EAELES’ Disease – A Distressing Mystery

Eales’ disease is an idiopathic; occlusive periphlebitis affecting the peripheral retina of young healthy males. Retinal changes are characterized by perivasculitis, peripheral ischemia and neovascularization leading to recurrent vitreous hemorrhage and other sequelae.

In 1880, Henry Eales – a British Ophthalmologist first observed the disease entity characterized by idiopathic vitreous hemorrhage in young males. He observed seven males of ages 14 – 29 years, all of them having history of headache, epistaxis and constipation. The disease was considered to be vasomotor neuritis till five years later Wardsworth explained its association with retinal inflammation. Though cases of Eales’ disease have been reported in Europe and North America but for unknown reasons it is rare in the developed world and more commonly reported in the Indian Subcontinent. Its incidence in India is 1 in every 200 – 250 ophthalmic patients. Eales’ disease targets healthy young adults, mostly males (Male predominance is up to 97.6%) with an age group ranging from 20 – 30 years. The disease is bilateral 50–90% of the time and thus causes significant socioeconomic burden.

The etiology is still obscure. Eales’ disease is considered to be an immunological response to some exogenous exposure. Favourable response to systemic steroids, histopathological evidence of inflammatory cells in vitreous and epiretinal membrane (ERM) and altered levels of immune markers point towards immunological mechanism behind it. Serum electrophoresis of patients with Eales disease was conducted at Armed Forces Institute of Ophthalmology (AFIO), Rawalpindi and 30% of them were found to have raised seric proteins and gamma globulins, 85% had raised ESR pointing towards possible role of immune process. Hypoxia induces increased expression of vascular endothelial growth factor (VEGF), which provides vasoproliferative stimulus. Oxidative stress with oxygen and lipid free radicals are also said to cause retinal inflammation. Mycobacterium Tuberculosis DNA has been isolated from pathological samples of patients of Eales’ disease. However, it was observed in India that only 1.3% of TB patients (active or healed) had Eales’ disease. Hypersensitivity to tuberculoprotein has long been associated with the etiology of Eales’ disease. Positive Mantoux was observed in as high as 90% of Eales’ patients in some case series. On the contrary, it has also been reported in Mantoux negative patients. HLA phenotyping was also studied at AFIO, which has suggested genetic basis of its etiology since HLA DR – 3, AI, B8. B5 and DR – 15 (2) were found to be raised in Eales’ patients. Eales’ disease is also known to have associations with haematological and neurological diseases.

Retinal changes in Eales’ disease patients may show retinal periphlebitis, later arteries may also be involved. Obliteration of inflamed vessel may lead to ischemia and neovascularization, which is observed in 80% of Eales’ patients. Neovascularization elsewhere (NVE) is commoner than neovascularization disc (NVD). Capillary drop out of 20 area and 60 disc area cause NVE and NVD respectively. New vessels may bleed to cause recurrent vitreous hemorrhage. Fibrovascular proliferation, retinal detachment (RD), uveitis, CMO, secondary BRVO, optic atrophy, NVI and rubeotic glaucoma are other sequelae. Macula is generally involved in later stages except in Central Eales’ disease.

Treatment is purely symptomatic and is stage dependent. Recurrent vitreous hemorrhage is the hallmark of this disease. Stage of inflammation is amenable to oral corticosteroid therapy. Ischemia and neovascularization are treated by photocoagulation and observation. Vitreous hemorrhage requires observation and then photocoagulation with vitrectomy. Complications require sophisticated procedures. Empirical anti tuberculosis treatment has been tried for severe phlebitis and massive infiltration with nodule formation. Systemic steroids, posterior subtenon and intravitreal triamcinolone acetonide (IVTA) have been advocated. In one study IVTA was given in 12 patients of Eale’s disease. At eight weeks, 10 of them showed reduction in leakage. Photocoagulation is the mainstay of treatment for stage of ischemia and neovascularization. Sectoral scatter for NVE and PRP for NVD is recommended. In a study at AFIO, 99 Eales’ patients were recruited over...
three years to ascertain the usefulness of laser photocoagulation in managing asymptomatic eyes. 90% (39) of the patients receiving photocoagulation (n = 43) showed visual improvement while 21% (9) of the control group (n = 43) showed improvement14.

Vitrectomy is performed for non clearing vitreous hemorrhage, tractional RD threatening macula, vitreous membranes with or without RD and combined tractional and rhegmatogenous RD. Patients who had photocoagulation prior to surgery show better prognosis. Vitrectomy is also found useful in managing asymptomatic fellow eyes of treated Eales’ patients15. Anterior retinal cryoablation is applied for clearance of vitreous hemorrhage and ablation of ischemic peripheral retina or areas of NVE and is usually reserved as an adjunct to photocoagulation in Eales’ disease. Anti VEGF therapy is a promising new option as an adjunct to other therapies. A large prospective study has been planned at AFIO in a bid to explore the role of intravitreal bevacizumab in Eales’ disease study its demographics, establish a treatment protocol and open new vistas for research. It is the 21st century with a bitter reality that Eales’ is still an agonizing dilemma – an unrevealed mystery that awaits our joint endeavours to allay the distress of affected young males of this region.

REFERENCE

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