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Study of long term course and visual prognosis in Eales disease: A retrospective study

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Abstract

Purpose: To study the long term course and visual prognosis in Eales disease

Methods: Patients who had been examined between 1984 – 1989 were included in the study.

Results: A total of 34 patients were followed up of a minimum period of 10 years. The retinal leisons found during the initial examination was vitreous haemorrhage (34.5%) NVE (32.7%) vascular sheathing (30.9%) FVP (29.1%) NVD (14.5%) NVD and NVE (10.9%) & RD (5.4%) of eyes. visual acuity improved in 23.6% maintained in 58.2% & decreased in 18.2% of eyes. The complications were cataract, Tractional RD, Rubeosis iridis, Neovascular glaucoma & Phthisis bulbi.

Conclusion: The most important elements in dealing with Eales disease are early diagnosis, periodic follow up and adequate therapy depending upon the stage of the disease.

Keywords: Eales Disease, Fibrovascular Proliferation (FVP), Neovascularization of disc (NVD), Neovascularization elsewhere (NVE), Retinal detachment (RD).

Introduction

Eales' disease is an idiopathic Obliterative vasculitis that predominantly affects the peripheral retina of young adults. Retinal changes include perivascular phlebitis, peripheral non perfusion, and neovascularization. Visual loss is characteristically caused by bilateral recurrent vitreous haemorrhage and its sequelae. In 1880 and 1882 Henry Eales[(1,2)] a British ophthalmologist described the clinical picture of recurrent retinal haemorrhages in young adults. His seven patients were all young men (age, 14 to 29 years). They had in common a history of headache, epistaxis, variation in peripheral circulation, dyspepsia, and chronic constipation. Eales believed it to be a vasomotor neurosis wherein constriction of the alimentary vessels and compensatory dilatation of those of the head, lead to rupture of retinal and nasal vessels with consequent haemorrhage. However, retinal vasculitis was not described by Eales. Wadsworth[(3)] described the associated signs of retinal inflammation five years later. In the century that has followed, Henry Eales has been honoured with the eponym for the disease characterized by idiopathic recurrent vitreous haemorrhage in otherwise young and healthy adults. The reeponym has been retained because of its widespread recognition and the emphasis that it gives to the common clinical features and certain significant therapeutic implications.

Since Eales' disease was originally thought to be a disease of retinal veins, Elliot[(4)] initially suggested that the disease be called "periphlebitis retinae." However, near equal prevalence of venular and arteriolar inflammation was later documented and hence, Kimura et al,[(5)] Keith Lyle and Wybar,[(6)] and Cross' preferred to call it "retinal perivasculitis" or even "retinal vasculitis" since the vessel wall itself is involved in inflammation and not merely its surrounding connective tissue. Eales' disease has been reported from the United Kingdom, the United States, and Canada in the later half of 19th and early 20th century. But for unknown reasons, it is now rare in more developed countries and is more commonly reported from the Indian subcontinent. The reported incidence in India is one in 200 to 250 ophthalmic patients.[(7)]

Eales' disease predominantly affects healthy young adults. There is a male predominance. The peak age of onset of symptoms is 20 to 30 years. Most patients' symptoms related to vitreous haemorrhage, such as floaters, cobwebs, blurring, of vision. Others have blurring of vision associated with retinal vasculitis but without vitreous haemorrhage. Many patients complain of symptoms in only one eye, but detailed fundus examination of the fellow eye reveals the early changes associated with Eales' disease such as periphlebitis, vascular sheathing, or peripheral non-perfusion. Eventually, between 70 to 80% of patients develop bilateral involvement though the extent of retinal involvement may not be to the same extent. Duke-Elder' noted 90% bilateral retinal involvement and others[(8)] have noted only 50% bilateral involvement.

Aim of the Study

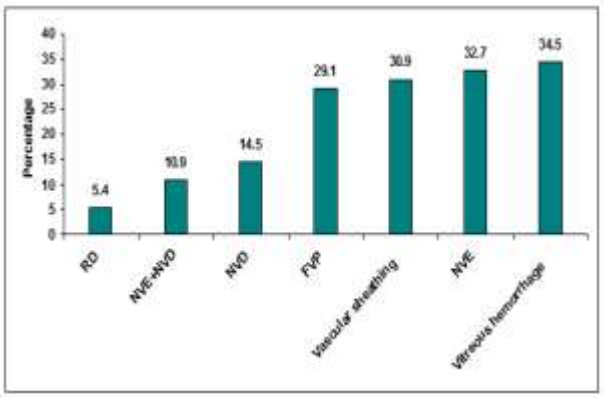
The aim of the study is to find out the clinical course and visual progmosis in eales disease in 10 years follow up. This is a retrospective case study of 34 patients and a total of 55 eyes were included in the study. The period of the study is between 1984 to 1989. The inclusion criteria is patient diagnosed as eales disease with a minimum of 10 years follow up. A detailed history and complete ophthalmic examination including slit lamp examination and fundus examination was done by

indirect ophthalmoscopy. Those patients whose fundus was visible underwent fluorescein angiography, the other B-scan ultrasound. Routine lab investigations like complete blood count, ESR, routine urine examination and mantoux test were done when necessary. Patients were either followed or treated with one or more: medical therapy(oral corticosteroids, anti-tubercular therapy), Photocoagulation, pars plana Vitrectomy, cryotherapy and retinal detachment surgery. Visual improvement was defined as improvement of vision of two Snellen lines or more than the pre-treatment vision at last follow up. Visual stabilization was defined as maintenance of same vision without improvement or detoriation of vision less than two Snellen lines as compared to pre-treatment vision.

Results

In our study all the patients were males and the mean age of presentation was 27.18 years (range from 10 to 48 years). The total number of patients was 34. The disease was bilateral in 21(61.8%) patients and unilateral in 13(38.2%) patients. The mantoux positivity rate was 50% and association with tuberculosis was found in 6(17.6%) patients. Floaters and sudden diminution of vision was the most common symptom. Vitreous haemorrhage was the most common presentation in 19(34.5%) eyes. This is followed by NVE in 18(32.7%), Vascular sheathing in 17(30.9%), Fibrovascular proliferation (FVP) in 16(29.1%), NVD in 8(14.5%), NVE and NVD IN 6(10.9%) and retinal detachment in 3(5.4%) of eyes.

Major retinal lesions at the initial examination



The treatment modality applied were – Photocoagulation in 32(58.2%), PPV in 18(32.7%), RD surgery in 3(5.4%), cryotherapy in 2(3.6%) eyes. A total of 15 eyes received no treatment.

The Treatment modality given:

Treatment	No. of eyes	%
Photocoagulation	32	58.2
PPV	18	32.7
RD Surgery	3	5.4
Cryotherapy	2	3.6
No Treatment	15	27.3

The initial and final visual acuity of 55 eyes:

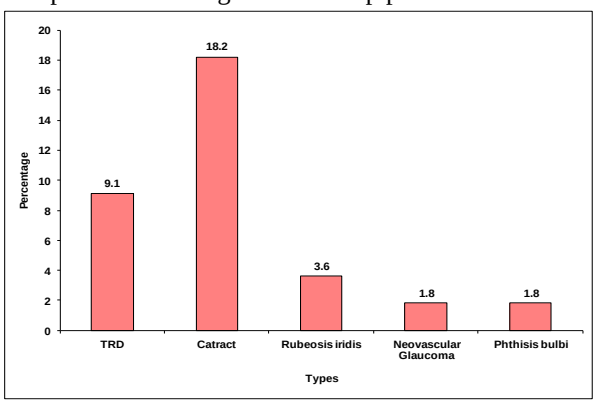
Visual Acuity	Initial		Final	
	No. of eyes	%	No. of eyes	%
NLP	.		9	16.4
PL	8	14.5	-	-
HMCF	2	3.6	2	3.6
CF 1-3m	6	10.9	4	7.3
6/60	5	9.1	3	5.5
6/36	2	3.6	1	1.8
6/24	1	1.8	3	5.5
6/18	5	9.1	3	5.5
6/12	3	5.5	2	3.6
6/9	7	12.7	8	4.5
6/6	16	29.1	20	36.4

The mean visual acuity at presentation was1.004 log units and after the mean follow up1.323 log units. P value =0.08.

Visual acuity changes according to treatment modality:

Treatment	Visual Acuity					
	Decreased		Maintained		Increased	
	N	%	N	%	N	%
Photocoagulation	12	21.8	5	9.1	15	27.3
PPV	6	10.9	2	3.6	10	18.2
Cryotherapy	-	0	2	3.6	-	0
RD Surgery	1	1.8	1	1.8	1	1.8

Complications during the follow up period:



Discussion

Eales disease is an idiopathic obliterative vasculitis that involves the peripheral retina of young adults. It may start with retinal oedema, followed by progressive cuffing of venules, peripheral retinal vessel non-perfusion and retinal neovascularization. Disc and retinal neovascularization is observed in 80% cases. Bleeding from neovascularization is one of the major causes of visual loss.

In Eales disease, males were commonly affected 9, 10, usually bilateral 9, 11 and mostly affects the young adults.

Features	Atmaca LS et al**	SN Study
Study period	1970-1991	1984-1989
Total no. of patients	130	34
Mean age	29.6	27.18
Sex	96.9% Males	100% Males
Bi laterally	73%	61.8%
Mean follow up	142.4 months	140 months

In a study by Atamca ls et al Vascular sheathing was found in 42.2%, NVE in 32.9%, Vitreous haemorrhage in 19.1%, FVP in 6.2%, NVD in 4%. NVD & NVE in 3.1% and RD in 4.4%. Whereas in our study major (SN) retinal lesion was Vitreous haemorrhage with 34.5%.This is followed by NVE in 32.7%, Vascular sheathing in 30.9%, FVP in29.1%, NVD in 14.5%, NVD and NVE in 10.9% and RD in 5.4% of eyes.

The study by Atamca ls et al, Photocoagulation was the most common treatment modality given in 57.2% of eyes, PPV in 14.5%, RD surgery in 2.5%, Cryotherapy in 0.4%, where no treatment was given in 21.6% of eyes. In our study (SN study) also Photocoagulation was the most common treatment modality given in 58.2% of eyes, PPV in 32.7%, RD surgery in 5.4%, Cryotherapy in 3.6%, and no treatment was given in 27.3% of eyes. In our study most of the patients presented in the stage of neovascularization and vitreous haemorrhage. Photocoagulation is the treatment of choice in the proliferative stage of the disease 13.There was complete regression of neovascularization after the mean follow up.

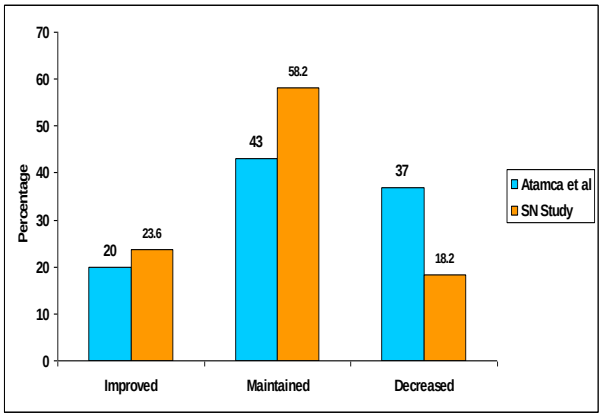
In our study, 2 patients received systemic corticosteroids, anti-tubercular therapy(ATT) in 6 patients and systemic corticosteroid and ATT in 1 patient. In patients with positive Mantoux test and active perivasculitis treatment with a combination of oral corticosteroids and and anti-tubercular therapy have been recommended 12.

In our study (SN) cataract was the most common complication IN 18.2% whereas Tractional RD was the most common complication in a study by Atamca ls et al.

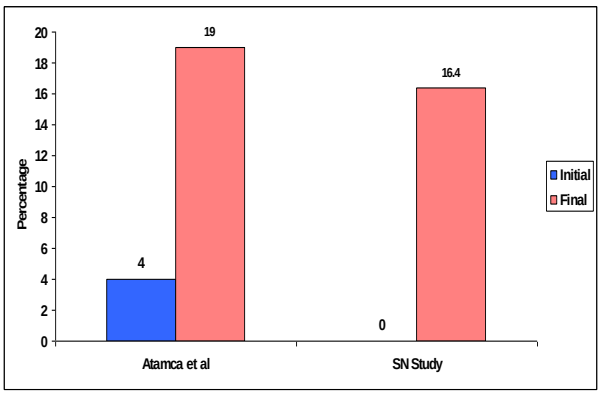
Complications	Atmaca LS et al**	SN study
Cataract	7.2%	18.2%
Tractional RD	10.2%	9.1%
Rubeosis iridis	3%	3.6%
Neovascular Glaucoma	1.7%	1.8%
Phthisis bulbi	0.8%	1.8%

In our study, visual acuity maintained in 58.2% of eyes whereas in Atamca ls et al visual acuity maintained in 43% of eyes. In both the study visual acuity is maintained.

Visual prognosis after the mean follow up:

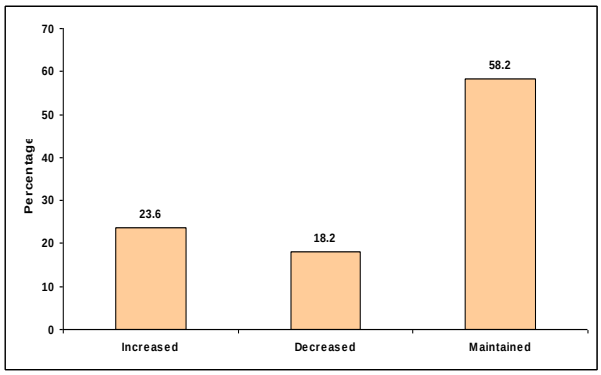


Number of NPL(no perception of light) eyes after the mean follow up:



In our study 16.4% of eyes became NPL whereas in Atamca ls et al 19% of eyes became NPL after the mean follow up.

Visual prognosis after the mean follow up:



Major retinal lesions at final examination:

Retinal Lesion	No. of eyes	%
Vitreous haemorrhage	3	5.5
Vascular sheathing	-	-
NVE	-	-
NVD	-	-

Conclusion

The most important elements in dealing with Eales disease are early diagnosis, periodic follow up and adequate therapy depending upon the stage of the disease.

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