

Coats' disease

Retinal telangiectasia

DESCRIPTION

Coats' disease is a developmental, non-inherited, progressive and almost invariably unilateral retinal vascular disease. There is a variable pattern of retinal telangiectasia, with focal dilatation of retinal vessels that are incompetent and leak fluid. Exudative retinal detachment is the most significant among multiple possible complications of this condition.

Central vision may also be impaired when retinal edema and exudates affect the macula. Coats' disease is one of the causes of leukocoria (a white pupillary reflex) in children, of which retinoblastoma is the most important diagnosis requiring exclusion.

Milder forms of the retinal telangiectasias include idiopathic juxtafoveal retinal telangiectasia and Leber military aneurysms.

SYMPTOMS

The patient or family may notice reduced vision in one eye, strabismus or leukocoria.

SIGNS

Signs are highly variable. Involvement may be focal or diffuse, and is often more pronounced in the temporal fundus. Affected areas contain irregularly dilated and tortuous vessels (telangiectasia), often with saccular, 'light-bulb' venous outpouchings (aneurysms).

These abnormal vessels leak fluid, the serous component of which is preferentially reabsorbed.

As a result, areas of yellow, lipid-rich exudate progressively accumulate beneath and within the areas of edematous retina, usually over a period of several years. Exudates also tend to accumulate at the macula, possibly because of the increased metabolism and activity of the pigment epithelial pump in this area.

Continued leakage often leads to exudative retinal detachment and the formation of a pale mass, causing leukocoria. Rarely, proliferation of the retinal pigment epithelium creates a darker lesion which may resemble melanoma.

This mass may displace the lens and ciliary body anteriorly, causing secondary glaucoma.

Visual acuity in the affected eye is severely reduced in these advanced cases. Other complications of advanced disease include retinal or vitreous hemorrhage, ischemia and neovascularisation.

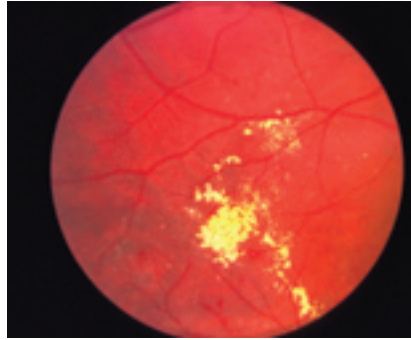


FIGURE 1. 'Lightbulb' venous outpouching and yellow lipid exudate

PREVALENCE

Rare. More common in males. Most cases are diagnosed before age 20.

SIGNIFICANCE

Early diagnosis and treatment of peripheral lesions may prevent extension and damage to the macula. Retinoblastoma must be excluded with caution, particularly in young children.

DIFFERENTIAL DIAGNOSIS

Retinoblastoma, persistent hyperplastic primary vitreous, choroidal hemangioma, branch retinal vein occlusion, toxocariasis.

SEE ALSO

Retinal Detachment – Classification, Retinitis Pigmentosa.

MANAGEMENT

Ocular tests, imaging investigations

Fluorescein angiography demonstrates coarsening of the capillary network with distal capillary closure, beading of venules and distal capillary non-perfusion.

Laser

When retinal oedema or exudates are detected, consideration is given to treatment. Laser photocoagulation of ischaemic retina is the most common modality; the objective is to promote reabsorption of exudates. As an example, an ideal candidate for laser photoco-

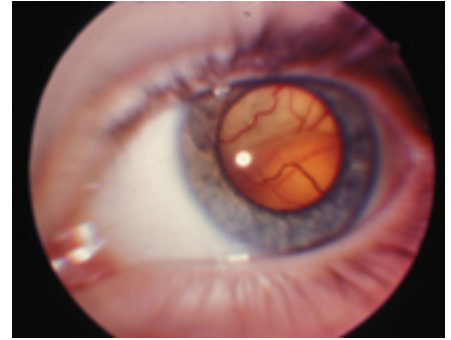


FIGURE 2. Leukocoria in a child with Coats' disease

agulation is a child with reduced central vision in association with focal vascular abnormalities not involving the macula. Laser treatment may retard or prevent progression, and occasionally improves vision. Multiple treatment sessions are often required. Photocoagulation is less effective once retinal detachment has occurred.

Surgery

Other treatment options include cryotherapy, drainage of subretinal exudates, and scleral buckling surgery.

Review

Review at three-month intervals is recommended for eyes with mild vascular dilatation only. Six-monthly review is appropriate once stability has been demonstrated.

The full series of these articles will be available in the book *Posterior Eye Disease and Glaucoma A-Z* by Bruce AS, O'Day J, McKay D and Swann P. £39.99 For further information click on the Bookstore at www.opticianonline.net.

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