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Epidemiology of Eales Disease in the Central Western India

Review Article

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Abstract

Objective: To investigate the clinical features and propose a new staging system based on the clinicopathological correlation and formulate new guidelines for management of Eales' disease in healthy young males.

Design: Prospective cohort study.

Setting: Vitreo Retinal Department of a tertiary eye care center in Western Central India.

Participants: Seventy-four eyes diagnosed with Eales' disease.

Materials and methods: From 2004 to 2010, patients clinically diagnosed with Eales' disease were enrolled in this study using specific inclusion and exclusion criteria. We examined the patients' anterior and posterior segments thoroughly. We performed the necessary ocular and systemic investigations. We divided the investigations into three stages: inflammatory, ischemic and complications and the patients were treated accordingly. We treated the patients using medical management, photocoagulation or pars plana vitrectomy. The patients were monitored according to standard schedules and formats. All information was documented using a pre-tested online format and statistical analyses were performed using SPSS ver. 17. A p-value < 0.05 was considered to indicate significance.

Outcome measures: Visual acuity.

Results: The cohort comprised 74 cases with a mean age of 30 ± 8.73 years. The visual acuity of the presenting cohort was < 3/60 in 64.9% of eyes. The final visual outcome was > 6/12 in 40 eyes (54.1%), 6/60 to 6/18 in 14 eyes (18.9%) and < 1/60 in the remaining 9 eyes at a mean follow-up of 592 days. The visual parameters differed significantly pre-and post-treatment. We evaluated the visual outcome following surgical management.

Conclusion: We studied epidemiological facts about anterior and posterior segment findings.

Key Words: Eales Diseases; Grading System; Periphlebitis; Vitreous Hemorrhage; Systemic Corticosteroids.

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Introduction

Eales' disease was first described by Henry Eales, a British ophthalmologist, in 1880 and 1882. [1,2] He identified this disease in seven male patients ranging in age from 14 to 29 years with recurrent retinal hemorrhages. In addition, these patients had histories of headache, variation in peripheral circulation, chronic constipation, and epistaxis. [2]

Eales' believed that these symptoms were caused by vasomotor neurosis with constriction of the alimentary vessels and compensatory dilatation of the retinal and nasal vessels. Eales' did not describe retinal vasculitis. [2] However, 5 years later, Wadsworth (1887) described signs of retinal vasculitis. [3] Eales' has subsequently been honored with the eponym for idiopathic retinal Periphlebitis. [4]

Eales' disease is an idiopathic inflammatory vasoproliferative disorder of the retinal veins, which leads to the occlusion of these vessels. [5] It affects primarily healthy young adults between the

ages of 20 and 40 years. It affects primarily males, (80-90%), but can affect females. Elder et al. (1971) reported the possibility of bilateral involvement in 90% of cases. [6] It may rarely affect the arterioles. This vascular occlusion, induced by the inflammation of veins, can lead to proliferative vascular retinopathy and subsequent sequelae, such as recurrent vitreous hemorrhage, and tractional or combined retinal detachment. [5,7]

Eales' disease was common a century ago, but its incidence has declined in western countries. However, Eales' disease is widespread in the Indian subcontinent and certain areas of the Middle East. It is usually seen unilaterally, but can present bilaterally. Eales' disease is characterized by three overlapping stages of venous inflammation (vasculitis), occlusion, and retinal neovascularization. Diagnosis is mostly clinical and requires exclusion of other systemic or ocular conditions that could present with similar retinal features. Recurrent vitreous hemorrhage is the hallmark of Eales' disease. [7]

Treatment options include systemic steroids during the inflammatory stage, retinal photocoagulation usually in the proliferative stage to the non-perfused retina, and early vitrectomy for recurrent vitreous hemorrhage. [6,7,11] Oral steroids are the primary agents used most frequently for the management of Eales' disease, but prolonged systemic use of steroids is associated with serious ocular and systemic side effects. [11] Photocoagulation is the mainstay of treatment in the proliferative stage of Eales' disease. [12] Intravitreal steroids and anti-VEGF have been used in many posterior segment ocular pathologies. [13,14] Visual prognosis is good if treated early in the course of the disease. [7,12,13] Recently, anti-metabolite therapies, especially methotrexate, have been used in Eales' disease with some success. [12]

The prognosis of patients with Eales' disease is variable and depends on the availability of medical care. Many of the patients in the region in which this study was conducted live in areas that are underserved by medical care. Therefore, the treatment outcome may also vary. This study assessed the demographics, clinical presentation and treatment outcome of Eales' disease in Central West India, which is inhabited by mainly tribal populations.

Methods

This study was a prospective interventional case series, designed to evaluate the epidemiology of Eales' disease patients in a region of Central Western India; data were collected from January 2004 to December 2012. The study was conducted in a tertiary care teaching hospital and a tribal center for ophthalmic care, which serves the second largest tribal belt in India. It is located at the junction of three states, Gujarat, Rajasthan and Madhya Pradesh.

Approval from the Institutional Ethics Committee was obtained and all efforts were made to adhere to the guidelines of the Declaration of Helsinki.

Patients were briefed about the procedures of the study and consent was obtained before enrolment.

Patients suspected to have Eales' disease aged between 20 and 40 years underwent exhaustive ocular and medical evaluation for confirmation of a diagnosis of Eales' disease. Our cohort comprised patients diagnosed with Eales' disease with no other systemic or ocular disease.

Subjects with a history of diabetes mellitus, hypertension, collagen vascular disease, sarcoidosis, Bechet's disease, systemic lupus erythematosus, Coats' disease, or syphilis were excluded from the study.

Ophthalmic examinations included best-corrected visual acuity using Snellen's chart, intraocular pressure measurement by applanation tonometry, and anterior segment evaluation by slit-lamp examination.

Evaluations of the posterior segment at the initial and each subsequent visit were performed by direct and indirect ophthalmoscopy, slit-lamp biomicroscopy with 90-D and three-mirror lenses, and fundus fluorescein angiography.

Investigations included complete blood count with sickle-cell screening, erythrocyte sedimentation count, liver function test, fasting plasma sugar level, blood coagulation profiles, Mantoux test using 2 TU/0.1 mL purified protein derivative (PPD), and chest X-ray.

Ultrasonography was performed to detect fractional retinal detachment in eyes affected by vitreous hemorrhage with no fundus view. The location and extent of retinal involvement due to Eales' disease, neovascularization, and fibrovascular traction were documented in all patients by fundus drawing and digital fundus color photography. Optical coherence tomography (OCT) was performed using high-density OCT (Cirrus, 2000, Carl Zeiss Meditec, CA,) as required after it has became available.

The inflammatory stage was treated with medical management, periocular steroids, systemic steroids or methotrexate. Complete blood counts and liver function tests were performed at least once per month in patients prescribed methotrexate. Prior to prescribing methotrexate, a thorough medical consultation was performed by a medical oncologist. The ischemic stage was treated with anti-VEGF or photocoagulation according to availability. The stage of complications was treated with surgical management. The details were documented in a pre-tested, online Eales' disease form and then exported to a Microsoft Excel™ spreadsheet.

Data were analyzed using the SPSS software (ver. 17.0; IBM SPSS Inc., Chicago, IL, USA). Univariate parametrical analyses were used. A p-value of < 0.05 was considered to indicate statistical significance.

Results

The cohort comprised 74 male patients with a mean ages of 30 ± 8.73 years (Table 1). Demographic factors including age, diet, smoking, religion and race were evaluated (Table 2). No significant differences in the demographic factors were observed, with the exception of age of presentation ($p = 0.02$).

Within the cohort, the presenting visual acuity was $< 3/60$ in 64.9% of eyes. The final visual outcome was $> 6/12$ in 40 eyes (54.1%), 6/60 to 6/18 in 14 eyes (18.9%) and $< 1/60$ in 9 eyes at a mean follow-up of 592 days.

We found the anterior segment normal in 65 (87.8%) cases (Table 3). We then evaluated the findings in the posterior segment (Table 4). The treatments prescribed were systemic steroids (64.9%), anti-metabolites (35.1%), photocoagulation (13%), pars plana vit-

rectomy (32.1%) and revision vitrectomy (9.5%) (Table 4).

We evaluated visual parameters pre- and post-treatment, and found a significant difference (p = 0.000, Table 5). We next compared visual outcome following surgical management, and again found a significant difference (p = 0.000, Table 6).

Discussion

Our cohort comprised 74 cases with a mean age of 30 ± 8.73 years. Other reports with a similar demographic profile, including age and gender reported similar findings. [7,15-17] In this study, we have simplified the classification according to the clinical findings. In the inflammatory stage (stage 1), patients with vitritis and vasculitis were treated with systemic and periocular steroids. In the ischemic stage (stage 2), patients with neovascularization and ischemia were treated with laser or anti-VEGF therapy. During the stage of complications (stage 3), cases of traction or mixed retinal detachment were treated with pars plana vitrectomy. Saxena et al. reported a classification and grading system, the proposed classification system may provide guidelines for the management of Eales' diseases. [15] Periphlebitis and vitreous hemorrhage have been reported, but the incidences of vitritis, vasculitis and vitreous hemorrhage have not been described. [7,15-17]

We observed total posterior vitreous detachment in 24.3% of cases. Badrinath et al. reported multifocal vitreo retinal adhesions in 87.8% of cases. [18,19] There was no difference in the response to treatment in the Mantoux-positive and -negative groups. [20]

Oral corticosteroids and deep sub-tenon triamcinolone, used for quadrants two and three reported by Biswas et al., has a significant effect on visual outcome. [21,22] We used anti-metabolites in the form of methotrexate in 35.1% of cases .[7,22] As suggested by many investigators we did not use anti-tubercular therapy for the treatment of Eales' disease.[7] We used laser photocoagulation in the ischemic stage (stage 2) or in those with neovascularization, as reported by Biswas et al. [7,23,24]

In the current study, vitrectomy was performed in 32.1% of cases, which is in contrast to the 6–18% reported by Badrinath et al. [19] This may be due to the greater number of patients being in the stage of complications (stage 3) in our study.

Table. 1 Age Category

AGE CATEGORY	NUMBER (N)	PERCENT (%)
11 TO 20	4	5.4
21 TO 30	42	56.8
31 TO 40	15	20.3
41 TO 50	11	14.9
51 TO 60	2	2.7
Total	74	100.0

Table. 2 Demographic Results

	NUMBER(N)	PERCENT(%)
Hindu Tribal	65	87.8
Socio Economic Status Poor	65	87.8
Maize Eater	66	89.2
Smokers	32	43.2
Bilateral	64	86.5
No Systemic Disorder	72	97.3
Follow Up In Days	592 +/- 374.4	

Table. 3 Clinical Features in Anterior Segment

FINDINGS	NUMBER(N)	PERCENT(%)
Cataract	2	2.7
Chemosis	2	2.7
Chemosis, Rubeosis Iridis	1	1.4
Leucokoria	1	1.4
Normal	65	87.8
Pterygium	2	2.7
RAPD	1	1.4
Total	74	100.0

Table. 4 Clinical Features In Posterior Segment

CLINICAL FINDINGS	PRESENT		NO VIEW	
	NUMBER(N)	PERCENT(%)	NUMBER(N)	PERCENT(%)
Normal Anterior Segment	65	87.8		
Vitreous Haze 2 Quadrants	20	27	23	31.1
Vitreous Haemorrhage	20	27	23	31.1
Vasculities-St	48	64.9	16	21.6
Vasculities-It	38	51.4	18	24.3
Vasculities-Sn	25	33.8	25	33.8
Vasculities-In	16	21.6	25	33.8
Rubeosis Iridis	6	8.1	10	13.5
NVE/NVD	23	31.1	15	20.3
Macular Edema	24	32.4	17	23
Retinal Detachment	32	43.4		
Systemic Steroids	48	64.9		
Antimetabolites	26	35.1		
Laser	10	13.5		
Pars Plana Vitrectomy	24	32.1		
Revision Pars Plana Vitrectomy	7	9.5		

NVD-Neovascularization Disc; NVE-Neovascularization Elsewhere

Table. 5 Comparative Study Of Vision Following Medical Mamnagment Of Eales' Disease

POST TREATMENT VISION	PRE TREATMENT VISION					Total
	<1/60	1/60 TO 3/60	6/60 TO 6/36	6/24 TO 6/18	6/12 TO 6/9	
<1/60	14	4	3	0	0	21
1/60 TO 3/60	6	8	0	0	0	14
3/60 TO 5/60	2	0	0	0	0	2
6/60 TO 6/36	5	4	2	1	1	13
6/24 TO 6/18	3	1	1	5	1	11
6/12 TO 6/9	0	1	0	6	1	8
6/6 TO 6/5	0	0	0	0	5	5
Total	30	18	6	12	8	74

P=0.000

Table.6 Comparative Study Of Vision Following Surgical Management Of Eales Diseases

POST OPERATIVE VISION	SURGERY PERFORMED		Total
	NO	PP VIT	
<1/60	5	16	21
1/60 TO 3/60	8	5	13
3/60 TO 5/60	2	0	2
6/60 TO 6/36	10	3	13
6/24 TO 6/18	11	0	11
6/12 TO 6/9	7	0	7
6/6 TO 6/5	5	0	5
Total	48	24	72

P=0.000 PP VIT-PARS PLANA VITRECTOMY

Conclusion

We have studied newer aspects of demographic details, anterior segment findings and posterior segment findings. We have found that final visual outcome significantly improved following medical or surgical treatment.

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