI)[19,20] is useful to differentiate retinoblastoma from Coats' disease, but is less superiorto CT and ultrasonography.

High-resolution Doppler ultrasound[21] has been used in some instances to demonstrate real-time imaging and to delineate specific structural abnormalities which are not detectable by CT or MRI.

Cytology and biochemical analysis of the subretinalfluid can provide accurate information in the diagnosis of Coats' disease. The subretinal fluid contains cholesterol crystal and pigment-laden macrophages in the absence of tumor cells. However it is of little help in a clinical noninvasive setting.[18,22]

Differential Diagnosis[4]

Any condition that produces leukocoria and strabismus, which includes retinoblastoma, parsplanitisleading to proliferative retinopathy, Norrie's disease, congenital cataract, persistent hyperplasticprimary vitreous, familial exudative vitreo-retinopathy, retinopathy of prematurity, toxocara endophthalmitis and retinitis pigmentosa with Coats' response.

Management

The goal of treatment is to obliterate the telangiectaticvessels and to allow reabsorption of exudation to preserve anatomical integrity of the eyeball and to preserve or improve visual function. The treatment modality depends on the location and severity of the lesion and age of the patient. If the lesions are limited in extent and do not threaten macular vision, patients can be followed safely without any treatment. However, if exudation is extensive and progressive, threatening central vision and if there is significant peripheral retinal detachment, treatment is indicated.

Laser photocoagulation has become the treatment of choice for Coats' disease. Development of fluoresce in angiography led to the realization that many treatment failures were results of inability to detect abnormal vessels on routine clinical examination.[23,24]

Focal laser photocoagulation is performed directly to leaking telangiectatic vessels. Usually a larger spot size (300-500 micron) is used to achieve moderate intensity burns.[22] Indirect ophthalmoscopy mountedlaser86,87 is used to treat more peripheral or anterior retinal lesions.[25,26]

When the disease involves the peripheral retina, is complicated by exudative retinal detachment or where due to massive exudation and thickening of the retina, it is difficult or impossible to produce a laser reaction, cryotherapy is recommended. After treatment, exudates begin to be reabsorbed within 6 weeks, if the abnormal vasculature has been successfully eliminated.
Laser and Cryotherapy can lead to severe inflammation, macular pucker, vitreous hemorrhage, tractional or rhegmatogenous retinal detachment.

In cases of bullous retinal detachments when thecryo treatment of vascular anomalies is not possible drainage of subretinal fluid followed by cryotherapy and scleral buckling was shown to be effective.

The possible surgical complications in treating Coats' disease include vitreous and choroidal hemorrhage, inflammation and exacerbation of exudation, fibrosis with progressive retinal detachment, cataract formation, endophthalmitis, neovascular glaucoma and phthisis bulbi[25]

CONCLUSION

As the disease is often not diagnosed and treated until significant macular exudation occurs, central vision is frequently poor. Even after resolution of exudates with successful treatment, significant sub retinal fibrosis persists. Good visual acuity can be maintained in patients with mild vascular anomalies, those who do not require treatment, or those diagnosed and treated before macula is involved.[27] Amblyopia therapy may be helpful in improving vision and should be considered in young patients after successful treatment and resolution of exudates.

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