

Case Report

Eales disease: A rare primary perivasculitis of the retina

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Introduction

Retinal vasculitis is a sight-threatening condition characterised by inflammation of the retinal vessels. It can occur as a primary disease or present in association with various diseases. Eales disease is a rare primary perivasculitis of the retina [1]. Here we present a case of a young female presenting with reduced visual acuity who was diagnosed with Eales disease and treated successfully.

Case Presentation

A 24 year old, previously healthy female from Jaffna in the Northern Province of Sri Lanka presented to the ophthalmology clinic of the Jaffna Teaching Hospital with a four week history of reduced vision and a one week history of floaters in her right eye. She had no ocular trauma or infection. She is a mother of two children and both pregnancies were uneventful. She was given the Bacillus Calmette–Guérin (BCG) vaccination at birth according to the extended program of immunization (EPI) schedule.

On examination, the best corrected visual acuity was 6/18 in the right eye and 6/9 in the left eye. Both pupils were equally reactive to light. Fundusoscopic examination revealed bilateral retinal vasculitis and periphlebitis with mild to moderate vitreous hemorrhage in the right eye (Figures 1a and 1b).

Blood pressure was 130/80 mmHg. All peripheral pulses were felt. BCG vaccine scar was present. Other system examination was unremarkable. Laboratory investigations including full blood count, urine full report, erythrocyte sedimentation rate, C-reactive protein, fasting plasma glucose, serum electrolytes, liver biochemistry and serum creatinine were within the normal range. Chest radiograph was normal with no evidence of active or past tuberculosis or sarcoidosis.

Tuberculin skin test was negative with a reading of 5mm. Anti-nuclear antibody, perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA) and cytoplasmic anti-neutrophil cytoplasmic antibodies (C-ANCA) were negative. Antiphospholipid antibodies were within the normal limits. Venereal Disease Research Laboratory test (VDRL) was non-reactive. Antibody testing for toxoplasmosis was negative. Plasma homocysteine level was marginally elevated (15.26 µmol/L).

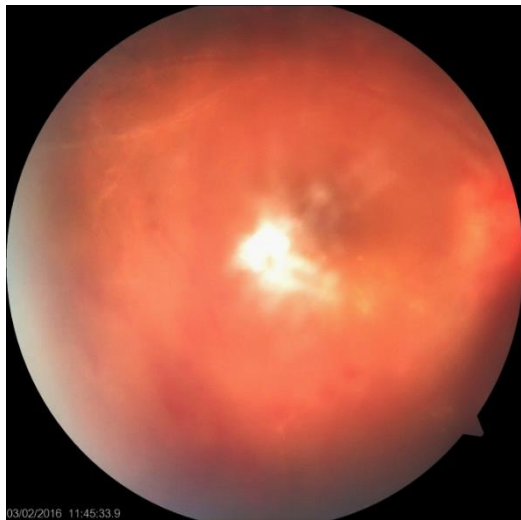


Figure1a : Funduscopy showing retinal vasculitis and periphlebitis in left eye

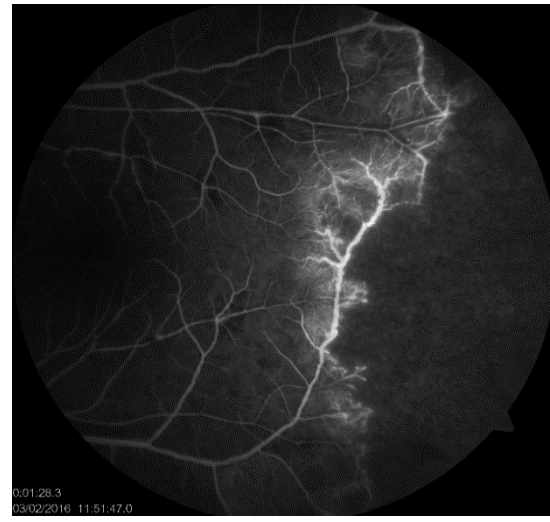


Figure1b : Funduscopy showing retinal vasculitis and periphlebitis with mild to moderate vitreous haemorrhage in right eye

A fluorescein angiogram (FFA) was done which showed the presence of peripheral ischaemia as evidenced by retinal peripheral capillary non perfusions (capillary dropouts) with periphlebitis in both eyes (Figure 2).



left eye



right eye

Figure 2 : Fundus fluorescein angiogram (FFA) showing retinal peripheral capillary non perfusions with periphlebitis

The diagnosis of Eales disease was made, based on the typical fundoscopic appearance and reasonable exclusion of primary and secondary causes of vasculitis. Patient underwent laser photocoagulation to the peripheral retina of both eyes and was given bilateral subtenon injections of depomedrol followed by oral prednisolone 90mg (2mg/kg).

Oral prednisolone was continued at a dose of 90 mg/day for a further two weeks and gradually tapered over three months. She was given folic acid for 6 months and three doses of vitamin B12 intramuscular injection. She did not have any features of relapse and vision was stable on review 6 months after the initial presentation. She was lost to follow-up thereafter.

Visual acuity improved in the right eye 6/9 and left eye 6/6 3 weeks from the initiation of treatment. Her right eye vitreous haemorrhage cleared and retinal vasculitis resolved.

Discussion

Diagnosis of sight-threatening retinal vasculitis in a young female is challenging for both the treating clinician and the patient. Retinal vasculitis can present either as an isolated idiopathic disease or as a manifestation of infective or neoplastic conditions. It is also associated with systemic inflammatory or autoimmune disorders [2].

This disease entity was first reported as a case series of recurrent retinal haemorrhage in young adults by Henry Eales, who believed it to be a part of a vasomotor neurosis [3]. Patients usually present with symptoms of reduced visual acuity, floaters or vitreous hemorrhage in one eye, although fundoscopic changes are often seen bilaterally [1,4,5]. Eales disease is an idiopathic venous occlusive disease which is characterized by three overlapping stages of retinal phlebitis, peripheral nonperfusion and retinal neovascularization [1]. Another staging system includes a fourth stage characterised by rubeosis iridis, neovascular glaucoma, complicated cataract and optic atrophy [6].

The pathophysiology and aetiology of Eales disease is still controversial and it has been recognised as a primary vasculitis of unknown aetiology [1]. Demonstration of tubercle bacilli in the pathological specimens of patients with primary vasculitis by Gilbert and Stock in 1935 and 1937[7,8] and an increased number of cases in tuberculosis endemic areas in patients with positive Mantoux tests raised a suspicion of an association between tuberculosis and Eales disease. In 2013, Biswas et al., proposed that the term “presumed tuberculous retinal periphlebitis” be substituted instead of Eales disease [9]. Further the need for corticosteroid treatment with the risk of activation of latent tuberculosis is a concern for the treating clinician.

Higher frequency of certain human leukocyte antigen types (HLA) in patients with Eales disease has been observed and has given rise to the hypothesis that individuals with HLA predisposition may develop retinal vasculitis triggered by an inactive mycobacterial antigen [10]. In addition to tuberculoprotein hypersensitivity, HLA association and oxidative stress, hyperhomocysteinaemia is also reported in Eales disease [11]. An elevated homocysteine level was observed in our patient as well.

The diagnosis of Eales disease is made by the typical fundoscopic and fluorescein angiographic findings. Several conditions including leukaemia, sarcoidosis, tuberculosis, syphilis, toxoplasmosis, systemic lupus erythematosus, Wegener’s granulomatosis, Behcet’s disease and viral retinitis can mimic the vasculitic phase of the disease. Diabetic

retinopathy and branch and central retinal vein occlusion are the common mimickers of the proliferative phase [1].

The treatment of Eales disease depends on the stage of the disease. A full course of anti-tuberculous treatment (ATT) of 9 months along with a short course of glucocorticoids is recommended by some investigators [9], whereas some reserve ATT for patients with acute phlebitis with massive infiltration, nodule formation and complete obliteration of venous segments [1]. Vitamin B12 and folate treatment is recommended in cases of hyper homocysteinaemia until the homocysteine level returns to normal [9].

Conclusion

Our patient who presented with a four week history of reduced vision in the right eye was diagnosed to have Eales disease with the typical fundoscopic appearance. Laboratory investigations within the normal range including inflammatory markers, normal chest radiograph, negative autoimmune screening and negative antibodies for most common pathogens, along with the prompt response to corticosteroid treatment, further supports the diagnosis of Eales disease. The initial reduction of visual acuity in the right eye was due to vitreous haemorrhage and it improved with treatment to 6/9 in the recovery stage of haemorrhage. A negative Mantoux test, normal inflammatory markers, normal chest radiograph and early disease stage on presentation prompted us to treat with steroids alone, without institution of antituberculosis treatment. Marked symptomatic improvement in vision as well the resolution of retinal vasculitis as evidenced by fundoscopy alleviated the need for ATT. The need for ATT treatment must be decided on an individual basis. Supplementation with vitamin B12 and folate may have a role in the treatment of Eales disease.

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