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CASE REPORT

A 23-year old female patient with Eales Disease: case report

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ABSTRACT

During the 19th century, Henry Eales described the clinical features of recurrent retinal hemorrhages in young patients. The condition was later established as a vaso-occlusive inflammatory disease of the peripheral retina that predominantly affects healthy young male individuals, especially in developing countries. We present the case of a 23-year-old female with recurrent floaters, sometimes accompanied by blurring of vision, in the left eye. Flourescein angiography of the left eye showed vitreous hemorrhage on the inferior and temporal portions of the retina. Hematologic and autoimmune diseases were ruled out as possible causes of the condition during the first few years. Further systemic work-up revealed increased erythrocyte sedimentation rate and a positive Mantoux test. A diagnosis of Eales Disease was made on the sixth year of follow up. The eye condition has been managed with intermittent focal laser treatment and short courses of topical prednisolone acetate. Despite the delay in the establishment of the diagnosis and the patient's refusal to undergo vitrectomy, the recurrent condition has been tolerated by the patient who has been functional at work most of the time.

Keywords. recurrent vitreous hemorrhage, retinal vasculitis, retinal neovascularization, photocoagulation

INTRODUCTION

In 1880, Henry Eales, a British ophthalmologist, described a condition characterized by recurrent vitreous hemorrhages among young men.[1] For the succeeding centuries, many have followed Eales' findings, and further studies emerged regarding the immunologic, molecular, and biochemical aspects of Eales Disease that implicate *Mycobacterium tuberculosis* genome, human leukocyte antigen, retinal autoimmunity, and radical-mediated damage in the pathogenesis of the disease.[2] In one study in India, one out of 200-250 ophthalmic patients was reported to have Eales Disease.[3] The condition primarily affects males,[3 4] but in this report, we describe the clinical features, diagnostic and therapeutic management, and outcomes of an otherwise healthy young female who we eventually diagnosed as having Eales Disease.

CLINICAL FEATURES

A 23-year-old female came to our clinic in 2008 with a 2-year history of floaters in her left eye. There were no accompanying systemic signs and symptoms. The patient had no history of ocular trauma or previous tuberculosis infection. She tolerated the condition until she noticed a marked decrease in visual acuity, which prompted her to seek consultation. The patient works as a nurse. She had unremarkable past medical, family, and social history. A review of systems was also unremarkable, save for her eye complaint. Visual acuity of the left eye was 20/80. A fundus examination on presentation revealed hemorrhages at the infero-temporal portion of the retina of the left eye. The right eye findings were within normal limits. The patient's clinical course is summarized in Table 1.

DIAGNOSTIC AND THERAPEUTIC APPROACHES

A fluorescein angiogram (FA) of the left eye done in 2008 (Figure 1) revealed vitreous hemorrhages on the inferior and temporal parts of the retina. Capillary dropouts were seen on the inferior portion of the retina on the AV phase of the angiogram. Neovascularizations were also noted, mostly located at

IN ESSENCE

Eales Disease is an inflammatory vaso-occlusive disease of the retina. It has been associated with tuberculosis or tuberculin protein exposure.

This disease among otherwise healthy young individuals is characterized by neovascularizations in the retina and subsequent vitreous hemorrhages.

Laser photocoagulation usually controls the formation of neovascularizations, and anti-inflammatory drugs decrease intra-ocular inflammation. Non-resolving hemorrhages may require vitrectomy. In general, prognosis is good among patients receiving prompt diagnosis and adequate treatment.

Table 1 Chronology of signs and symptoms, physical examination findings, diagnostics, therapeutics and outcomes*

Timeline	Signs and symptoms	Physical examination	Diagnostics	Working diagnoses	Therapeutic management	Outcome
2008	Floaters in inferotemporal portion of the eye	**VA:** 20/80; **SLIO:** hemorrhages in the inferotemporal portion of the retina	**FA:** vitreous hemorrhages in the inferior and temporal portions of the retina, capillary dropouts and neovascularizations in the inferior portion of the retina	Vitreous hemorrhage	Focal laser treatment	VA improved to 20/20 after 9 months
2010	Blurring of vision and recurrence of floaters	**VA:** 20/80; **SLIO:** new-onset vitreous hemorrhage	**FA:** new retinal neovascularizations; **WBC:** leukocytosis, 11 x 10^9 per liter; **ESR:** raised, 26 mm/hr; **ANA:** normal; **RF:** normal; **CRP:** normal	Vitreous hemorrhage, possibly secondary to any of the following: tuberculosis, systemic lupus erythematosus, Eales Disease, leukemia	Focal laser treatment; ketorolac 1 drop three times a day on the affected eye for 1 month; oral prednisone 50mg/kg/day for 1 month, then tapered to 10mg/kg/day for 6 weeks	VA improved to 20/20 after 3 months
2013	Blurring of vision and recurrence of floaters	**VA:** 20/30	**FA:** new vitreous hemorrhages and retinal neovascularizations; **ESR:** raised, 29 mm/hr; **ANA:** normal; **RF:** normal; **CRP:** normal	Strongly considering Eales Disase	Focal laser treatment; topical prednisolone acetate 4 times a day for 1 month	The patient had the same VA, but floaters were minimized and bleeding was controlled
Early 2014	Asymptomatic	---	---	The patient was referred to another ophthalmologist who also considered Eales Disease	---	The need to establish past history or ongoing tuberculosis infection was reiterated to the patient
Late 2014	New-onset minimal floaters	**VA:** 20/30-2; **SLIO:** minimal new-onset vitreous hemorrhage	**CXR:** within normal limits; **Mantoux test:** positive	Eales Disease, Stage 3b	No intervention. The patient was monitored for new signs and symptoms	No change in VA; persistent minimal floaters
2015	Blurring of vision and minimal floaters	**VA:** 20/100; **SLIO:** new-onset vitreous hemorrhage, inflammatory cells noted on the posterior segment of the retina	---	Eales Disease, Stage 3b	Topical prednisolone acetate 1 drop every 4 hours for 1 month	Visual acuity improved to 20/40 after 1 month of treatment

* All signs and symptoms, physical examination findings, ophthalmologic investigation findings, diagnoses, management of the eye, and outcomes pertain to the left eye. The patient's right eye has been normal throughout the disease course. ANA – antinuclear antibody; CRP – C-reactive protein; CXR – chest X-ray; ESR – erythrocyte sedimentation rate; FA – fluorescein angiography; RF – rheumatoid factor; SLIO – slit lamp indirect ophthalmoscopy; VA – visual acuity; WBC – white blood cells.

the inferotemporal area of the retina. Our working diagnosis at this point was vitreous hemorrhage in the left eye. Focal laser treatment improved the visual acuity of the affected eye.

The floaters in the left eye recurred almost yearly over the next seven years, sometimes accompanied by blurring of vision in the same eye. Visual acuity in the left eye fluctuated between 20/30 and 20/100. Repeat funduscopic examinations and FAs in 2010 and 2013 revealed new-onset hemorrhages and retinal neovascularizations. During these years, we attempted to establish the underlying cause of the recurrent retinal hemorrhages. We knew that there was no past or ongoing trauma to the left eye. We were able to rule out systemic lupus erythematosus (normal antinuclear antibody, rheumatoid factor and C-reactive protein). The slightly elevated white blood cell count (11 x 10^9 per liter) was not convincing enough to think of a hematologic condition, such as leukemia. The raised erythrocyte sedimentation rates (26-29 mm/hour) in 2012 and 2013 were not specific enough to point us to a particular explanatory condition, either.

We requested for chest radiograph and Mantoux test to check if the patient had tuberculosis, but she was apprehensive for some time about her condition and did not comply with the added diagnostics. However, after we were able to rule out trauma, systemic lupus nephritis, and leukemia, we were already strongly considering Eales Disease in 2013. Early in 2014, another ophthalmologist who saw the patient was also considering Eales Disease. Later in 2014, we were finally able to convince the

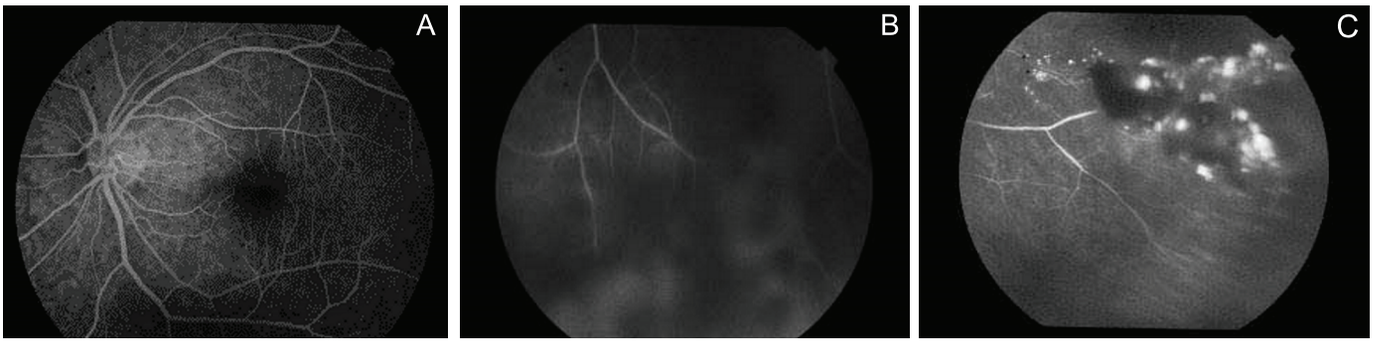

Figure 1 (2008) Fluorescein angiogram of the left eye, showing capillary dropouts on the inferior portion of the retina on the AV phase of the angiogram (A). Vitreous hemorrhages were seen on the inferior and temporal areas of the retina (B and C). Neovascularizations were also seen, mostly located in the inferotemporal portion of the retina.

patient to have the pending diagnostics done. The chest X-ray result was within normal limits, but the Mantoux test was positive. We then decided on a diagnosis of Eales Disease, Stage 3b at that point.

Focal laser treatments were repeated in 2010 (Figure 2) and 2013 (Figure 3). Oral steroids and topical ketorolac were added to the treatment in 2010, and visual acuity of the left eye returned to normal in that year. While on focal laser treatment in 2013, the patient was also prescribed with topical prednisolone acetate. Floaters were minimized subsequent to the treatment, but visual acuity persisted to be decreased since 2013. During this time, we noted fibrous proliferation at the inferior and temporal portions of the retina (Figure 3). Since this can cause traction on the retina that could lead to retinal detachment, we advised the patient to undergo pars plana vitrectomy in the left eye, but she opted for conservative therapy. After establishing the diagnosis of Eales Disease, we prescribed intermittent short courses of topical prednisolone acetate when the hemorrhages would recur.

OUTCOMES

Follow-up eye examinations have been aimed at closely monitoring any new or recurrent vitreous hemorrhages and new blood vessel formations. As of 2015, the patient reported seeing minimal floaters, however, eye examinations showed controlled inflammation and neovascularization. Currently, the patient is functional at work as a nurse and performs

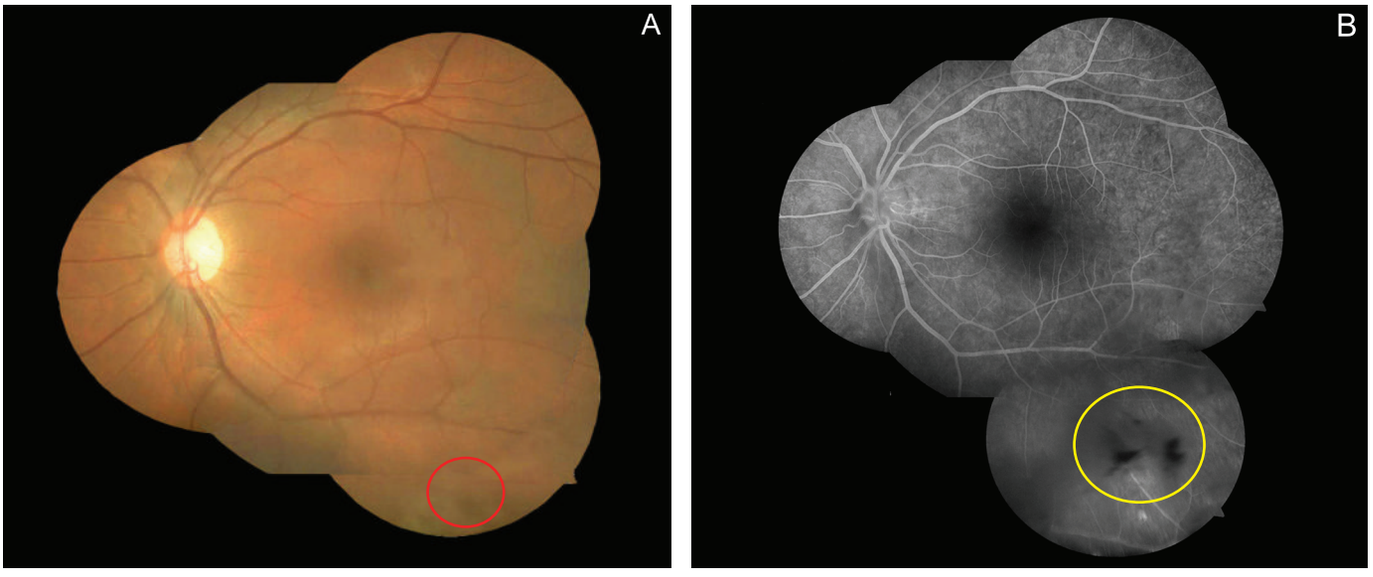

Figure 2 (2010) Fundus photo (A) shows new-onset vitreous hemorrhage at the inferotemporal portion of the retina (A: red ring). Laser marks are also visible in the area. Fluorescein angiogram (B) shows new-onset blocked hypofluorescence (B: yellow ring) that corresponds to the vitreous hemorrhage seen in the fundus photo. Hyperfluorescence seen on the late phase of the angiogram shows new-onset leakage due to new-onset neovascularizations elsewhere.

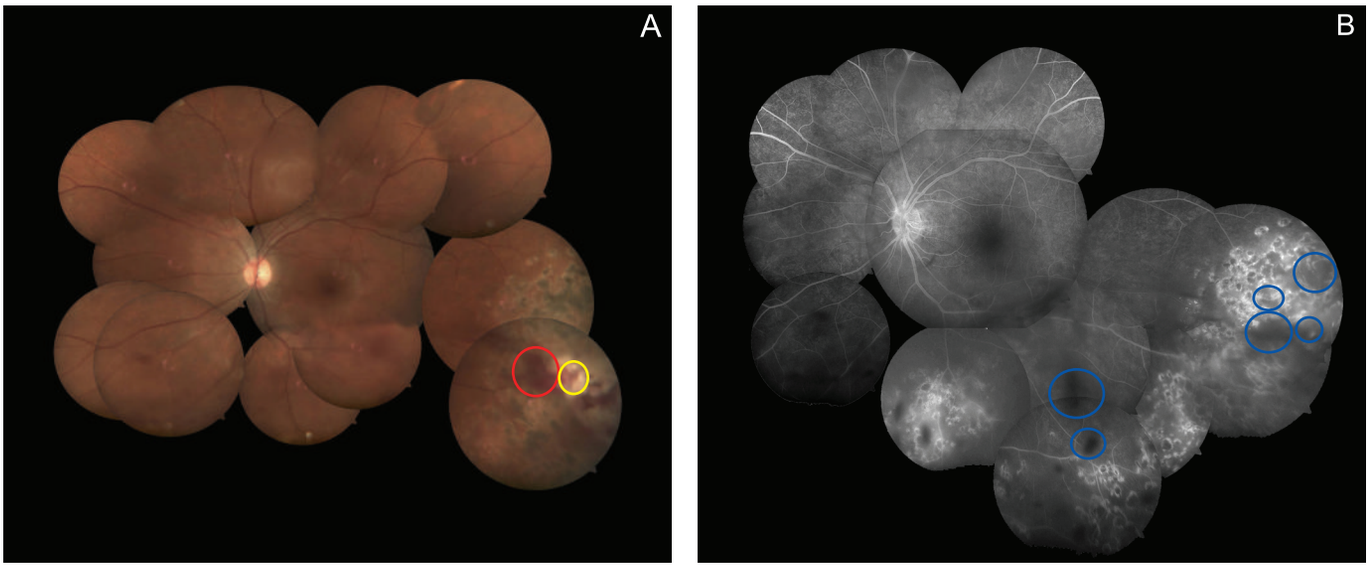

Figure 3 (2013) Fundus photo (A) shows new-onset vitreous hemorrhage on areas with laser marks (A: red ring). Fibrous proliferations (A: yellow ring) were also noted on the same area. Fluorescein angiogram (B) shows new-onset multiple blocked hypofluorescence (B: blue rings) that correspond to the vitreous hemorrhages seen in the fundus photo. Hyperfluorescence seen on the angiogram shows staining on the laser marks and leakage due to new-onset neovascularizations elsewhere.

activities of daily living with ease.

DISCUSSION

Our patient came due to floaters on her left eye. This is consistent with the initial presentation of patients with Eales Disease. Funduscopic examination and FA findings in our patient proved the presence of hemorrhages and neovascularizations in the affected eye. Over several years, and despite repeated laser treatments and corticosteroid courses, the floaters recurred, sometimes accompanied by blurring of vision. We requested laboratory examinations that would help us rule out autoimmune and hematologic diseases as possible causes of the vitreous hemorrhages. When most of the laboratory results came out normal, we tried to establish a possible previous exposure to tuberculosis. However, our patient was hesitant about having a tuberculosis work-up, and we had to respect her decision, so the performance of the chest X-ray and Mantoux test was rather delayed. The Mantoux test, which turned out to be positive, confirmed the patient's exposure to tuberculosis and helped us decide on a diagnosis of Eales Disease.

Eales Disease is an inflammatory, vaso-occlusive retinal disease usually affecting young adults. It is believed to be associated with exposure to tuberculosis or tuberculin protein.[2] As in our patient's case, the onset of symptoms typically occurs during the second to third decade of life.[5-7] The inflammatory process of the retinal vessels leading to vaso-occlusion is still not fully understood. However, this pathologic process leads to retinal ischemia that causes neovascularization. The newly formed vessels tend to bleed easily, and this plays a major role in decreasing the patient's vision.[8] Eighty percent of patients with Eales Disease develop neovascularization either on the disc or elsewhere,[9] and bilateral involvement of the eyes is common.[5-7]

The classification system of Eales Disease is useful in assessing disease severity and in monitoring the effects of medical, laser or surgical treatment.[10] It also provides a method of categorization based on both funduscopic and fluorescein angiographic variables that have a bearing on visual outcome prognostication. According to the classification system, our patient who experiences vitreous hemorrhages has Eales Disease, Stage 3b.[10]

The treatment of Eales Disease is usually geared towards alleviating the symptoms of the disease. This is achieved by controlling retinal perivasculitis and decreasing the risks of vitreous hemorrhage from new blood vessels.[3] For almost all neovascularizations in the retina, regardless of the cause and if there is no contraindication, laser photocoagulation, which prevents proliferation of new vessels in the ischemic retina, is the mainstay of treatment.[11] Observation and corticosteroids are the ideal treatment options for inactive retinal vasculitis and active

perivasculitis, respectively. Our patient was treated with corticosteroids and underwent laser photocoagulation, which improved her visual acuity. However, the recurrence of symptoms led to new tissue formation that would benefit from pars plana vitrectomy. For non-resolving vitreous hemorrhage, pars plana vitrectomy can be perfomed.[12] Vitrectomy ensures a clear optical axis that facilitates photocoagulation, relieves vitreous traction, and may promote regression of new blood vessels.[13] Having early vitrectomy – within 3 to 6 months from the onset of vitreous hemorrhage – yields improved visual acuity and favorable prognosis.[14] We explained the benefits of vitrectomy to the patient, but she was not willing to undergo the surgical procedure.

Eales Disease has a generally good prognosis and, with appropriate treatment, rarely leads to blindness.[5 7]

Our patient with Eales Disease has been coming to our clinic to be relieved of two rather bothersome symptoms in her left eye – floaters and blurring of vision. We were able to observe our patient long enough to document the recurrent nature of her symptoms. While we were giving her intermittent symptomatic treatments for the symptoms, we offered to do pars plana vitrectomy in the left eye to provide a more definitive treatment for her condition. She refused such surgical treatment, and she somehow came to stand having to go through laser photocoagulation and apply topical corticosteroids on her left eye every time these treatments are indicated.

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Patient consent

Obtained

Reporting guideline used

CARE Checklist (http://www.care-statement.org/downloads/CAREchecklist-English.pdf)

Article source

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Competing interests

None declared

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