Coats' Disease: Symptoms, Diagnosis, and Treatment

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Coats' Disease is a rare eye disorder described as an idiopathic retinal vascular disorder. It affects the retina by causing an abnormal growth of blood vessels, which, if left untreated, could lead to partial visual impairment (Shields et al., 2019). It was first identified in 1908 by George Coats. He observed that the primary signs of the disease were retinal exudation and telangiectasia. Many studies have termed Coats' Disease sporadic since it had no linked systemic abnormalities. It is mainly prevalent in young males, and symptoms typically present before age 15. In most cases (95%), it presents unilaterally (affects only one eye) (Helmi et al., 2022). The pathophysiology of Coats' Disease happens in phases. First, the blood-retina barrier collapses, leading to a leakage of blood into the retina. There is also a loss of pericytes and endothelial cells. Next, there is leakage into the vessel wall. This leakage causes vessel wall thickening, aneurysms, and vessel obliteration. The damaged vessels may eventually occlude and cause ischemia and neovascularisation in a few cases (Helmi et al., 2022).

Patients with Coats Diseases often present with various symptoms. These symptoms include vision loss, strabismus (crossed eyes), xanthocoria (yellow-orange pupillary reflex), nystagmus (spontaneous involuntary eye movements), or pain (Helmi et al., 2022). The primary symptom of the disease is decreased vision (visual impairment), which occurs progressively. The extent of this impairment is often dependent on the extent of retinal detachment. As mentioned, Coats' Disease is often unilateral. In many cases, this means that there is unequal vision between the two eyes. This usually results in strabismus, where the eyes appear crossed. In advanced stages, patients with Coats Disease experience pain in the eyes and redness of the pupil owing to redness due to secondary complications. Another symptom of Coats' Disease is leukocoria,

where light reflects off the abnormal blood vessels in the retina, giving the pupil a whitish appearance.

Coats' Disease is very difficult to diagnose accurately. More than 50% of cases are often misdiagnosed owing to their close presentation as retinoblastoma (Oli et al., 2021). Other differential diagnoses usually include Familial Exudative Vitreoretinopathy (FEVR), retinal detachment, Noorie disease, and persistent hyperplastic primary vitreous (PHPV) (Shields et al., 2019). To accurately diagnose Coat's Disease, some useful diagnostic tools include Fundus Fluorescein Angiography (FFA), Optical Coherence Tomography (OCT), Ocular Ultrasonography (USG), Computed Tomography (CT), and Magnetic Resonance Imaging (MRI). FFA helps pinpoint the abnormal or leaking vessels that indicate the diagnosis of Coats Disease. OCT is used to assess the extent of the damage to the retina and the accumulation of fluids, which is characteristic of Coats' Disease. MRI helps to rule out differential diagnoses like retinoblastoma in advanced stages of Coats' Disease. Once the disease has been definitively diagnosed, ophthalmologists can consider several treatment options.

Photocoagulation is one treatment option that can be used to manage Coats' Disease. It involves using laser therapy to repair leaking blood vessels in the retina. The resulting reduced blood in the retina will help preserve vision in the eye. Cryotherapy can also be used to manage the condition. This treatment option involves freezing the leaking vessels in the retina to prevent further leakage. Similarly, the reduced leakage preserves vision in the eye. Coats' Disease can also be treated using pharmaceuticals. For instance, intravitreal injections of drugs like bevacizumab (an anti-VEGF drug) can help reduce the frequency of damaged vessels and reduce leakage (Oli et al., 2021). Finally, in advanced cases where there is, as is in many cases, severe retinal detachment, surgery on the eye could be an option to remove vitreous gel.

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