Dissertation on

CLINICAL PROFILE OF RETINAL VASCULITIS AT A TERTIARY EYE CARE CENTRE AND OUTCOMES FOLLOWING MANAGEMENT

Submitted in partial fulfillment of requirements of

M.S. OPHTHALMOLOGY
BRANCH - III

REGIONAL INSTITUTE OF OPHTHALMOLOGY
MADRAS MEDICAL COLLEGE
CHENNAI- 600 008

THE TAMILNADU

DR.M.G.R. MEDICAL UNIVERSITY

CHENNAI

APRIL 2017

INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE, CHENNAI 600 003

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CERTIFICATE OF APPROVAL

To
Dr. Smriti Jain
Post Graduate in M.S. Opthalmology
Regional Institute of Ophalmology and Hospital for Ophtalmology
Madras Medical College
Chennai 600 003

Dear Dr. Smriti Jain,

The Institutional Ethics Committee has considered your request and approved your study titled " **CLINICAL PROFILE OF RETINAL VASCULITIS AT A TERTIARY EYE CARE CENTRE AND OUTCOMES FOLLOWING MANAGEMENT" NO.-20052016**

The following members of Ethics Committee were present in the meeting hold on **03.05.2016** conducted at Madras Medical College, Chennai 3.

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2. Dr.Isaac Christian Moses.,M.D.,Dean,MMC,Ch-3	:Deputy Chairperson
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11.Thiru S.Govindasamy, BA.,BL,High Court,Chennai	: Lawyer
12.Tmt.Arnold Saulina, MA.,MSW.,	:Social Scientist

We approve the proposal to be conducted in its presented form.

The Institutional Ethics Committee expects to be informed about the progress of the study and SAE occurring in the course of the study, any changes in the protocol and patients information/informed consent and asks to be provided a copy of the final report.

Member Secretary – Ethics Committee

MEMBER SECRETARY
INSTITUTIONAL ETHICS COMMITTEE
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CERTIFICATE

This is to certify that this dissertation entitled "**CLINICAL PROFILE OF RETINAL VASCULITIS AT A TERTIARY EYE CARE CENTRE AND OUTCOMES FOLLOWING MANAGEMENT"** is a bonafide record of the research work done by **DR SMRITI JAIN**, post graduate in Regional Institute of Ophthalmology, Madras Medical College and Research Institute, Government General Hospital , Chennai-08, in partial fulfilment of the regulations laid down by The Tamil Nadu Dr. MGR University for the award of M.S. Ophthalmology Branch III, under my guidance and supervision during the academic years 2014-2017.

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DECLARATION BY THE CANDIDATE

I hereby declare that this dissertation entitled, **"CLINICAL PROFILE OF RETINAL VASCULITIS IN A TERTIARY EYE CARE CENTRE AND OUTCOMES FOLLOWING MANAGEMENT"** is a bonafide and genuine research work conducted by me under the guidance of **Prof. Dr. K. Sridhar, M.S., D.O.,** Chief. Department of Vitreo-Retina services, Regional Institute of Ophthalmology and Government Ophthalmic Hospital, Chennai-600008.

Date: **Dr. Smriti Jain**

Place:

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I express my sincere thanks and gratitude to **Prof. Dr. M .K. MURALIDHARAN, M.S.,M.Ch.,** Dean, Madras Medical College for permitting me to conduct this study.

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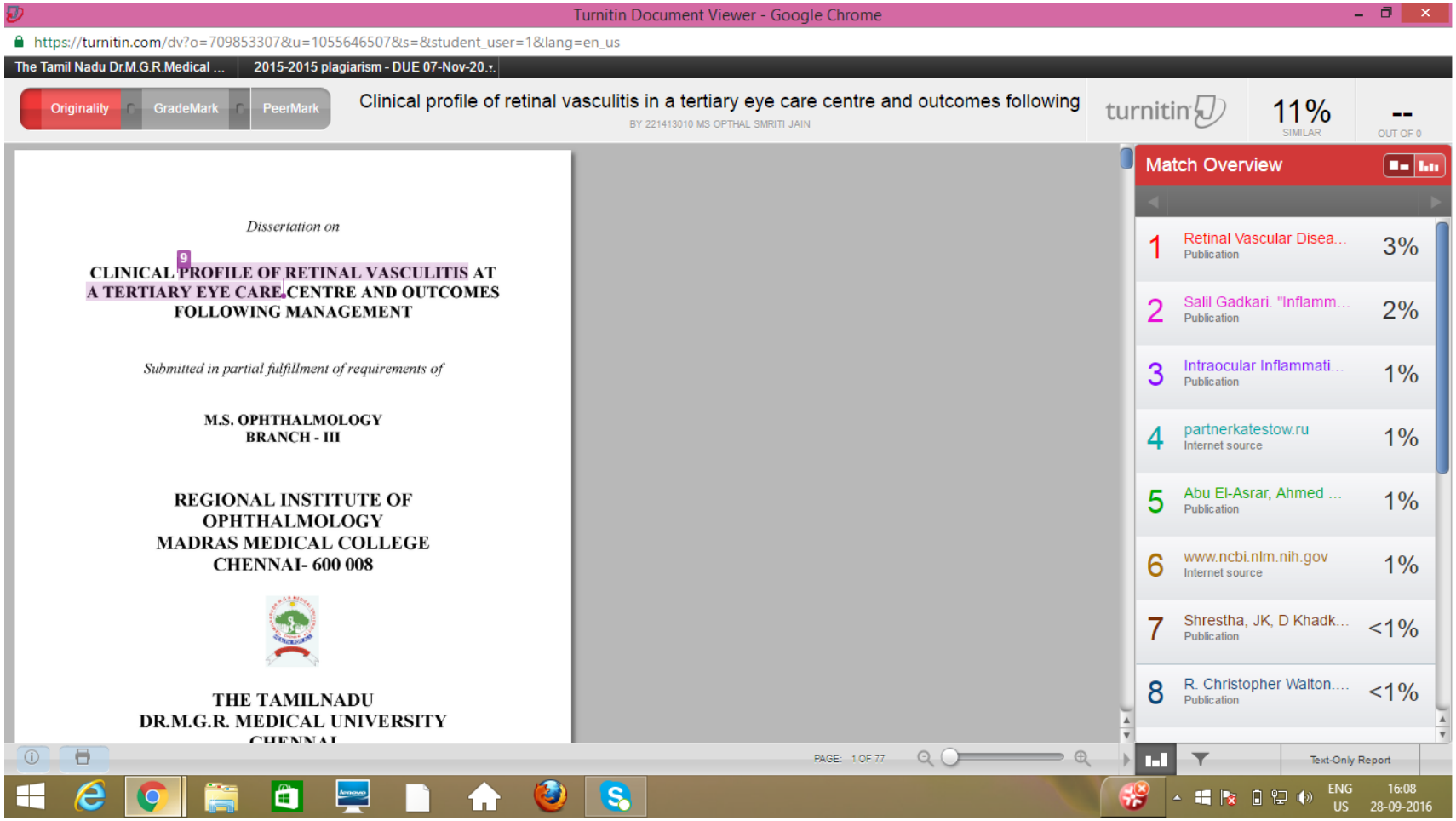
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CONTENTS

INTRODUCTION

Retinal Vasculitis is a group of inflammatory disorders of the eye characterised by retinal vascular inflammation along with intra-ocular inflammation. It preferentially affects the veins, but rarely arteries or arterioles or both veins and arteries can be affected. It is most commonly associated with presence of retinal haemorrhages or vitreous haemorrhages. Recurrence of vitreous haemorrhages is a common feature which might later get complicated by development of retinitis proliferens followed by retinal detachment, complicated cataract and ultimately secondary glaucoma. Ophthalmoscopic examination and fundus fluorescein angiography along with other investigations play a key role in the diagnosis and management of retinal vasculitis.

HISTORICAL ASPECTS

The first attempt to discuss in detail the clinical syndrome of recurrent haemorhages was made by HENRY EALES (1880-82).

PERLS(1873) and ANGELUCCI(1878) studied the histological appearance of periphlebitis.

WADSWORTH (1887) was the first to describe the clinical picture of perivasculitis.

AXENFELD and STOCK (1909-11) drew attention to the etiological significance of tuberculosis.

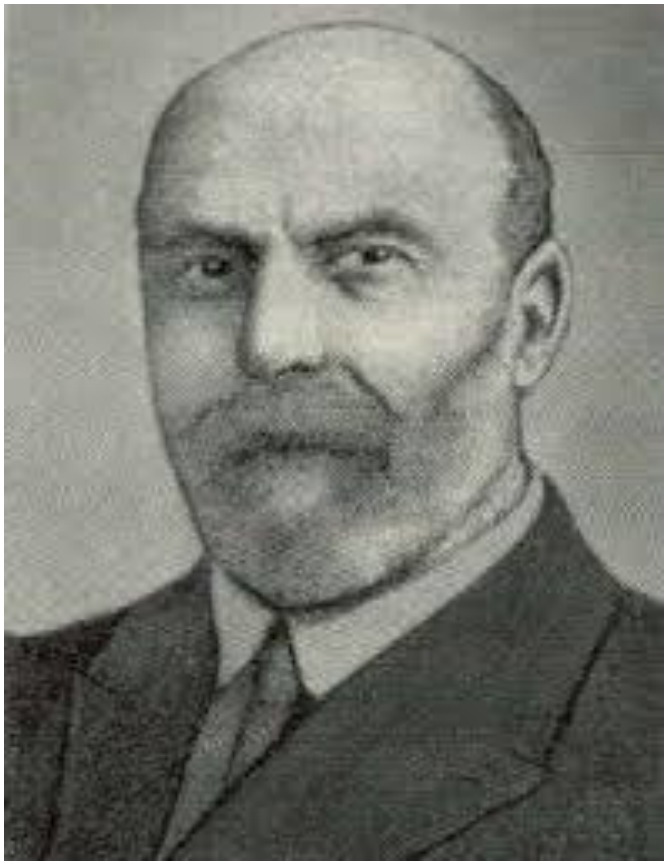


Fig 1-HENRY EALES

CLASSIFICATION OF RETINAL VASCULITIS

DUKE ELDER'S CLASSIFICATION[1]

1. Vasculitis secondary to uveitis
2. Vasculitis secondary to systemic diseases
3. Apparently primary vasculitis

COGAN'S CLASSIFICATION[2]

1. Mild Papillophlebitis
2. Moderate vasculitis which is bilateral and affects arterioles and/or veins
3. Severe vasculitis- Arterioles involved more than veins

RECENT CLASSIFICATION[3]

1. Primary Vasculitis
2. Vasculitis secondary to systemic diseases
3. Vasculitis secondary to ocular diseases

CLINICAL FEATURES OF RETINAL VASCULITIS

SYMPTOMS OF RETINAL VASCULITIS- [3]

The patient can present with the following ocular symptoms-

1. Painless loss of vision
2. Floaters
3. Scotomata due to areas of ischemia
4. Less commonly alteration in color vision and metamorphopsia
5. Can be asymptomatic if lesions are involving the retinal periphery

In association to these systemic manifestations may include-

1. Genital or oral ulcerations
2. Joint pains
3. Chest pain and breathlessness
4. Neurological manifestations
5. Skin ulcerations

SIGNS OF RETINAL VASCULITIS- [3]

1. Sheathing of vessels
2. Obliteration and perivascular edema
3. Retinal hemorrhages
4. Occlusive retinopathy characterised by capillary non perfusion areas
5. Vascular architectural alterations like arteriolar/venule anastomosis and crossing of vessels across horizontal raphe
6. Recurrent vitreous haemorrhage
7. Optic disc edema or optic atrophy
8. Cystoid Macular Edema

PATHOGENESIS

Retinal vasculitis is considered to be an autoimmune phenomena wherein the CD4 T cells are responsible for the cell mediated immunity. These cells are found both within and surrounding the blood vessels. Some studies have demonstrated the role of humoral immunity and immune complex deposition too in the occurrence of retinal vasculitis.

PRIMARY VASCULITIS

The cases of retinal vasculitis which are not associated with any ocular or systemic diseases are termed as idiopathic retinal vasculitis or primary vasculitis. It includes-

1. Eales Disease
2. Idiopathic Retinal Vasculitis, Aneurysms and Neuroretinitis (IRVAN)
3. Frosted Branch Angiitis
4. Scleritis

EALES DISEASE

It was first described in 1880 by Henry Eales who described it in five young men with recurring vitreal and retinal haemorrhages associated with constipation and epistaxis[5].

It is an idiopathic, bilateral, asymmetric condition presenting as an obliterative perivasculitis[6]. It can affect multiple quadrants of the fundus. Mostly it starts at or anterior to the equator and then proceeds towards posterior pole. It mostly affects young males between 20-30 years of age.

Clinical Features

Patients may present with painless diminution of vision and floaters in both eyes but the involvement is asymmetric.

There will be no signs in the anterior chamber and presence of cells and flare mark the beginning of rubeosis iridis.

The following features are typical of Eales Disease-[7]

1. Vascular Sheathing or inflammation (Fig 2)
2. Retinal capillary non-perfusion which may or may not be associated with neovascularisation
3. Vitreous Hemorrhage (Recurrent)
4. Cystoid Macular Edema

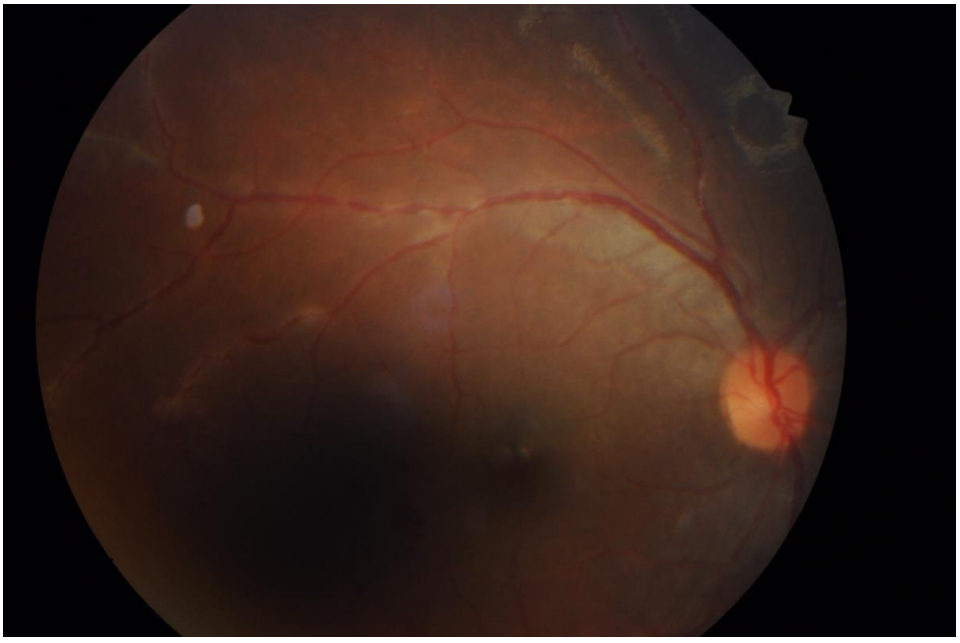


Fig 2- Sheathing of vessels in Eales Disease

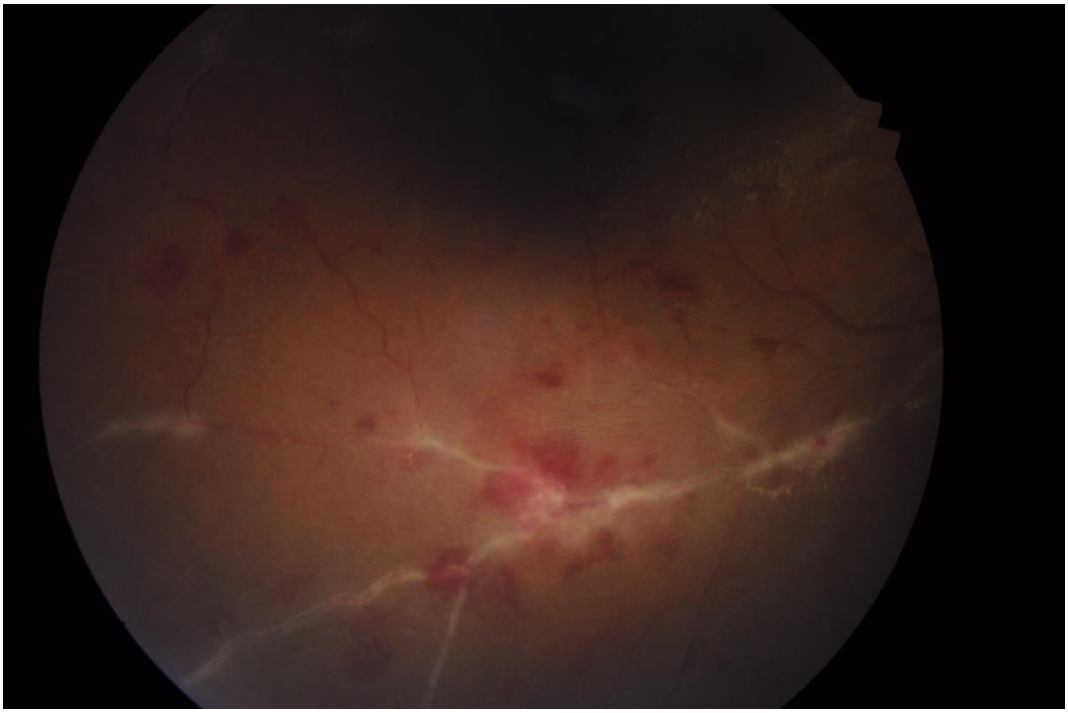


Fig 3- Sheathing with haemorrhages in Eales Disease

1) Inflammation

It is the commonest manifestation of Eales Disease presenting as vascular sheathing especially during early stages of the disease. Sheathing can be continuous or segmental with the presentation ranging from thin white lines to heavy exudates.[3]

The inflammation can lead to keratic precipitates , cells and flare in anterior chamber, vitreous cells and cystoids macular edema.

On fundus fluorescein angiography, the vascular sheathing is seen as hyperfluorescence around the vessel wall and staining of vessel wall (Fig 6)

2) Non Perfusion

Non perfusion can extend from far periphery to the posterior pole with the temporal retina most commonly affected. It is present in all patients with Eales Disease. Ghost vessels are seen in the non perfused areas. At the junction of non- perfused and perfused retina, microaneurysms, arterio-venous shunts and venous beading can be visualised[8]. Intra-retinal haemorrhages (Fig 3) occur in the affected area followed by venous tortuosity and formation of collaterals. Branch retinal vein occlusion is noted in some patients which does not necessarily follow the horizontal midline[9].

3) Neovascularisation

Non perfusion leads to formation of new vessels at the disc or elsewhere in the retina. Rubeosis iridis has also been reported. It often occurs at the junction of perfused and non perfused retina due to over-expression of Vascular Endothelial Growth Factor(VEGF). It is seen as leak on FFA(Fig 7). The commonest sequale of neovascularisation is

vitreous hemorrhage which in due course progresses to formation of fibrous scar tissue ultimately leading to tractional retinal detachment.

ETIOPATHOGENESIS

Multiple pathological, immunological, molecular biological and biochemical studies have indicated the role of HLA antigen, autoimmunity, mycobacterial genome and oxidative stress mechanism in the pathogenesis of vasculitis.

Higher phenotypic association of HLA B5, DR1 and DR4 was seen in patients with retinal vasculitis.[10,11]

Polymerase chain reaction using IS 6110 primers was done to detect the bacterial genome in vitreous fluid, epiretinal membrane and was statistically found in Eales patients.[11]

Biochemical studies show that the protein carbonyl group content increases with severity of Eales disease, indicating the role of oxidative stress in Eales disease.[12]

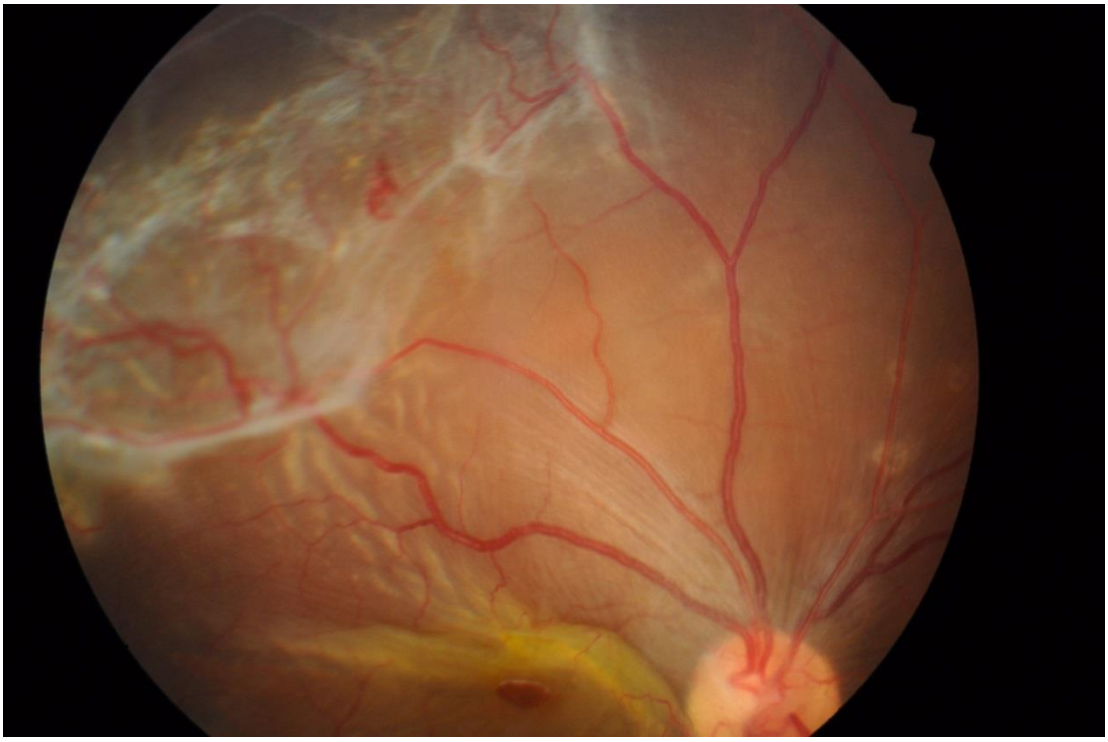


Fig 4 – Tractional detachment in Eales Disease

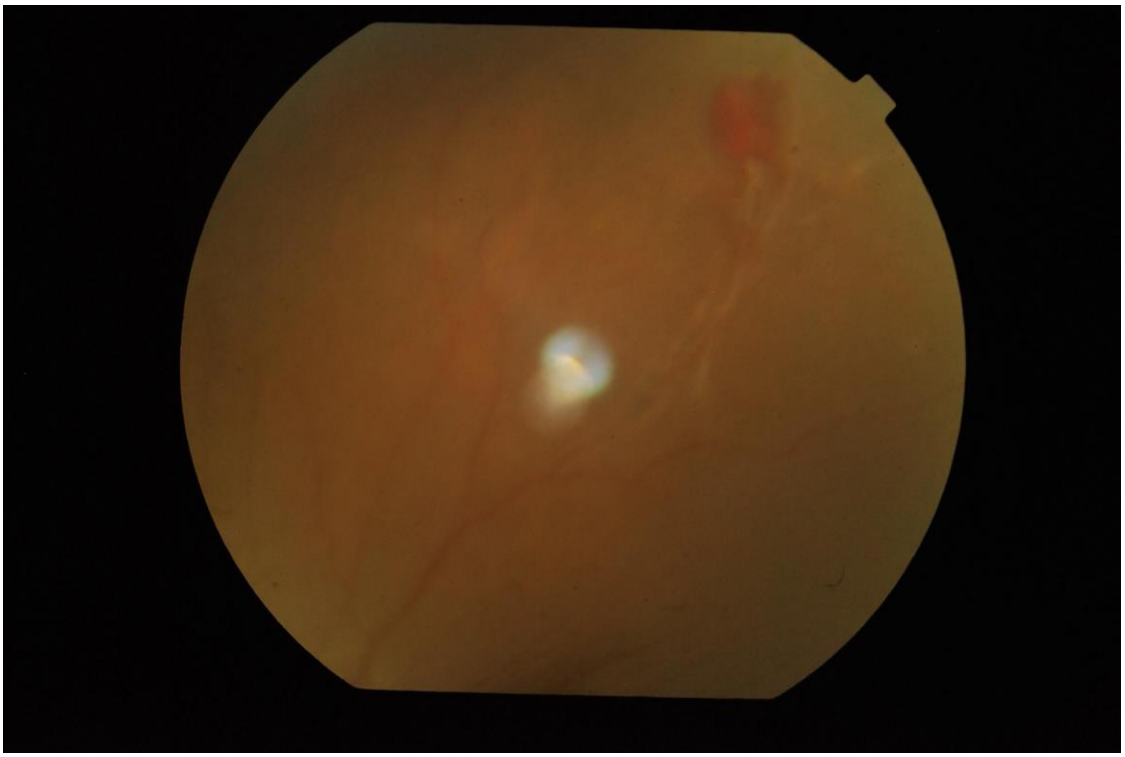


Fig 5- Neovascularisation in Eales Disease

CLASSIFICATION OF EALES DISEASE- [32]

Classification is based on the evolution and progress of the disease.

The classification divided the disease into four stages

Stage 1

Mild periphlebitis of peripheral retinal capillaries

Stage 2

Wide spread periphlebitis of the venous system

Stage 3

New vessel formation and vitreous hemorrhage

Stage 4

End result of multiple hemorrhages –Retinitis proliferans.(Fig 4)

NEWER CLASSIFICATION

This new system of classification divides the disease into central and peripheral, and peripheral type is further subdivided as

Stage 1a

Periphlebitis of small caliber vessels with superficial retinal hemorrhages.

Stage 1b

Periphlebitis of large caliber vessels with superficial hemorrhages.

Stage 2a

Areas of capillary non-perfusion.

Stage 2b

Neovascularization of disc and elsewhere. (Fig 5)

Stage 3a

Fibrovascular Proliferation

Stage 3b

Vitreous hemorrhage

Stage -4a

Traction Or Combined Retinal detachment

Stage-4b

Rubeosis iridis, Neovascular glaucoma, cataract and optic atrophy.

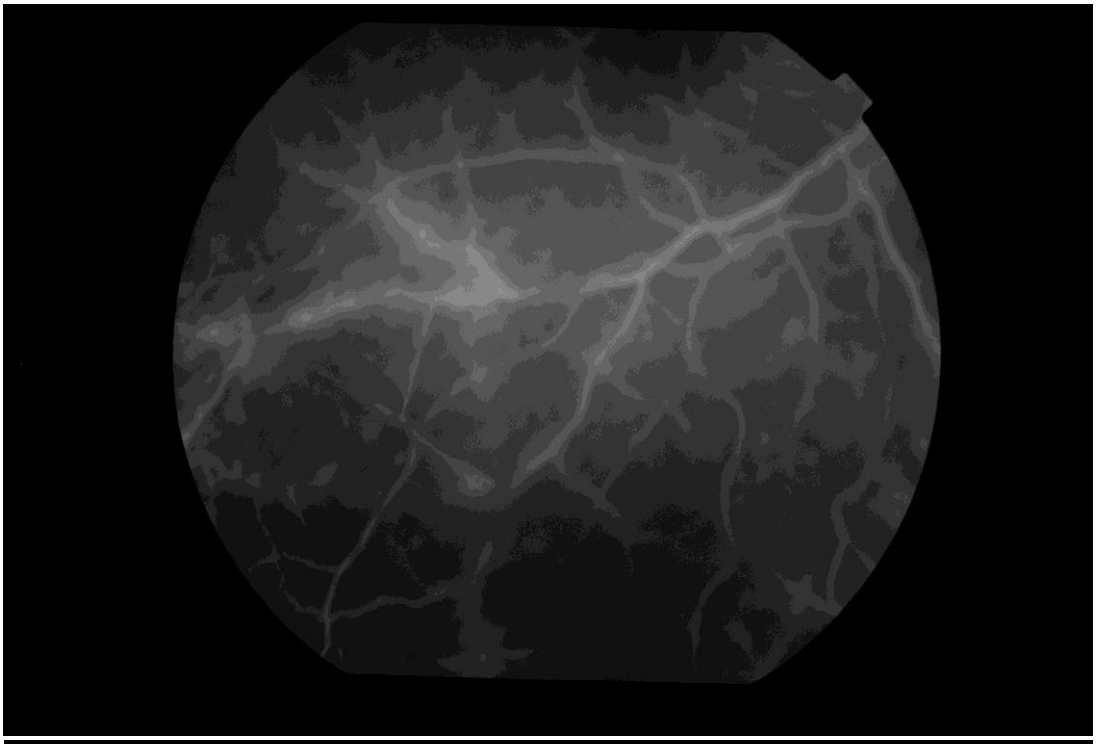


Fig 6- Vessel wall staining in Fundus Fluorescein Angiography in Eales Disease

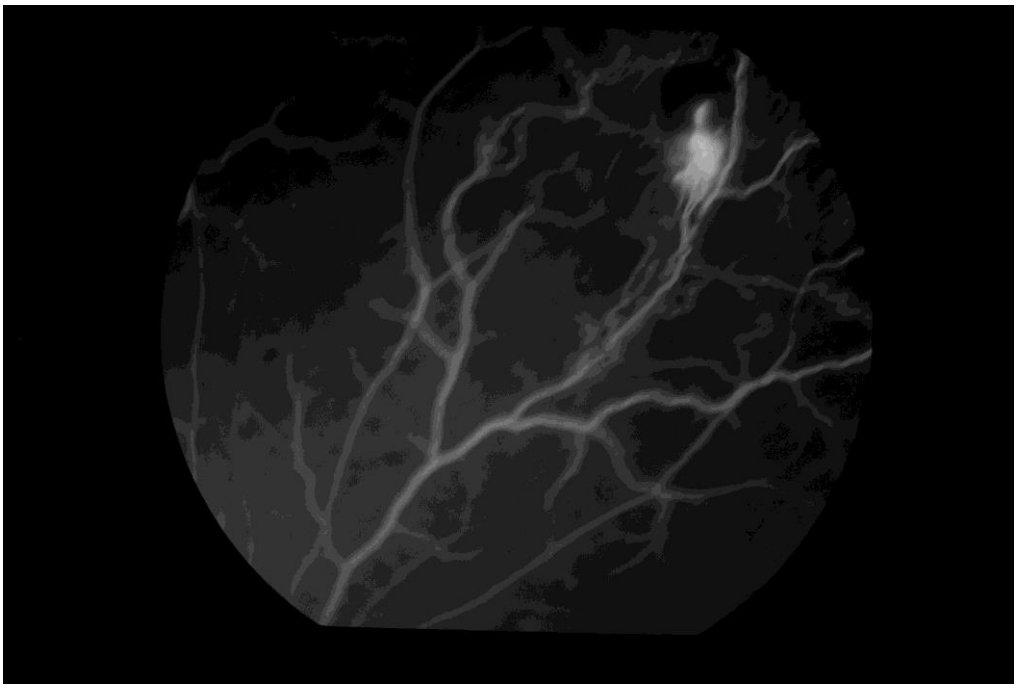


Fig 7- New vessel leak in Fundus Fluorescein Angiography In Eales Disease

DIFFERENTIAL DIAGNOSIS- [17]

- Diabetes Mellitus
- Sickle Cell hemglobinopathies
- Sarcoidosis
- Systemis Lupus Erythematosus
- Branch Retinal Vein Occlusion
- Central Retinal Vein Occlusion
- Coat's Disease
- Pars Planitis
- Dragged Disc Syndrome
- Macular Telengiectasia
- Cytomegalovirus retinitis
- Behcets Disease
- Toxocara
- Toxoplasma
- Syphilis
- Lyme's Disease

OTHER ASSOCIATED SYSTEMIC CONDITIONS[13]-

- Multiple sclerosis
- Cerebellar ataxia
- Myelopathy
- Hemiplegia
- Stroke
- Vestibuloauditary dysfunction
- Internuclear Ophthalmoplegia
- Internal carotid artery aneurysms
- Immunological disorders

MANAGEMENT

1. Medical Treatment

Corticosteroids in the form of oral prednisolone (1mg/kg) for acute inflammatory stage tapered over 6-8 weeks has to be started or we can administer periocular triamnicolone acetonide in the dose of 0.5-1ml (40mg/ml)

In patients with intolerance to steroids, oral methotrexate 12.5 mg/week for 12 weeks can be tried.

Anti tubercular therapy in patients with acute phlebitis with massive infiltration, nodule formation and obliteration of segment of veins has shown good outcomes.

2. Laser Photocoagulation

Laser photocoagulation is effective in stages of non perfusion (Fig 9,10) and neovascularisation. Focal, sectoral or panretinal photocoagulation can be given using laser indirect ophthalmoscope. (Fig 8,11)

Anchoring photocoagulation is applied at the base of tractional band to prevent further combined detachment.[15]

3. Pars Plana Vitrectomy

It is indicated in the following scenarios-

a. Non resolving vitreous haemorrhage(>6-8 weeks)

b. Tractional Retinal Detachment involving posterior pole

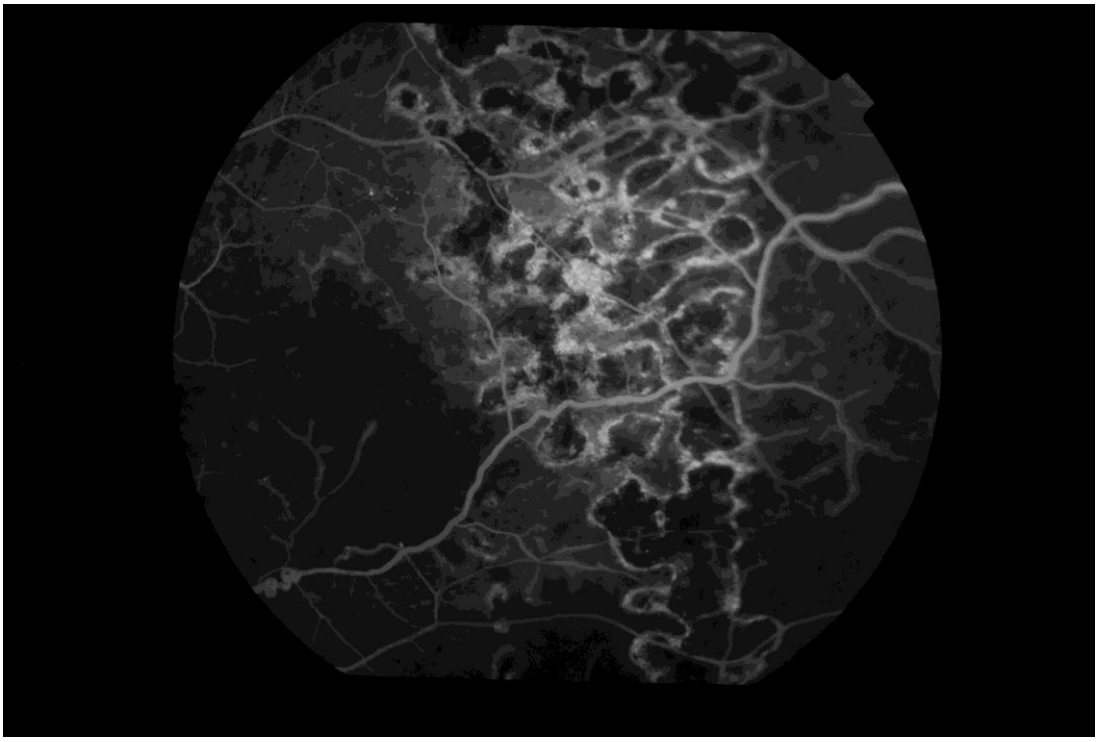


Fig 8-FFA post laser photocoagulation in Eales' Disease

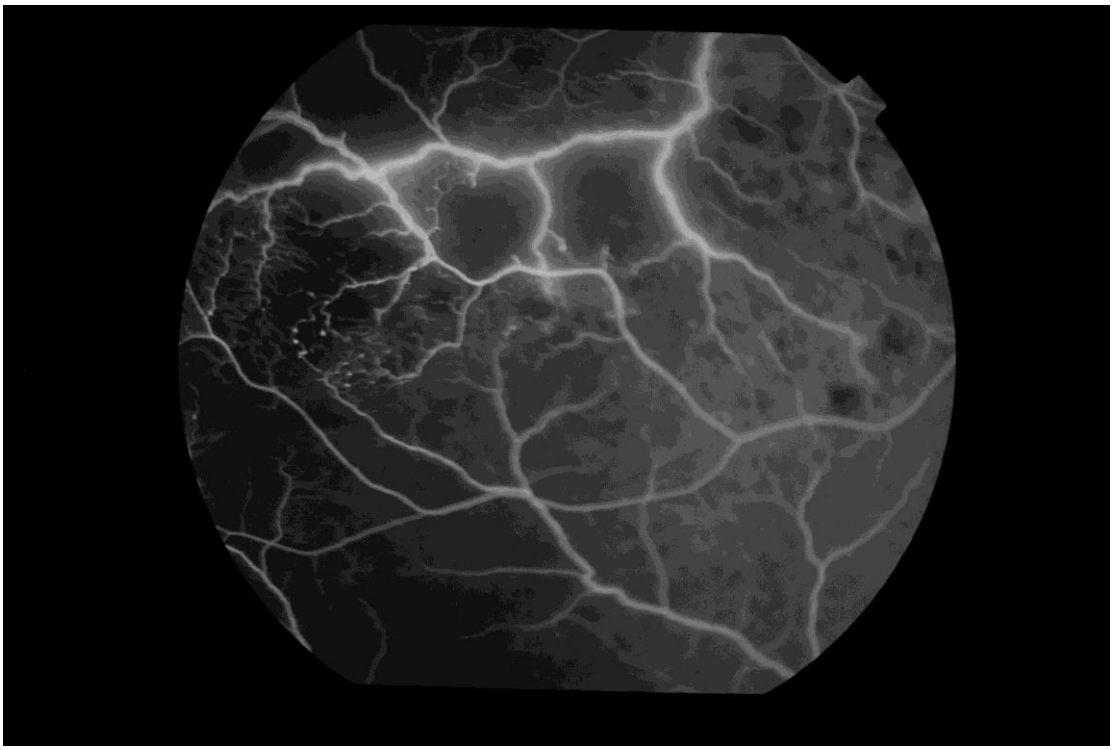


Fig 9-Capillary non perfusion areas with vessel wall staining in Eales' Disease

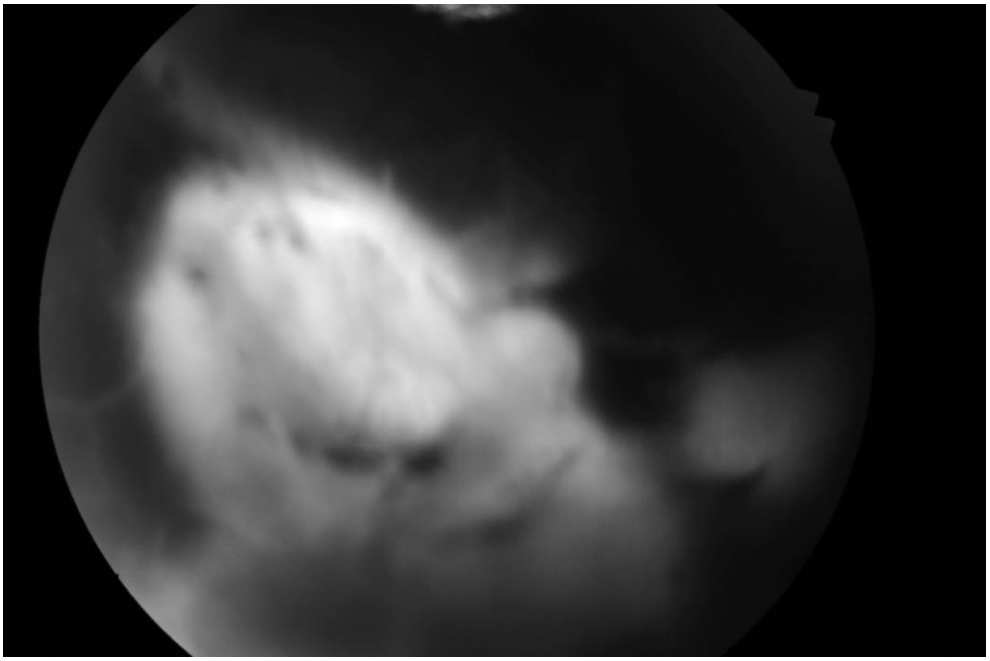


Fig 10- Areas of capillary non- perfusion and leaks on FFA in Eales Disease

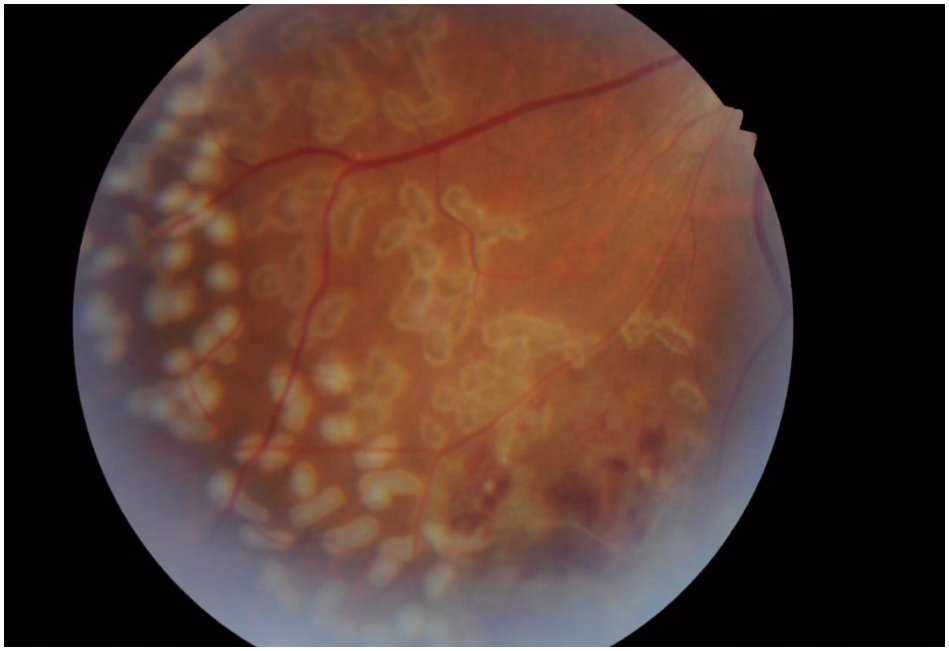


Fig 11-Fundus photo after application of laser photocoagulation to areas of leak and non-perfusion

c. Multiple vitreous membranes
d. Combined tractional and rhegmatogenous retinal detachment
e. Media Opacities

VISUAL PROGNOSIS

Inspite of extensive non perfusion, the posterior pole is spared in most of the cases preserving a good visual acuity. In cases of vitreous hemorrhage, within few weeks to months the hemorrhage settles inferiorly and gets re-absorbed restoring normal vision. Progression to neovascularisation, non resolving vitreous hemorrhage, retinal detachment and neovascular glaucoma can lead to permanent visual impairment.

IDIOPATHIC RETINAL VASCULITIS, ANEURYSMS AND NEURORETINITIS (IRVAN SYNDROME)

It is a rare condition seen in younger individuals. It is characterised by the presence of multiple saccular and fusiform aneurysms of the larger arterioles along with non perfusion in the periphery with neovascularisation, anterior uveitis and optic nerve head swelling.

The aneurysms tend to regress and appear at another location within a short period of time.

The disease has been reported in pregnant females with a hypercoagulable status.[16]

Laser photocoagulation in the areas of non perfusion is the current treatment of choice.

FROSTED BRANCH ANGIITIS

It is usually found in younger individuals who present with a sudden diminution of vision. It is characterised by the presence of florid translucent retinal perivascular sheathing involving venules more than the arterioles, retinal edema and intermediate uveitis. Most of the cases are bilateral with males and females showing equal affection.

Causes of frosted branch angiitis-

- Idiopathic
- Tuberculosis
- Syphilis
- Herpes Simplex type 2

- Ocular toxoplasmosis
- Influenza A exposure
- CMV retinitis
- HIV infection
- Aseptic meningitis
- Lymphoma
- Leukemia
- Saroidosis
- Multiple sclerosis
- Pars Planitis
- Systemic Lupus erythmatosus

Patients are responsive to steroids and photocoagulation with good visual outcome.[17]

SCLERITIS

The vasculature near the scleritis is integrally involved in inflammatory and pathological process. The inflammatory reaction in anterior scleritis with spill over leading to anterior or intermediate uveitis.

Posterior scleritis patients present with severe ocular pain with minimal anterior segment changes, minimal vitreous changes, choroidal folds and retinal thickening. Ultrasound examination helps in the diagnosis. Corticosteroid therapy helps to control the disease. But recurrences do occur. Whether Posterior scleritis is associated with a systemic disorder must be ruled out. Rheumatological work up should be ordered. Many patients respond to corticosteroids but relapse may occur on tapering the dose.

VASCULITIS SECONDARY TO SYSTEMIC DISEASE

1. Secondary to Infectious Disorders

- Bacterial disorders (Tuberculosis, Syphilis, Brucellosis, Lyme Disease)
- Viral disorders (Cytomegalovirus, Herpes simplex, Varicella Zoster, Acquired Immunodeficiency syndrome, Human T cell Lymphoma virus)
- Parasitic disorders (Toxoplasmosis, Toxocara (Fig 27))

2. Secondary to Neurological Disorders

- Multiple Sclerosis
- Micro-angiopathy of brain, retina and cochlea (Susac Syndrome)

3. Secondary to Malignancy

- Paraneoplastic syndromes
- Ocular lymphoma
- Acute Leukemia

4. Secondary to Systemic Inflammatory Disease-

- Behcet’s Diseae
- Sarcoidosis

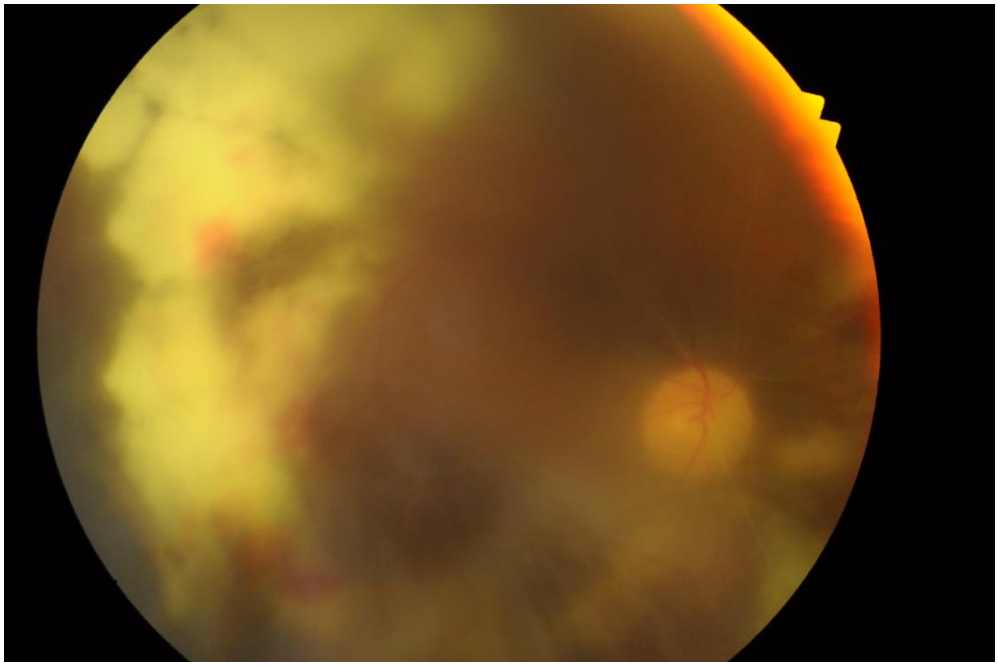


Fig 12-Vasculitis secondary to Acquired Immunodeficiency Syndrome (OD)

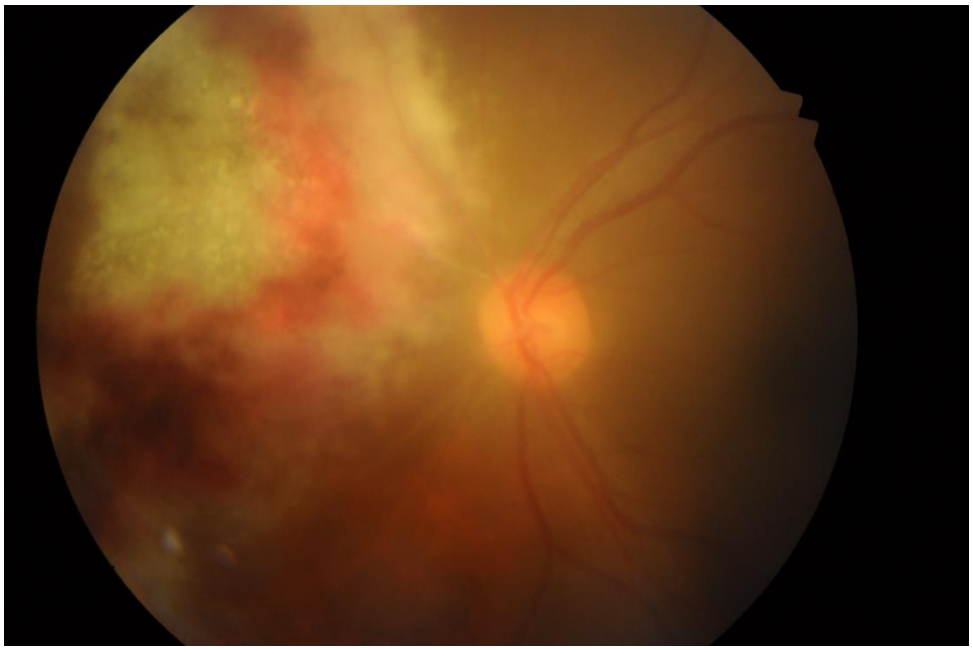


Fig 13- Vasculitis secondary to Acquired Immunodeficiency Syndrome (OS)

- Systemic Lupus Erythematosus (Fig 14,15,16,17)
- Wegener's Granulomatosis
- Polyarteritis nodosa
- Churg- Strauss Syndrome
- Polyarteritis Nodosa
- Relapsing Polychondritis
- Rheumatoid Arthritis
- HLA-B 27 associated uveitis (Fig 24,25)
- Crohn' Disease
- Post vaccination
- Takayasu's Arteritis
- Dermatomyositis
- Buerger's Disease
- Polymyositis

Retinal Vasculitis Secondary to Tuberculosis- (Fig 18,19,20,21)

Vasculitis in tuberculosis infection is very common. It may be either due to active systemic infection or due to type IV hypersensitivity reaction to tubercular antigen protein.[18] The inflammation leads to periphlebitis and sheathing of vessels with or without haemorrhages. The

vessels might get occluded leading to ischemia after few days. It can involve any quadrant of retina with presence of sheathing , haemorrhages and exudates. The ischemia will further cause formation of new vessels, vitreous hemorrhage and tractional retinal detachment. Chest radiograph and mantoux test can be employed for preliminary testing. Quantiferon TB gold test is more specific.[19] Once the cause is determined, patient can be started on anti- tuberculous therapy and oral corticosteroids with regular follow up.

Vasculitis secondary to Systemic Lupus Erythematosus- (Fig 14,15,16,17)

It's an immune complex disorder, which affects the eye with severe intraocular complication .Ocular involvement is common in the form of dryness.

In 7.5% of the patients with SLE retinal vasculopathy is mostly as microangiopathic changes with cotton wool spots. Cotton wool spots and retinal hemorrhages correlate with disease activity and are of negative prognostic sign. SLE patients with anti-phospholipid syndrome have increased risk for retinal vasoocclusive disease.[20]

The pathogenesis of SLE are complex and partially understood, include triggering agent such as infection might lead to activation of autoreactive T and B cells[21.] Treatment of microangiopathic changes should be chosen in the context of systemic disease. Remission is induced with intravenous cyclophosphamide and maintained with other immunosuppresants like azathioprine, Mycophenolate Mofetil and Rituximab are emerging therapies for SLE.

HIV Infection- (Fig 12,13)

The vasculitis in HIV infection can be caused by interaction between viral antigen and antibodies which are in the blood circulation. Higher the extent of immunosuppresion, more will be the chances of developing retinal vasculature involvement. The expression of p24 antigen over the surface of the virus is associated with advanced stage of disease. The patient can have lot of superficial and deep haemorrhages along with exudates and vascular sheathing. Finally it may all lead to vascular occlusion. Few soft exudates may be noted. Central retinal vein occlusion has also been reported.

Cytomegalovirus Infection- (Fig 22,23)

CMV is double stranded DNA virus from Herpes virus family. The involvement of retina with CMV infection mostly occurs as an opportunistic infection in immunocompromised patients or a congenital infection in children. Previously it was mre common with patient who have undergone organ transplant but now Acquired Immunodeficiency Syndrome is the most common cause when the CD 4 T cell count goes below 50 cells per mirolitre.

The virus reaches the eye through the bloodstream. It can present as small, white infiltrates in the retina arund the vessels with flame shaped haemorrhages which are scattered throughout along with sheathing of vessels. It has another granular variant which has less haemorrhages and more of retinal pigment epithium stippling. It starts in the periphery and spreads centrally so patients might have no visual complaints initially. Anterior chamber involvement is uncommon. All patients with a CD 4 count less than 50 should be screened for CMV regularly. Once diagnosed, the patients can be started on induction doses of intravenous ganciclovir followed by maintenance dose. Other options are Foscarnet, Cidofovir , Fomivirsen or Ganciclovir implant.

TOXOPLASMOSIS- (Fig 26)

The causative organism is Toxoplasma Gondii. Man is accidental host. Ocular involvement is very common presenting with characteristic chorio-retinitis foci with associated vitritis. Anterior chamber reaction may also occur. Old lesions may appear as an atrophic scar. Adjacent to the scar reactivation of lesion may occur in many cases. Retinal vasculitis may be noticed in some cases presenting as sheathing of vessels. Few cases have been reported which had only vasculitis as a presenting feature in toxoplasmosis instead of the typical activeretino-choroiditis foci. So, toxoplasmosis should be ruled out while evaluating a retinal vasculitis case even if typical features are absent. It can be seen in immunocompromised patients. Patients can be diagnosed by detecting toxoplasma antibodies in the serum. Patient should be started on antibiotics with oral corticosteroids. The associated anterior uveitis can be treated with topical steroids and cycloplegics.

VASCULITIS IN MULTIPLE SCLEROSIS

Multiple sclerosis is a chronic inflammatory demyelinating disease of central nervous system mostly affecting young adults. Ocular manifestations include Optic neuritis, Extra ocular muscle disturbance and intraocular inflammation.

It was reported by Rucker that 20% of patient with Multiple sclerosis have vascular sheathing as their association.[22]

Retinal vasculitis in multiple sclerosis patient is exclusively periphlebitis, retinal venous sheathing. It encompasses both the active and chronic form. The chronic form of sheathing appears typically as dense white strips over several branches of vascular tree.

Activity of periphlebitis is not correlated with Optic neuritis, systemic exacerbation or severity of disease.[23] It can lead to occlusive vasculitis with subsequent formation of neovascularisation.

BEHCET'S DISEASE

Ocular Behcet's is characterized by nongranulomatous uveitis with retinal vasculitis. The reported frequency of ocular manifestation is 83-95% in males and 67-73% in females.

Retinal vasculitis is of occlusive nature characterized by retinal edema, yellowish white exudates and hemorrhages. Behcet's disease is the only systemic vasculitis that affects both arteries and veins and this feature is pathognomic of Behcet's vasculitis.[24] Occlusive nature of vasculitis will lead to neovascularistion.

NECROTIZING VASCULITIS

Disorder with pathogenesis being linked to antineutrophilic cytoplasmic antibodies plus environmental and genetic factors.

POLYARTERITS NODOSA AND MICROSCOPIC POLYANGIITIS

SYSTEMIC COURSE OF PAN

- Neuropathy
- Nephropathy
- Cutaneous ulcers
- Gastrointestinal thrombosis and infarction
- Musculoskeletal pain
- Coronary arteritis
- CNS involvement

OCULAR MANIFESTATION OF PAN

- Microangiopathy with cotton wool patches
- Serous Retinal Detachment
- Retinal Vasculitis

- Artery occlusion
- Ischemic Optic Neuropathy

CHURG STRAUSS SYNDROME

- ACR criteria for diagnosis of Churg Strauss Syndrome
- Asthma, adult onset
- Eosinopilia >10% in the blood
- Sinusitis
- Pulmonary infiltrates
- Neuropathy
- Biopsy with vasculitis and Eosinophils

OCULAR MANIFESTATION

- Retinal vascular disorders like
- Ischemic Optic Neuropathy
- CRAO
- BRAO
- CRVO
- Retinal Vasculitis

WEGENER GRANULOMATOSIS

It’s a triad of Systemic necrotizing vasculitis(87%), necrotizing granulomatous involvement of respiratory tract and necrotizing glomerulonephritis.

ACR criteria for diagnosis

2 of 4

- Inflammation in the nose and mouth
- Abnormal urinary sediment
- Abnormal chest x-ray
- Granulomatous inflammation of arteries

OCULAR MANIFESTATION

- Orbit (13%)
- Eyelid and Nasolacrimal duct (13%)
- Episcleritis and Scleritis (11%)
- Keratitis (8%)
- Optic neuropathy (6%)
- Conjunctivitis (4%)
- Retinal vasculopathy

VASCULITIS DUE TO OCULAR DISEASES

- Pars planitis
- Choroiditis
- Posterior uveitis
- Behcet's retina sine systemic disease
- Birdshot retino choroidopathy
- Acute multifocal hemorrhagic retinal vasculitis

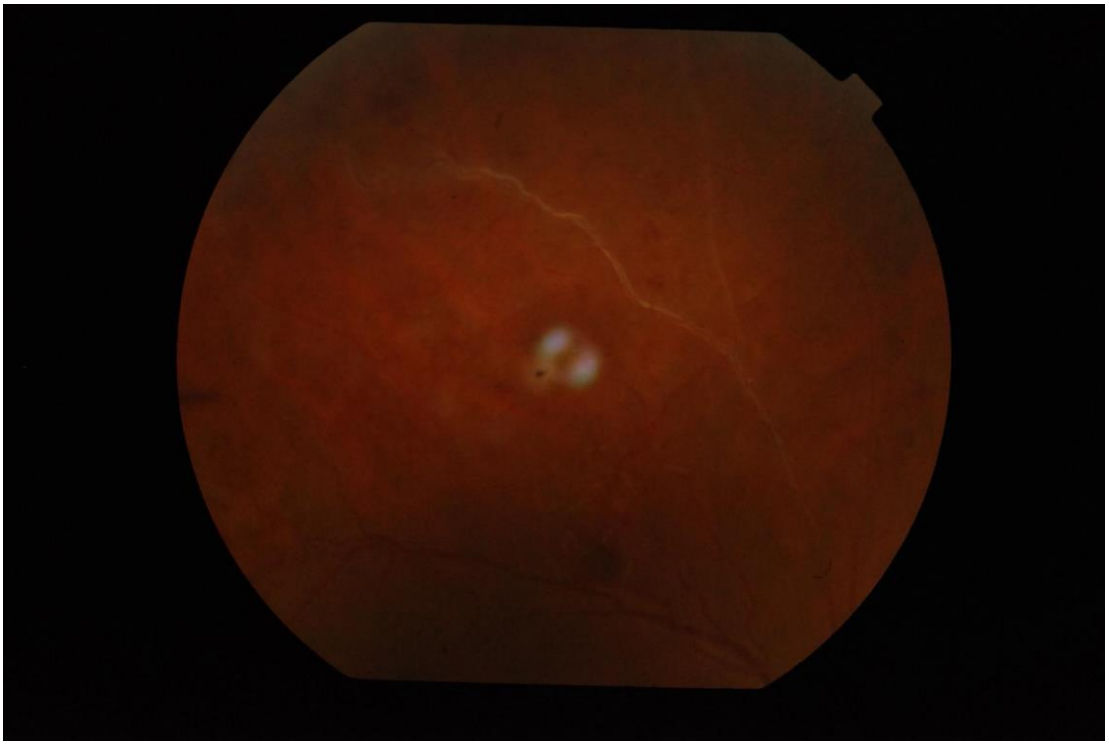


Fig 14-Vasculitis in Systemic Lupus Erythematosus (OD)

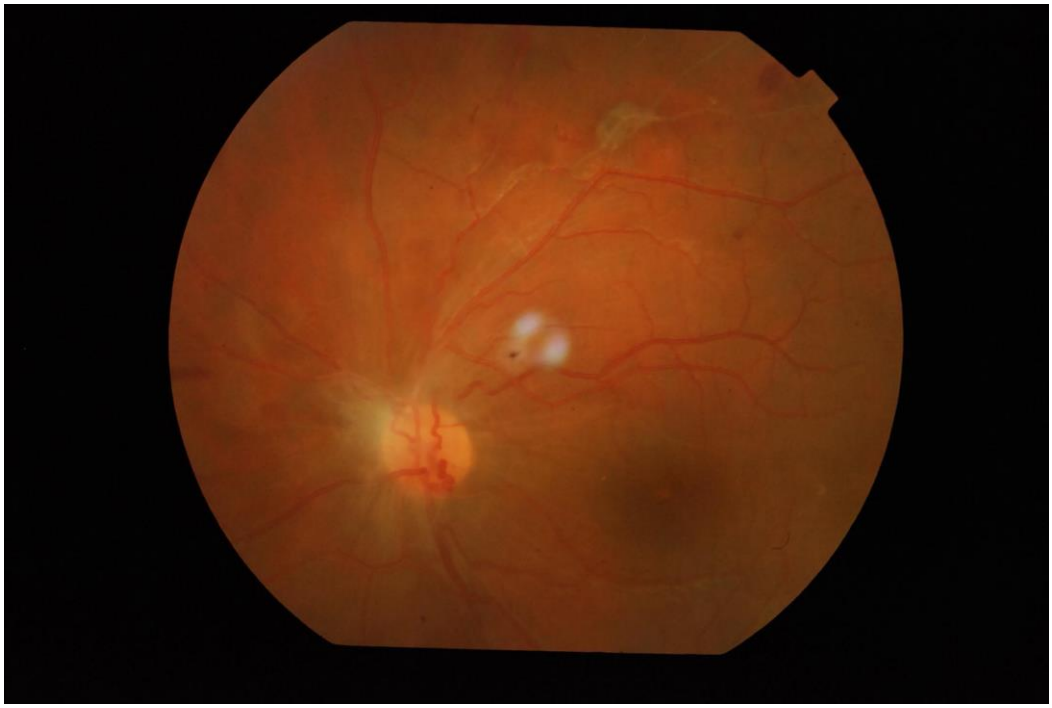


Fig 15- Vasculitis in Systemic Lupus Erythematosus (OS)

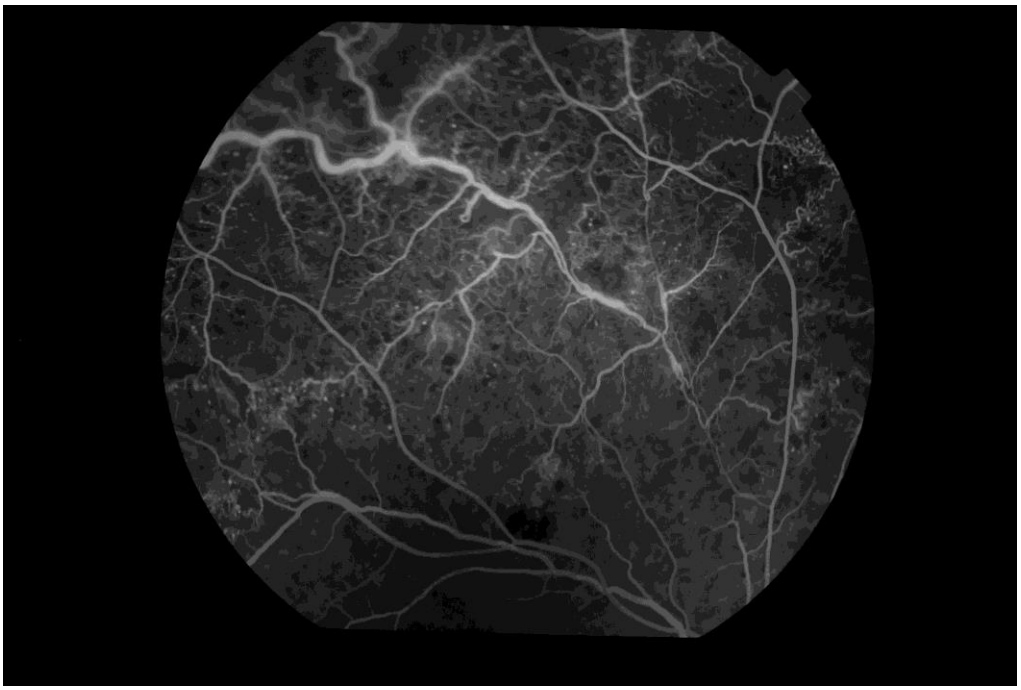


Fig 16-FFA showing staining of vessel walls in SLE

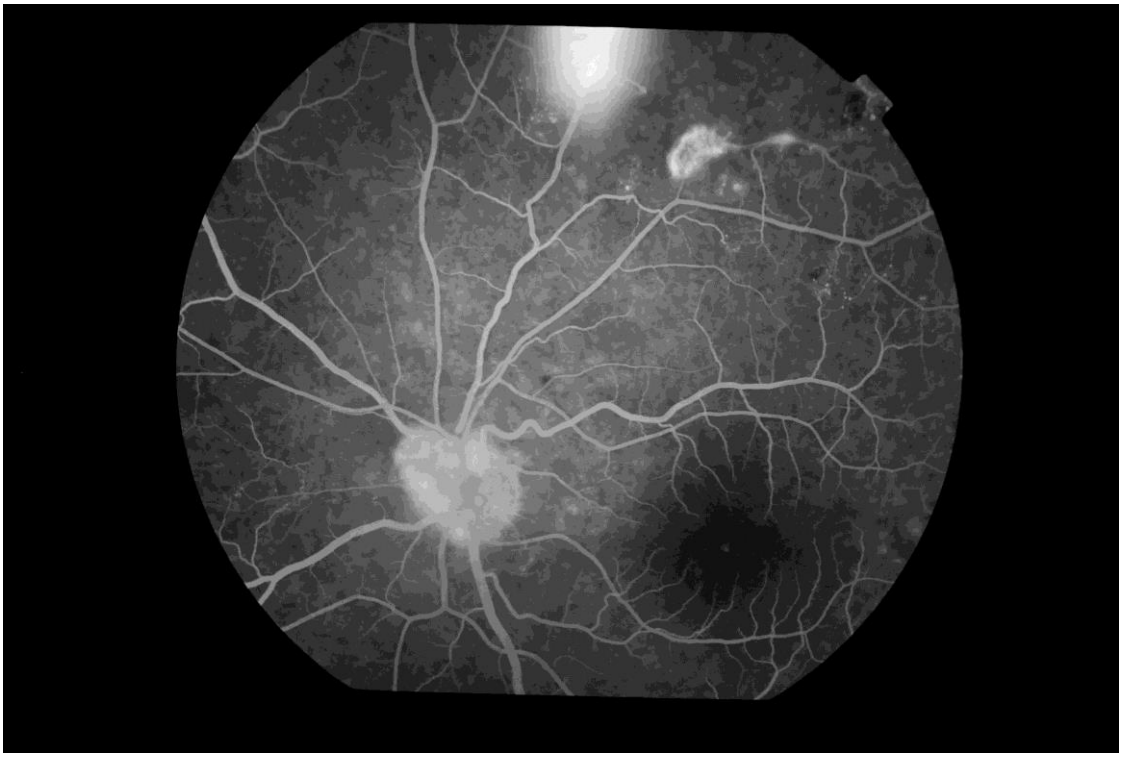


Fig 17-FFA showing leaks from NVE in SLE Vasculitis

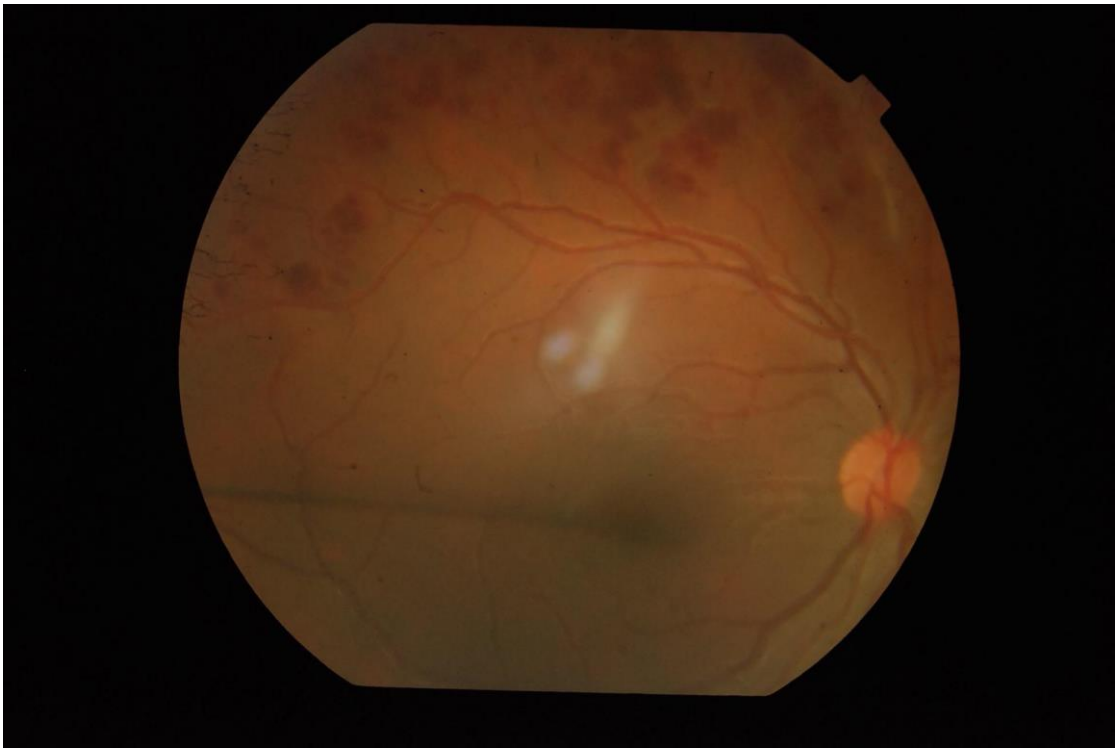


Fig 18-Sheathing with hemorhhages in vasculitis secondary to tuberculosis (OD)

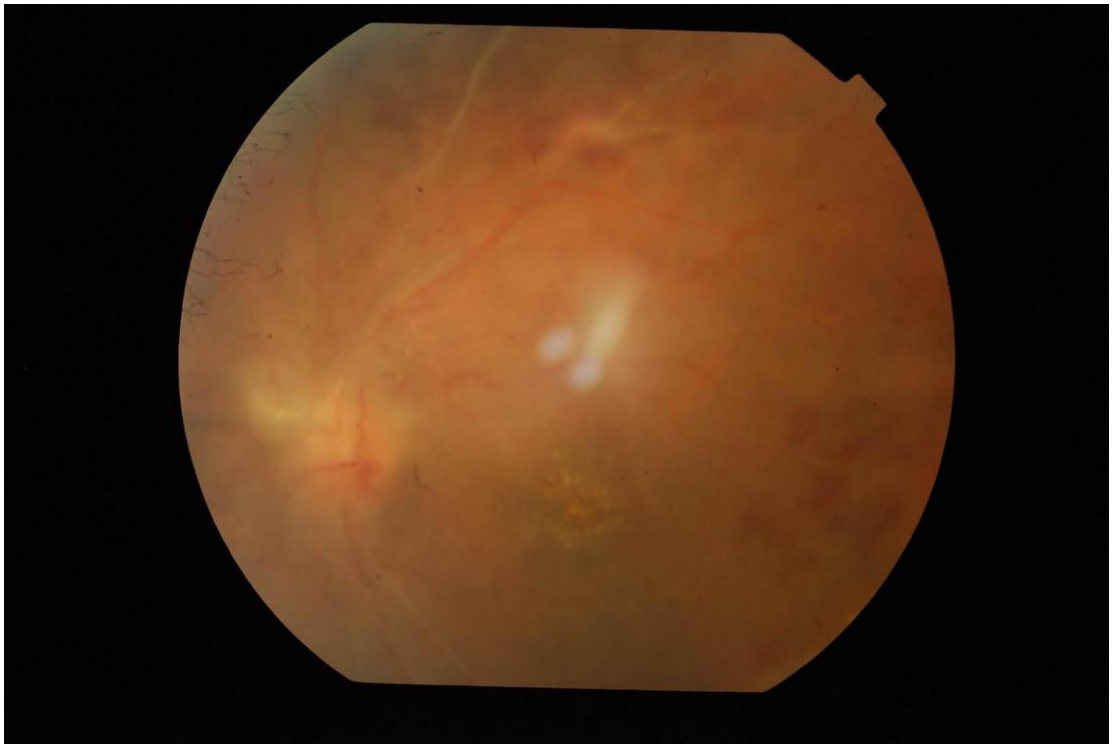


Fig 19-Sheathing with haemorrhages in vasculitis secondary to tuberculosis (OS)

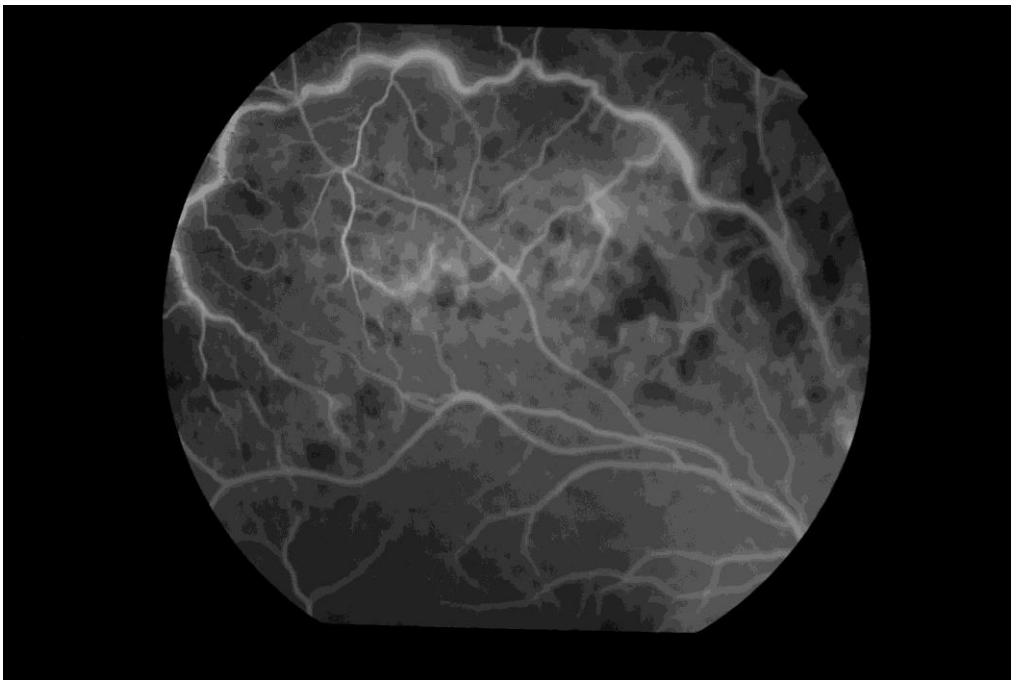


Fig 20-Vessel wall staining with blocked fluorescence due to haemorrhages on FFA(OD)

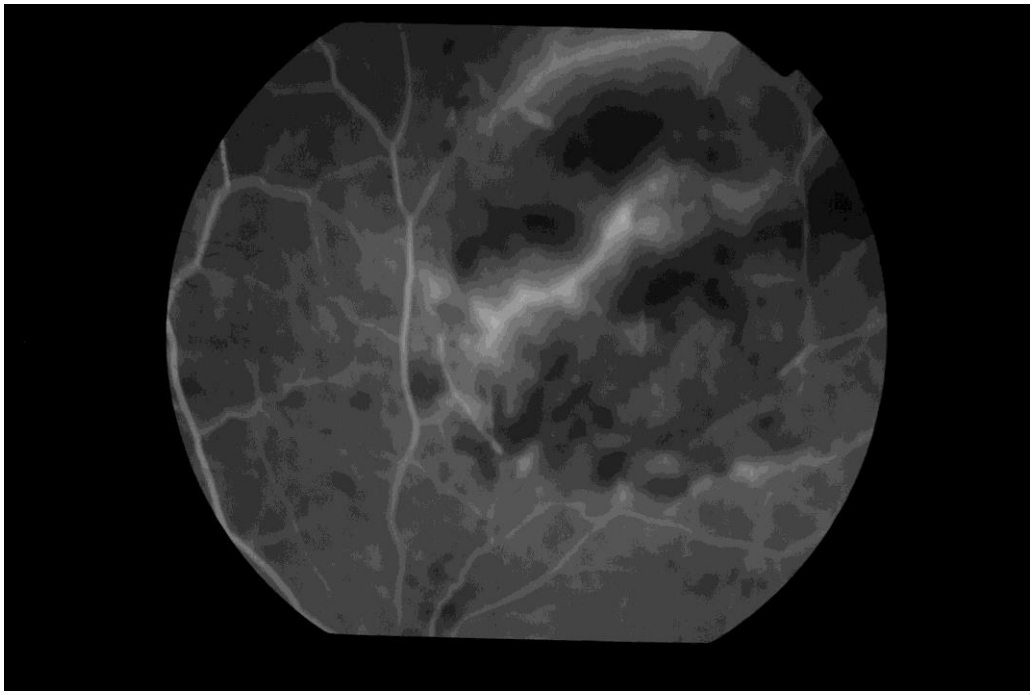


Fig 21-Vessel wall staining with blocked fluorescence due to haemorrhages on FFA(OS)

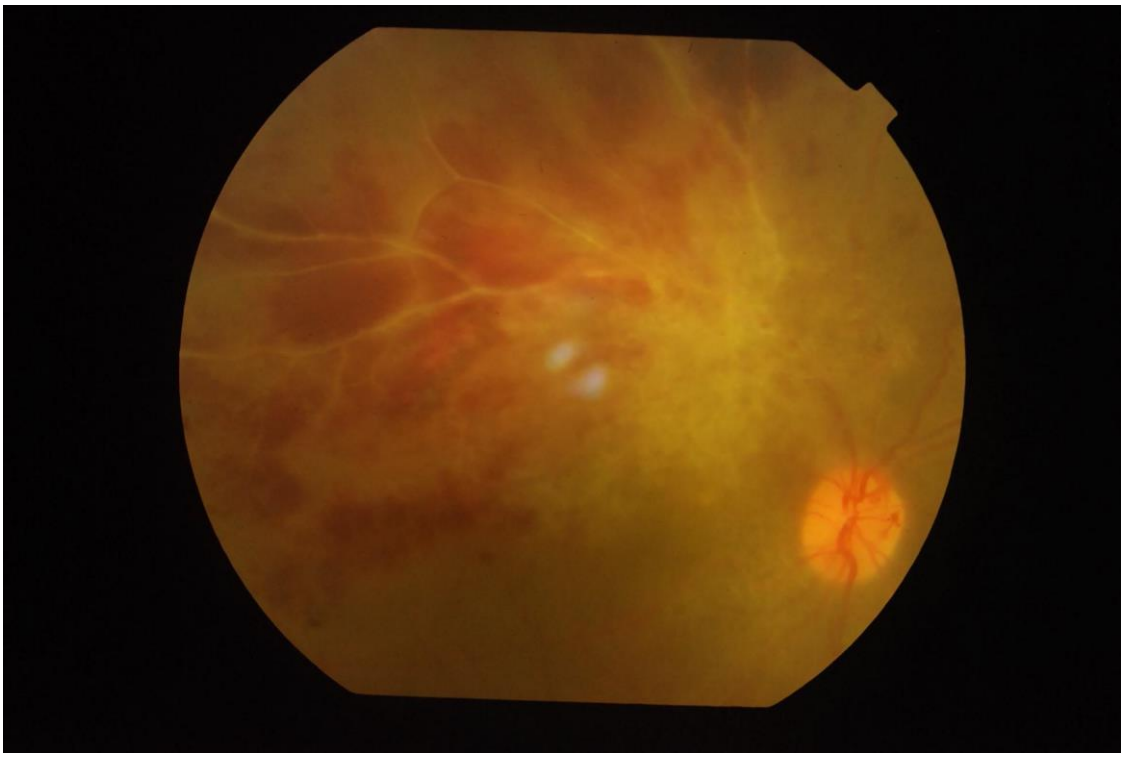


Fig 22-Vasculitis secondary to CMV infection (OD)

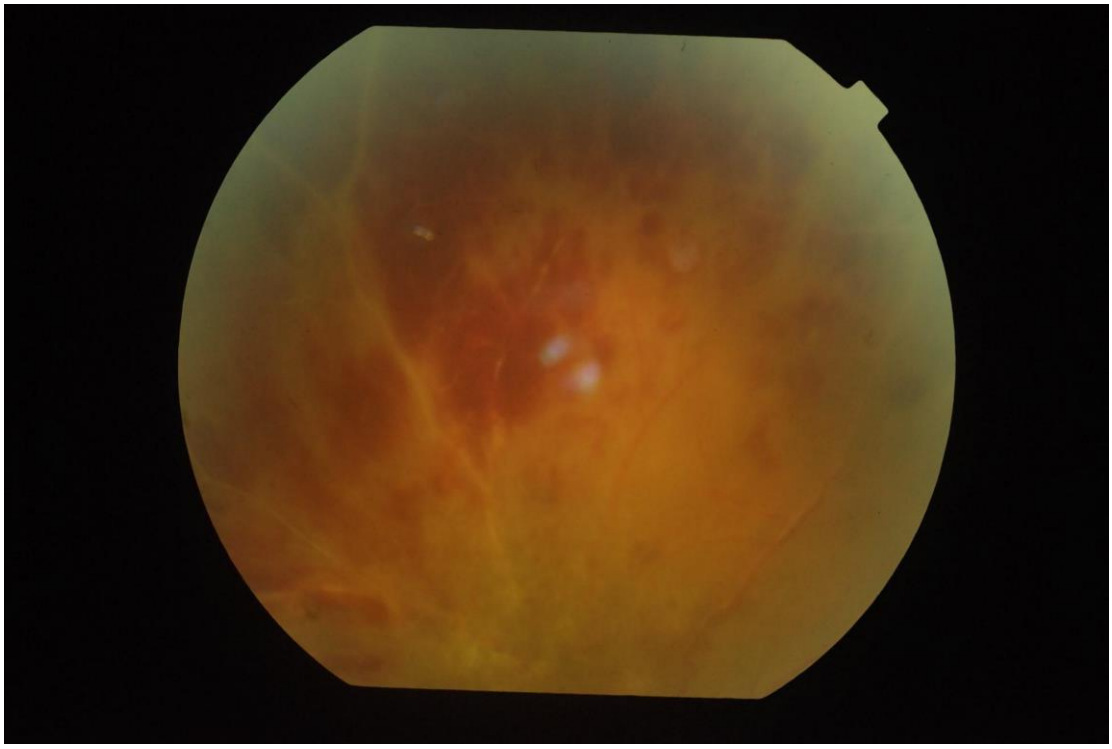


Fig 23- Vasculitis secondary to CMV infection (OS)

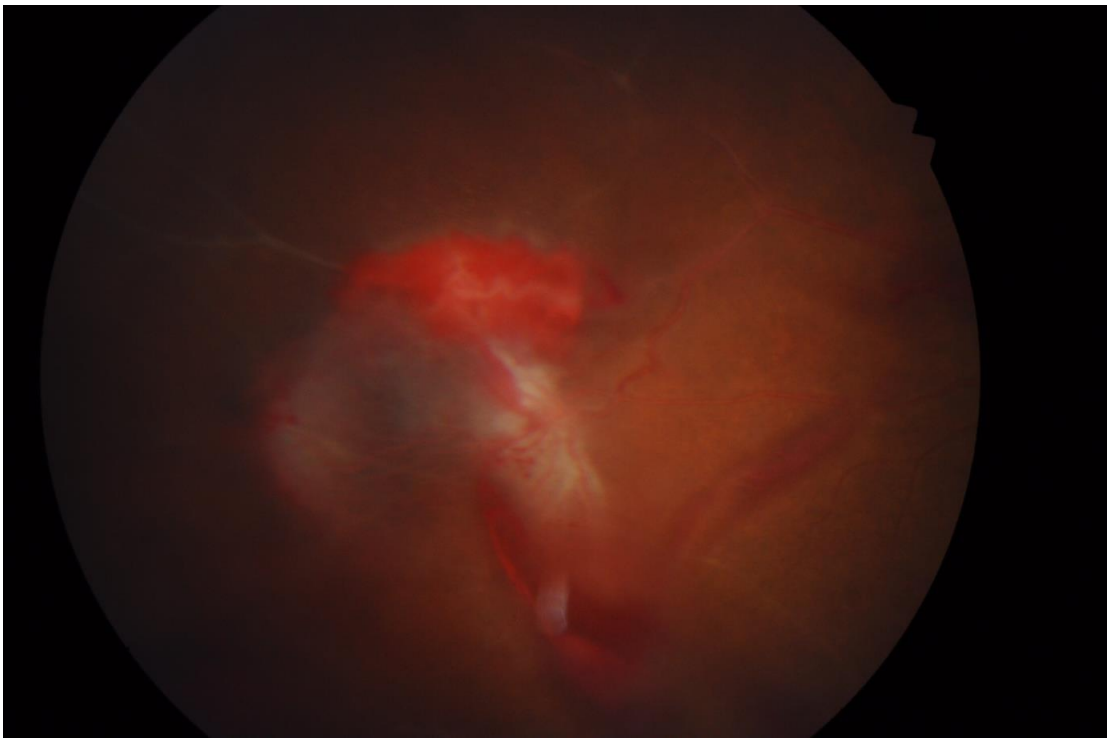


Fig 24-Vasculitis Secondary to Ankylosing Spondylitis

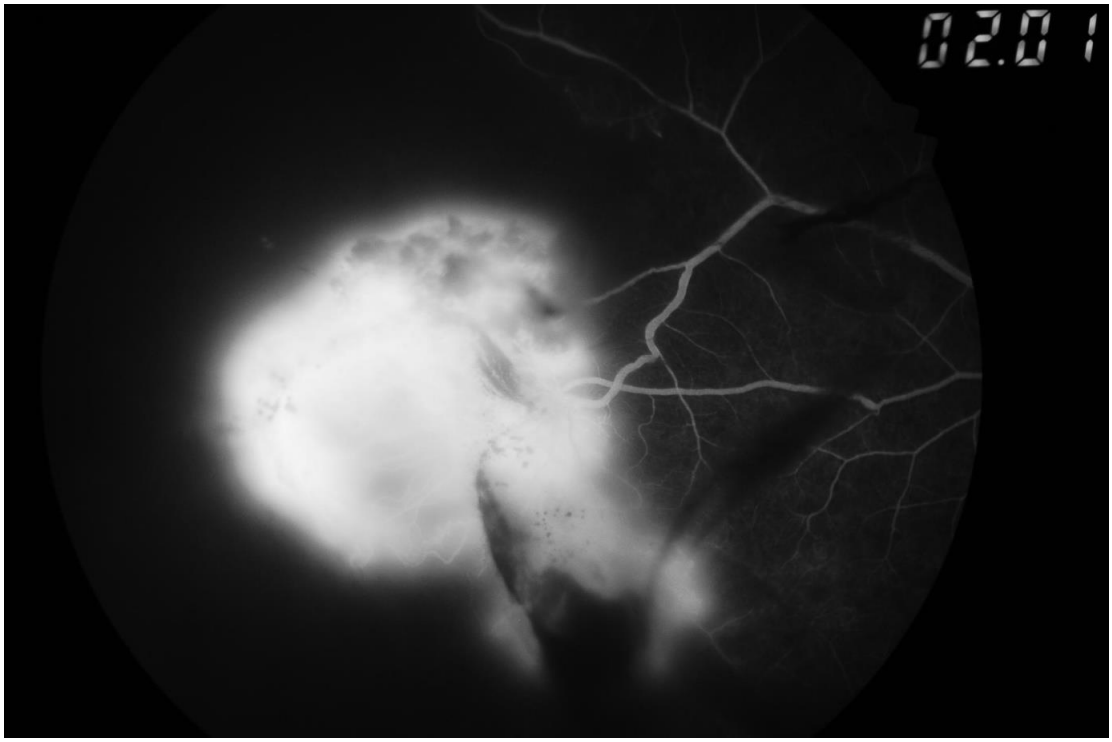


Fig 25-FFA showing corresponding areas of leakage from NVE and CNP areas

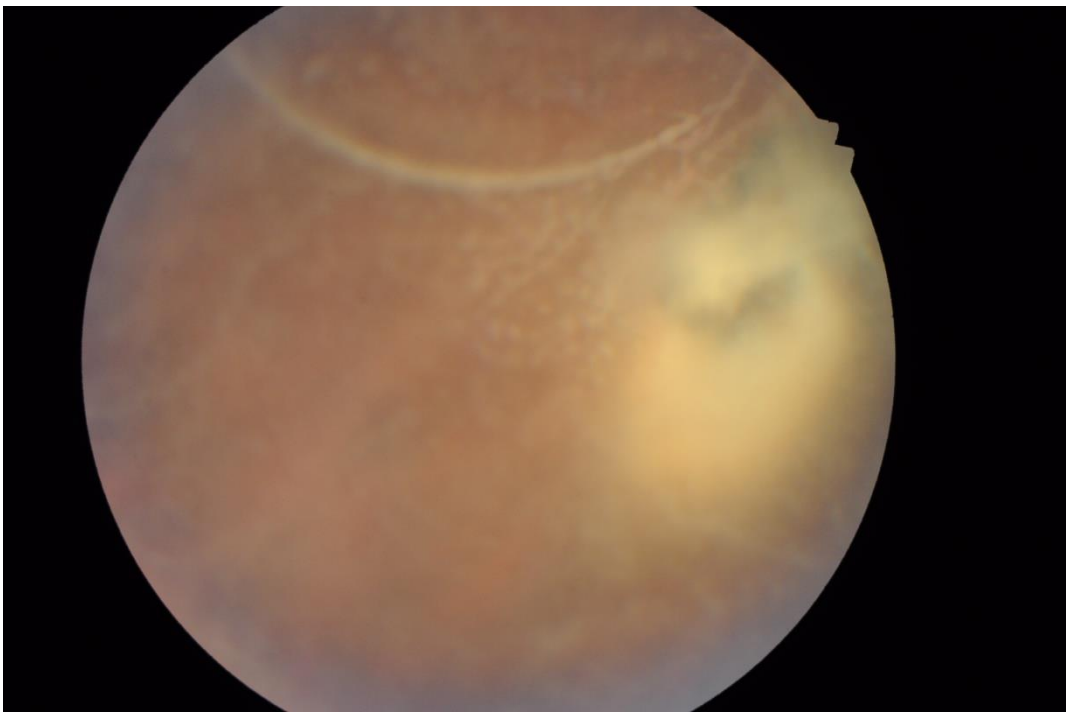


Fig 26-Toxoplasmosis with accompanying retinal vasculitis

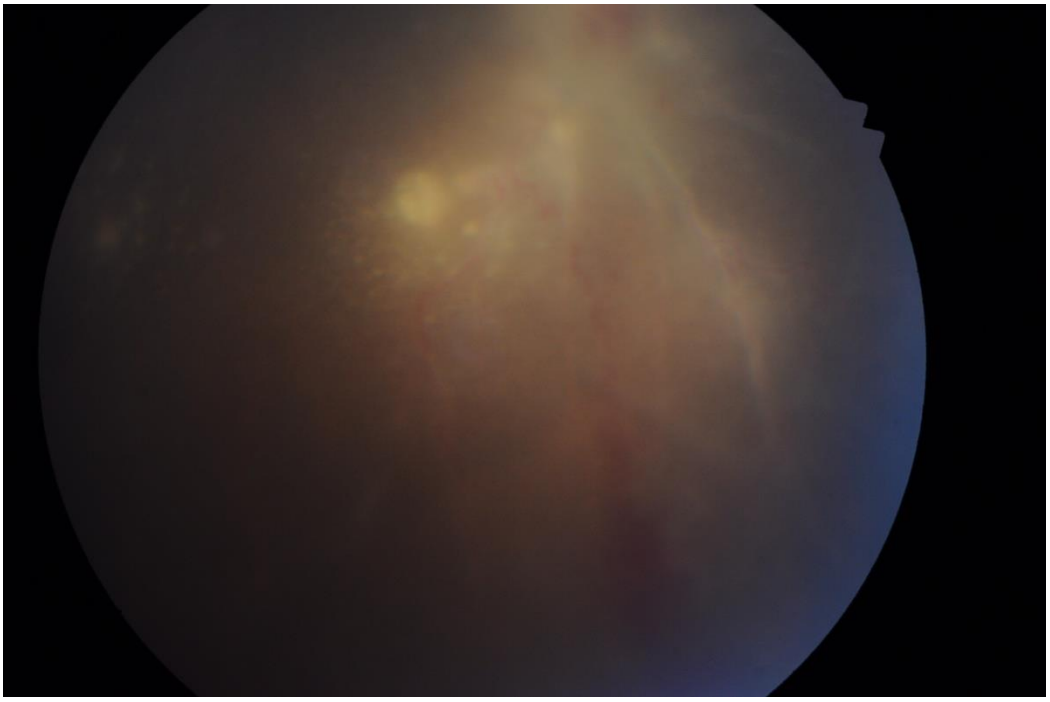


Fig 27-Toxocariasis with accompanying retinal vasculitis

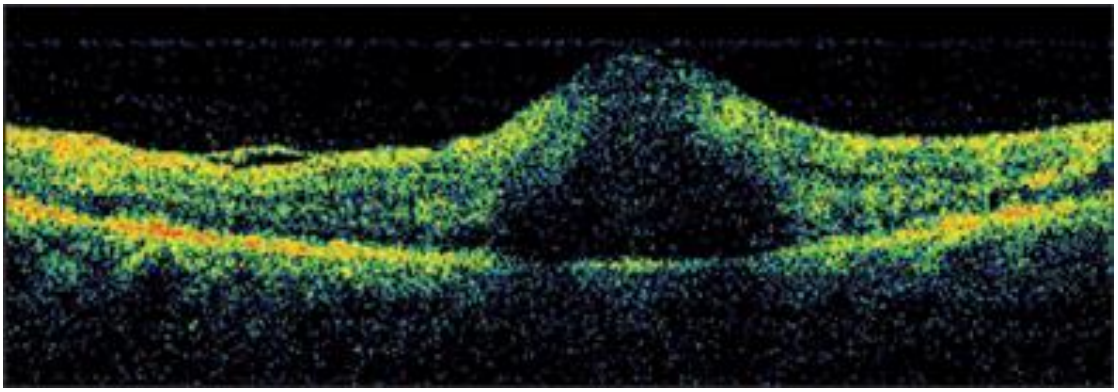


Fig 28-OCT showing macular edema in case of retinal vasculitis

DIAGNOSTIC TESTS IN RETINAL VASCULITIS

- Complete Blood Count with Differential Count
- Erythrocyte Sedimentation Rate
- C- Reactive Protein
- Renal and Liver Function Test
- Blood Sugar
- Urinanalysis
- VDRL and FTA- ABS test for Syphilis
- Mantoux Test
- TORCH Screening
- ELISA for Toxocara
- Serum ACE levels
- ELISA for HIV
- Rheumatoid Factor
- Antinuclear antibody
- Anti DNA
- Antineutrophil cytoplasmic antibody
- Antiphospholipid Antibody
- Human Leukocyte Antigen testing

- CSF analysis
- Vitreous Biopsy

IMAGING

- Chest X-Ray
- Sacro-illiac joint Xray
- Fundus Fluorescein Angiography
- Optical Coherence Tomography (Fig 28)
- B-Scan ultrasonography
- Magnetic Resonance Imaging

Role of Fundus Fluorescein Angiography in Retinal Vasculitis

After making a diagnosis of retinal vasculitis clinically, it is very essential to perform Fundus Fluorescin Angiography in eyes with a clear media. It helps in making the decision regarding management of the cases based on the findings.

We should look for leakage from the vessel walls and staining around the vessel walls which are signs of inflammation. Secondly, we should look for any areas of ischemia and capillary drop outs which will be seen as hypofluorescent areas which will guide our decision to start

laser photo-coagulation and anti –VEGF treatment. Next we should see if there are any new vessels which show early hyperfluorescence which increases in size and intensity during late phases. There might be evidence of macular edema in some cases which will be better visualised by optical coherence tomography. Presence of collaterals might be a feature in some cases with occlusion of the vessels. FFA can be repeated post laser to look for resolution of leaks from new vessels.

TREATMENT

ANTI-INFLAMMATORY

CORTICOSTEROIDS

- ✓ Systemic route (oral or intravenous administration)
- ✓ Sub-tenon's injection
- ✓ Intravitreal Triamcinalone
- ✓ Intravitreal implants

INTRAVITREAL ANTI-VEGF

IMMUNOMODULATORS

- ✓ Alkylating agents
- ✓ Anti-metabolite
- ✓ T-lympocytes modulators
- ✓ Biological response modifiers.

LASER THERAPY

Pan-retinal photocoagulation

SURGICAL THERAPY

Pars plana vitrectomy

SYTEMIC IMMUNOSUPPRESSANTS

The mainstay of treatment in vasculitis is either steroids or immunosuppressants. In case of an infective etiology, specific treatment of the infective agent with or without corticosteroids may be required. Anti-Tuberculous Therapy can be started for those testing positive for tuberculosis.

Steroids can be administered either systemically or periocularly. For mild to moderate inflammation, subtenon injection of corticosteroids (Triamcinolone acetonide 0.5-1ml; 40mg/ml) can be tried [25]. For moderate to severe inflammation oral prednisone in the dose of 1-2 mg/kg can be started. Severe cases with posterior pole involvement and refractory macular edema can be administered intra-venous methylprednisolone and intra-vitreal steroids.[26,27]

For patients who are non-responsive to steroids or steroid intolerant, immunosuppresssives can be given. Various drugs used in treatment include Methotrexate, Azathioprine, Cyclosporine, Mycophenolate Mofetil, Cyclophosphamide and Infliximab. The therapy is individualised based on the underlying cause.

In cases of associated anterior uveitis, topical steroids with cycloplegics can be started.

Laser Photocoagulation and intra-vitreal Anti VEGF injection

Retinal laser photo-coagulation should be given in cases with marked retinal ischemia with neovascularisation. Intra-vitreal antiVEGF can be injected concomitantly to arrest the progression of new vessels.

Vitrectomy

For patients with non resolving vitreous hemorrhage, Tractional Retinal Detachment and Epiretinal membrane formation.[28]

COMPLICATIONS

- Cystoid Macular Edema
- Macular ischemia
- Vein occlusions
- Vitreous hemorrhage
- Tractional Retinal Detachment
- Rubeosis Iridis
- Neovascular Glaucoma

COMPLICATIONS OF ORAL STEROIDS

FLUIDS, ELECTROLYTES

- Sodium retention
- Potassium loss
- Fluid retention
- Hypokalemic alkalosis
- Hyperosmolar coma

MUSCULOSKELETAL

- Muscle weakness
- Steroid myopathy
- Osteoporosis
- Aseptic necrosis of femoral and humeral head
- Tendon rupture

GASTROINTESTINAL

- Nausea
- Increased appetite
- Peptic ulcer
- Perforation of small and large bowel
- Pancreatitis

DERMATOLOGIC

- Poor wound healing
- Easy brusiability

NEUROLOGICAL

- Convulsion
- Headache
- Hyperexcitability
- Moodiness
- Psychosis

ENDOCRINE

- Menstrual irregularities
- Cushingoid state
- Suppression of adrenocortical pituitary axis
- Diabetes

OPHTHALMIC

- Cataract
- Glaucoma
- Central serous retinopathy
- Activation of herpes

OTHERS

- Weight gain
- Thromboembolism

AIM OF THE STUDY

PRIMARY OBJECTIVE

- To investigate the aetiologies, association with tuberculosis and other systemic illnesses

- Management of retinal vasculitis with either systemic steroids, intravitreal anti VEGF , laser photocoagulation , immunosuppressants ,vitrectomy or observation based on the case scenario

- Visual outcome following treatment of the vasculitis patient

SECONDARY OBJECTIVE

- To look for any complications following treatment
- Measures to provide rehabilitation for vasculitis patient

MATERIALS AND METHODS

50 cases of retinal vasculitis which attended the vitreo-retina clinic of Regional Institute of Ophthalmology and Government Ophthalmic Hospital, Egmore , Chennai between August 2015 and August 2016 for a period of 1 year were taken up for the study. It is a prospective study.

INCLUSION CRITERIA

1. Age >18 years

2. All cases of Retinal Vasculitis presenting with atleast one of the following features -

    a. Sheathing

    b. Perivascular inflammation

    c. Superficial and Deep Hemorrhages

    d. Staining of vessel wall on FFA

EXCLUSION CRITERIA

1. Age <18 years

2. Patient with pre existing ocular disease like diabetic retinopathy, vein occlusion , arterial occlusion, glaucoma etc

METHODOLOGY

Patient presenting to Retina Clinic were registered, evaluated and followed up during the study period.

A detailed history of the patient, visual acuity assessment, intraocular pressure measurement, Slit lamp examination, and fundus examination was done. Fundus Fluorescein Angiography and Optical Coherence Tomography was done for patients who had atleast one eye with clear media. B scan was done for patients without a clear media. All patients were subjected to following blood investigations-

1. Complete hemogram (Hemoglobin, Total WBC Count, Differential Count, Erythrocyte Sedimentation Rate)
2. Mantoux Test
3. Random Blood Sugar
4. VDRL
5. Elisa for HIV

All cases were referred to the following departments attached to Madras Medical College-

1. Rheumatalogy Clinic
2. Chest TB clinic

3. Dermatology OPD
4. STD clinic
5. ENT OPD
6. Dental OPD
7. Neurology OPD
8. Gynaecology OPD (female patients)

Patients were managed according to their presentation by either conservative treatment or surgical intervention.

- Patients with peripheral vasculitis and good visual acuity with no evidence of retinal non-perfusion were advised periodic observation

- Patients with peripheral vasculitis and good visual acuity and with less than 5 DD retinal non-perfusion , systemic steroids (T. Prednisolone 1mg/kg/day) was started which was tapered gradually.

- Immunosuppressants (Azathioprine 2mg/kg/day) planned for resistant cases (not responding to steroids treatment for period of 8 weeks)

- Patients with peripheral vasculitis with retinal non-perfusion of more than 5 DD laser photocoagulation was advised
- For cases with vitreous hemorrhage, observation and regular follow up was advised.
- For patients with non clearing vitreous hemorrhage and tractional retinal detachment vitrectomy with endophotocoagulation was advised

SCREENING PROCEDURES

- Detailed history of present illness
- Visual acuity using Snellen's acuity chart
- Slit lamp biomicroscopy of anterior segment
- Intraocular pressure using Goldmann Applanation tonometer
- Direct and Indirect Ophthalmoscopy
- Fundus Fluorescin Angiography
- Optical Coherence Tomography
- B scan
- Blood Investigations

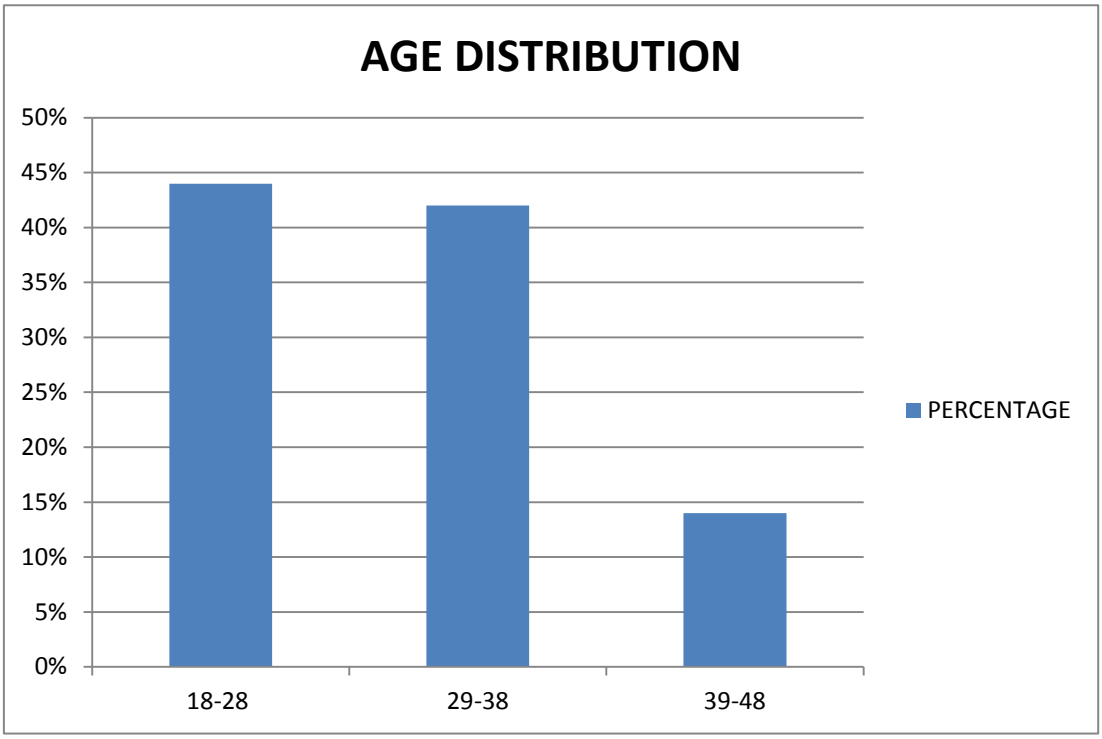
All patients planned for follow up at 1^{st},2^{rd}, 4^{th}week,3^{rd} month ,6^{th} month, 9^{th} month, 12^{th} month and the outcomes at the end of 3 months were analysed.

OBSERVATION AND ANALYSIS

1. AGE DISTRIBUTION-

TABLE 1

AGE GROUP(YEARS)	NO.OF CASES	PERCENTAGE
18-28	22	44%
29-38	21	42%
39-48	7	14%

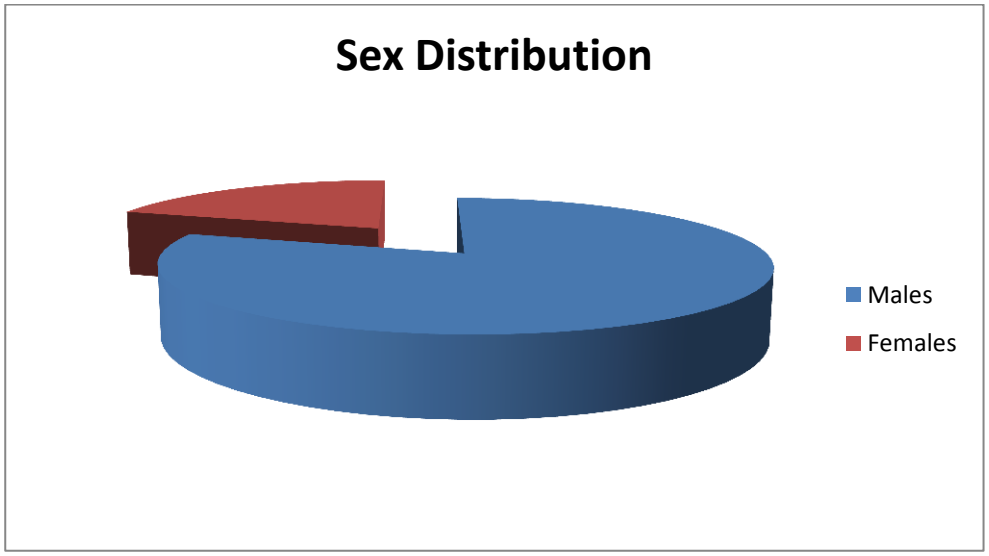


Amongst the 50 case of Retinal vasculitis which visited the retina clinic, maximum incidence was in the age group of 18-28 years (44%) followed by 29-38 years (42%) followed by 39-48 years (7%).

2. SEX DISTRIBUTION

TABLE 2

Sex	Number	Percentage
Males	40	80%
Females	10	20%

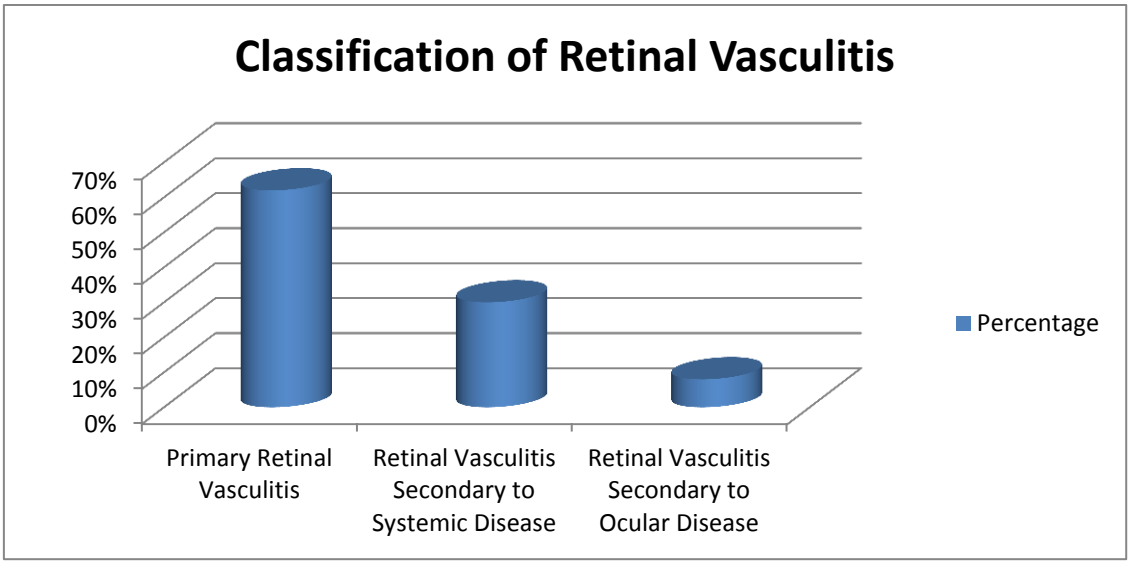


In our study, incidence of Retinal Vasculitis is more in males (80%) as compared to females (20%). Out of the 31 patients with Eales's Disease, 30 were male and only 1 was female. Out of 4 cases with retinal vasculitis secondary to ocular disease, 3 were males and 1 was female. Out of 15 cases with retinal vasculitis secondary to systemic disease, 7 were males and 8 were females.

3. CLASSIFICATION OF RETINAL VASCULITIS

TABLE 3

Disease	Number	Percentage
Primary Retinal Vasculitis	31	62%
Retinal Vasculitis Secondary to Systemic Disease	15	30%
Retinal Vasculitis Secondary to Ocular Disease	4	8%

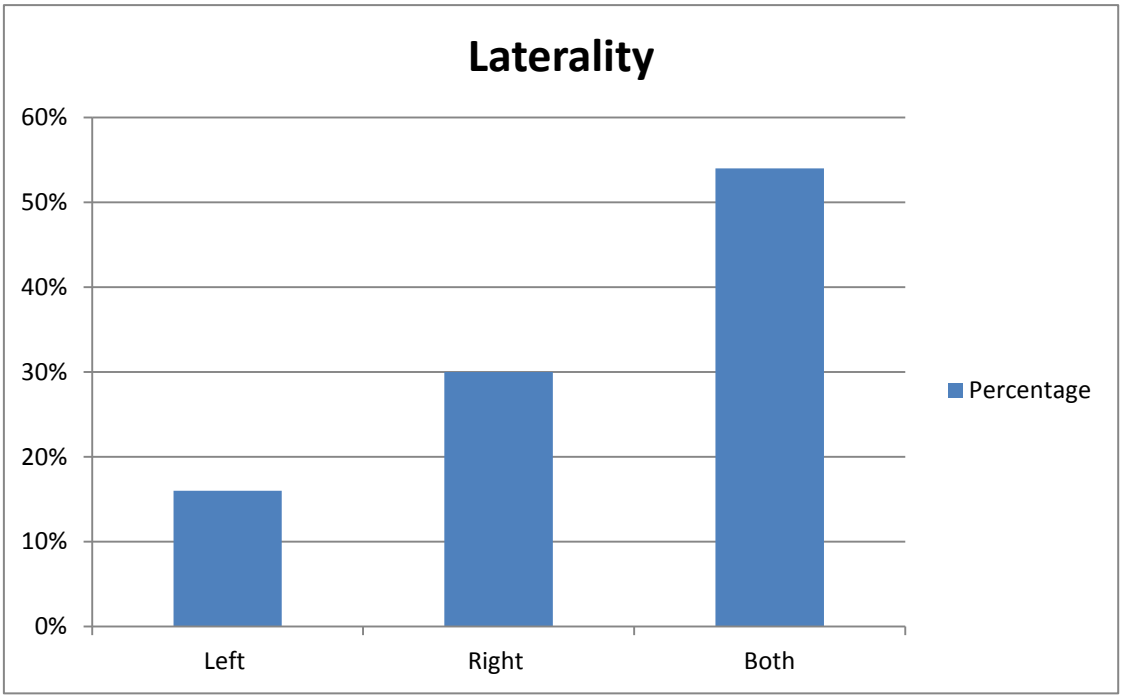


In our study, cases due to primary retinal vasculitis were maximum (62%), followed by those due to systemic diseases (30%) and due to ocular disease(8%).

4. LATERALITY

TABLE 4

Eye	Number	Percentage
Left	8	16%
Right	15	30%
Both	27	54%

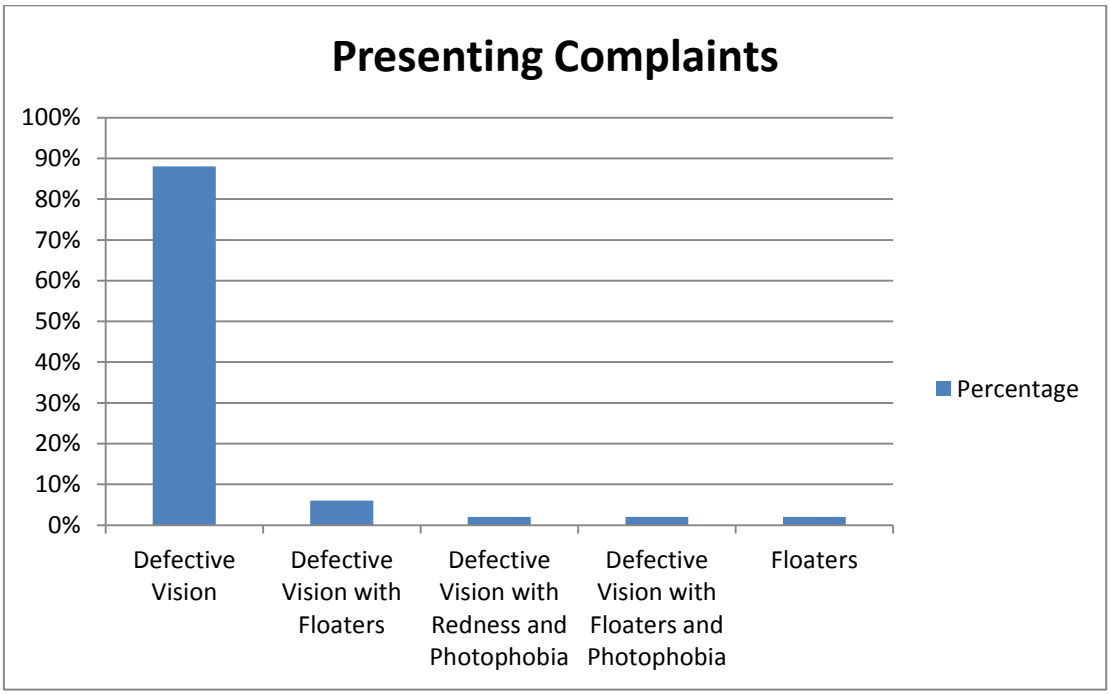


In our study, patients had predominant involvement of both the eyes (54%) followed by right eye (30%) then left eye (16%).

5. PRESENTING COMPLAINTS

TABLE 5

Presenting Complaints	No.	Percentage
Defective Vision	44	88%
Defective Vision with Floaters	3	6%
Defective Vision with Redness and Photophobia	1	2%
Defective Vision with Floaters and Photophobia	1	2%
Floaters	1	2%

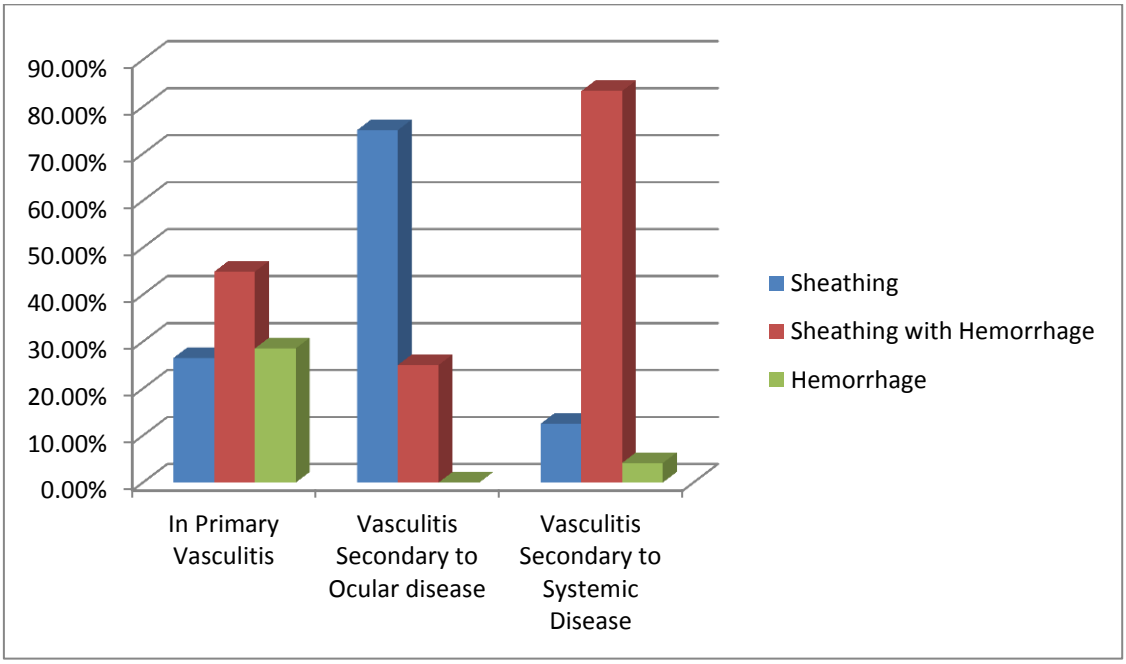


In our study, most of the patients presented with complaint of defective vision, in few cases associated with floaters, redness and photophobia.

6. CLINICAL SIGNS-

TABLE 6

Clinical Signs	Primary Vasculitis	Secondary to Ocular Disease	Secondary to Systemic Disease
Sheathing	13 eyes (26.53%)	3 eyes (75%)	3 eyes (12.5%)
Sheathing with Hemorrhage	22 eyes (44.90%)	1 eye (25%)	20 eyes (83.33%)
Hemorrhage	14eyes (28.57%)	0 (0%)	1 eye (4.17%)



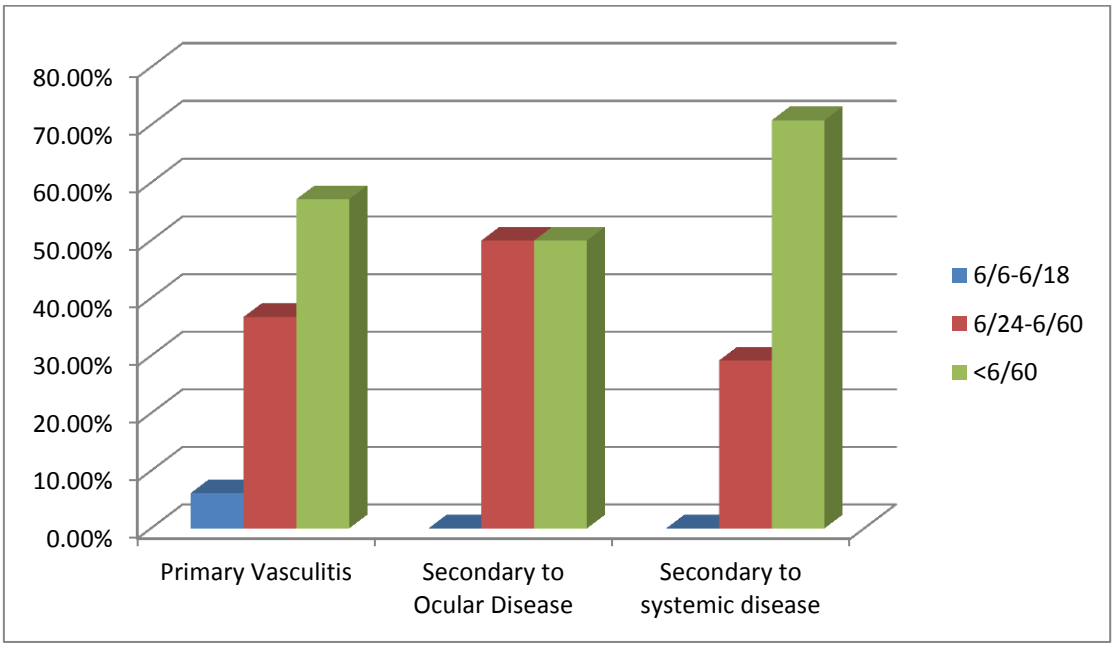
In our study, patients with Eales's Disease mostly presented with perivascular sheathing and haemorrhages, those secondary to systemic diseases also presented maximally with perivascular sheathing and haemorrhages and those secondary to ocular disease had sheathing as the predominant finding. Out of the 31 case of Eales Disease, 6 patients had vitreous hemorrhage at the time of presentation, 9 patients had associated neo-vascularisation, 7 patients had tractional bands and 2 cases were complicated with tractional retinal detachment.

Amongst the 15 cases secondary to systemic diseases, 1 patient had vitreous hemorrhage, 3 patients had neo-vascularisation and 3 patients had tractional bands.

7. BEST CORRECTED VISUAL ACUITY AT PRESENTATION-

TABLE 7

BCVA	Primary Vasculitis	Secondary to Ocular Disease	Secondary to systemic disease
6/6-6/18	3 eyes (6.12%)	0%	0%
6/24-6/60	18 eyes (36.73%)	2 eyes (50%)	7 eyes (29.17%)
<6/60	28 eyes (57.15%)	2 eyes (50%)	17 eyes (70.83%)

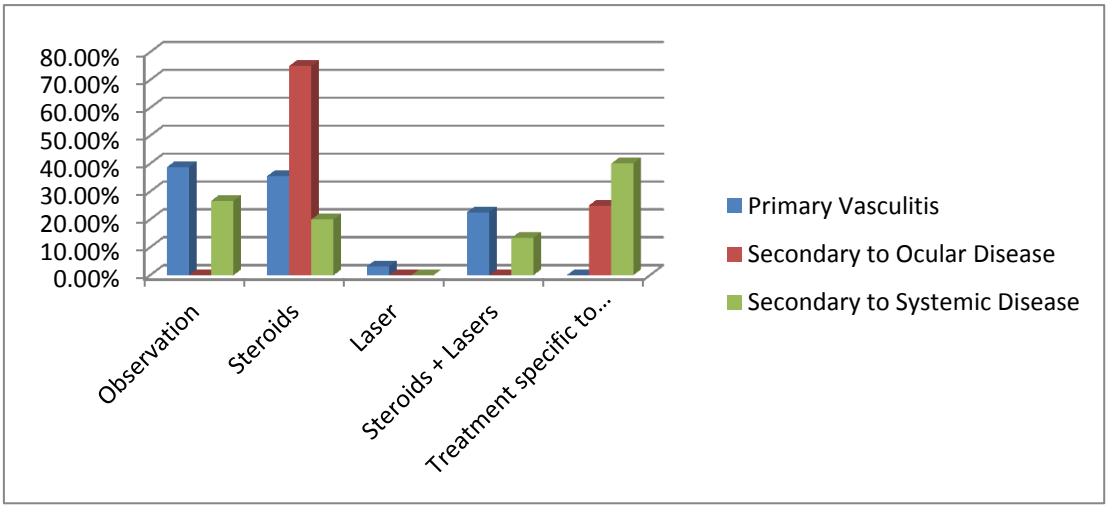


In our study, in patients with Eales's Disease, best corrected visual acuity at presentation was predominantly less than 6/60 (57.15%). In patients with vasculitis secondary to ocular disease, best corrected visual acuity was equally present in range of 6/24-6/60 and less than 6/60. In patients with vasculitis secondary to systemic disease, best corrected visual acuity was mostly less than 6/60 (70.83%).

8. TREATMENT MODALITY

TABLE 8

Treatment Modality	Primary Vasculitis	Secondary to Ocular Disease	Secondary to Systemic Disease
Observation	12 patients (38.71%)	0%	4 patients (26.67%)
Steroids	11 patients (35.48%)	3 patients (75%)	3 patients (20%)
Laser	1 patient (3.23%)	0%	0%
Steroids + Lasers	7 patients (22.58%)	0%	2 patients (13.33%)
Treatment specific to disease entity	0%	1 patient (25%)	6 patients (40%)



In patients with Eales Disease, those who presented with vitreous hemorrhage, tractional retinal detachment, tractional bands and minimal sheathing (12 patients) with no capillary non-perfusion areas observation was advised. Those patients who had extensive sheathing and haemorrhages with capillary non perfusion less than 5 DD (11 patients)

were started on T. Prednisolone 1mg/kg/day in tapered dosing. Steroids and laser photo-coagulation was the treatment for 7 patients with neovascularisation and capillary non-perfusion areas more than 5DD.

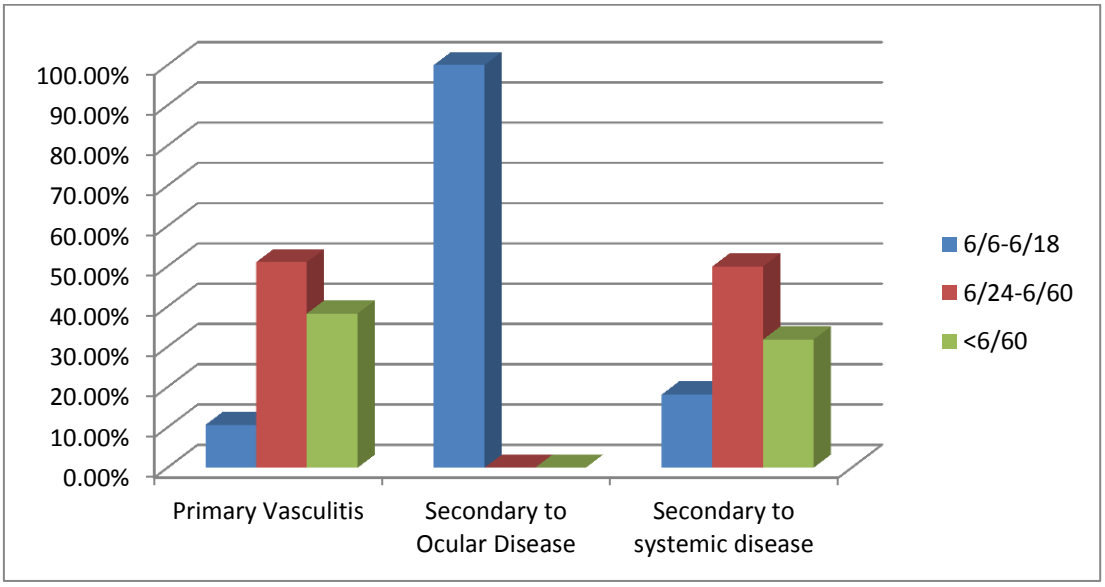
In patients with vasculitis secondary to ocular disease,2 patients were secondary to intermediate uveitis and 1 patient with choroiditis were started on oral steroids. 1 patient with choroiditis with associated tuberculosis was started on anti-tuberculous therapy and oral steroids.

In patients with vasculitis secondary to systemic disorders, 4 patients with associated acquired immunodeficiency syndrome were kept on observation. 1 patient with associated ankylosing spondylitis and 2 patients with systemic lupus erythematosus were started on oral steroids. 1 patient with ankylosing spondylitis and 1 with systemic lupus erythematous having neovascularisation were administered laser photo-coagulation along with oral steroids. 3 patients secondary to tuberculosis were prescribed anti-tuberculous therapy and oral steroids.1 patient with associated cytomegalovirus infection was started on IV Ganciclovir with oral steroids.1 patient with associated toxocara infection with vasculitis as well as anterior uveitis was started on T. Albendazole 15mg/kg/day for 10 days along with oral steroids and topical steroids with cycloplegics. 1 patient with associated toxoplasma infection with both vasculitis and anterior uveitis was administered T. Bactrim DS along with oral steroids and topical steroids and cycloplegics.

9. BEST CORRECTED VISUAL ACUITY AFTER 3 MONTHS-

TABLE 9

BCVA	Primary Vasculitis	Secondary to Ocular Disease	Secondary to systemic disease
6/6-6/18	5 eyes (10.64%)	4 eyes (100%)	4eyes (18.18%)
6/24-6/60	24 eyes (51.06%)	0%	11 eyes (50%)
<6/60	18 eyes (38.30%)	0%	7 eyes (31.82%)



In patients with Eales Disease, visual improvement was noted 3 months post treatment with 10.64% eyes in the range of 6/6-6/18 as compared to 6.12% eyes pre-treatment. In the range of 6/24-6/60, post-treatment there were 51.06% eyes as compared to 36.73 % pre-treatment. Percentage of patients in less than 6/60 range reduced to 38.30% from initial 57.15%.

In vasculitis secondary to ocular disease, dramatic improvement was seen, with 100% patients improving in the range of 6/6-6/18 as compared to 0% pre-treatment.

In vasculitis secondary to systemic disease, patients in the range of 6/6-6/18 increased to 18.18% compared to nil initially. Patients in the range of 6/24-6/60 increased to 50% from initial 29.17%. Patients with visual acuity less than 6/60 reduced to 31.82% compared to 70.83% initially.

2 patients were lost to follow up.

10. COMPLICATIONS-

5 patients developed weight gain following steroid administration and 2 patients complaint of gastritis following oral steroid administration.

DISCUSSION AND RESULTS

In this study on 77 eyes of 50 retinal vasculitis patients, 31 cases(62%) were due to Eales Disease , 15 cases (30%) were secondary to systemic disease and the remaining 4 cases (8%) were secondary to ocular disease.

Amongst the 50 case of Retinal vasculitis which visited the retina clinic, maximum incidence was in the age group of 18-28 years (44%) followed by 29-38 years (42%) followed by 39-48 years (7%). The mean age was 30.02 $\pm$ 12 years. In a study by Biswas Et al, mean age of presentation was found to be 33 $\pm$ 11. In a study by Donders in 1958 the average age for men was 28 years and for women it was 30 years.

In our study, incidence of Retinal Vasculitis is more in males (80%) as compared to females (20%). Out of the 31 patients with Eales's Disease, 30 were male and only 1 was female. Out of 4 cases with retinal vasculitis secondary to ocular disease, 3 were males and 1 was female. Out of 15 cases with retinal vasculitis secondary to systemic disease, 7 were males and 8 were females.

This male predilection in Eales Disease was supported by Elliot AJ:Thirty year observation of patients with Eales disease, in his study 93% were Male.[29]

In our study vasculitis secondary to systemic disease showed almost equal incidence in males and females.

In our study, patients had predominant involvement of both the eyes (54%) followed by right eye (30%) then left eye (16%). In patients with Eales Disease, bilaterality was seen in 18 out of 31 patients (58.06%) and unilateral disease was seen in 13 patients (41.93%) indicating towards bilateral presentation being more common than unilateral presentation. Duke Elder has reported that 90% patients with Eales Disease will have a bilateral presentation as a rule.[1]

In our study, most of the patients presented with complaint of defective vision (88%), in few cases associated with floaters, redness and photophobia. In 8 cases the defective vision was attributable to presence of vitreous hemorrhage. Out of these 8 cases, 7 were due to Eales Disease and 1 was secondary to systemic tuberculosis. Defective vision with floaters was reported in 3 cases (6% of cases) . All 3 patients presented with retinal vasculitis secondary to ocular disease. 2 cases had associated

intermediate uveitis and 1 had choroiditis secondary to tuberculosis. 2 patients with retinal vasculitis secondary to systemic toxoplasmosis and toxocariasis complaint of redness and photophobia due to associated anterior uveitis. Only 1 case reported only floaters and was diagnosed as having Eales Disease. Probably it was in the initial stage of inflammation at time of diagnosis.

In a study by Biswas et al, defective vision was found to be the commonest symptom similar to our study. But they reported higher incidence of floaters as compared to our study. [30]

In our study, patients with Eales's Disease mostly presented with perivascular sheathing and haemorrhages, those secondary to systemic diseases also presented maximally with perivascular sheathing and haemorrhages and those secondary to ocular disease had sheathing as the predominant finding. Out of the 31 case of Eales Disease, 6 patients had vitreous hemorrhage at the time of presentation, 9 patients had associated neo-vascularisation, 7 patients had tractional bands and 2 cases were complicated with tractional retinal detachment.

Amongst the 15 cases secondary to systemic diseases, 1 patient had vitreous hemorrhage, 3 patients had neo-vascularisation and 3 patients had tractional bands.

In their study, Biswas et al have also reported vascular sheathing as the commonest sign. But the study conducted by Saxena et al shows vitreous hemorrhage as the commonest sign.[31,32] In a study by Supanut et al also found vascular sheathing as the most common clinical sign involving veins more than the arteries[33,34].

In our study, in patients with Eales's Disease, best corrected visual acuity at presentation was predominantly less than 6/60 (57.15%) probably due to association of vitreous hemorrhage and tractional retinal detachment in these patients.

In patients with vasculitis secondary to ocular disease, best corrected visual acuity was equally present in range of 6/24-6/60 and less than 6/60. These patients mainly had extensive sheathing with haemorrhages. Associated vitritis could be a reason for vision in the range less than 6/60.

In patients with vasculitis secondary to systemic disease, best corrected visual acuity was mostly less than 6/60 (70.83%).

In patients with Eales Disease , those who presented with vitreous hemorrhage, tractional retinal detachment, tractional bands and minimal sheathing (12 patients) with no capillary non-perfusion areas observation was advised. Those patients who had extensive sheathing and haemorrhages with capillary non perfusion less than 5 DD (11 patients) were started on T. Prednisolone 1mg/kg/day in tapered dosing. Steroids and laser photo-coagulation was the treatment for 7 patients with neovascularisation and capillary non-perfusion areas more than 5DD. Observation was mainstay of treatment in these cases followed closely by oral steroids.

In patients with vasculitis secondary to ocular disease,2 patients were secondary to intermediate uveitis and 1 patient with choroiditis were started on oral steroids. 1 patient with choroiditis with associated tuberculosis was started on anti-tuberculous therapy and oral steroids. In these cases oral steroids were the mainstay of treatment. In case secondary to infectious etiology i.e tuberculosis anti-tuberculous therapy was warranted along with steroids.

In patients with vasculitis secondary to systemic disorders, 4 patients with associated acquired immunodeficiency syndrome were kept on observation . 1 patient with associated ankylosing spondylitis and 2 patients with systemic lupus erythematosus were started on oral steroids. 1 patient with ankylosing spondylitis and 1 with systemic lupus

erythematous having neovascularisation were administered laser photo-coagulation along with oral steroids. 3 patients secondary to tuberculosis were prescribed anti-tuberculous therapy and oral steroids.1 patient with associated cytomegalovirus infection was started on IV Ganciclovir with oral steroids.1 patient with associated toxocara infection with vasculitis as well as anterior uveitis was started on T. Albendazole 15mg/kg/day for 10 days along with oral steroids and topical steroids with cycloplegics. 1 patient with associated toxoplasma infection with both vasculitis and anterior uveitis was administered T. Bactrim DS along with oral steroids and topical steroids and cycloplegics. In these case also, oral steroids were the main treatment to decrease the inflammation in addition to the specific therapy for associated infectious and non-infectious causes which was essential to curb the systemic disease. Topical steroids and cycloplegics were helpful in treating the associated anterior uveitis in 2 cases. In the study by Biswas et al, corticosteroids were found to be the main treatment modality similar to our study. [35]

After a follow up period of 3 months, patients' visual acuity was reviewed. In patients with Eales Disease, visual improvement was noted 3 months post treatment with 10.64% eyes in the range of 6/6-6/18 as compared to 6.12% eyes pre-treatment. In the range of 6/24-6/60, post-treatment there were 51.06% eyes as compared to 36.73 % pre-treatment. Percentage of patients in less than 6/60 range reduced to 38.30% from initial 57.15%.

In vasculitis secondary to ocular disease, dramatic improvement was seen, with 100% patients improving in the range of 6/6-6/18 as compared to 0% pre-treatment.

In vasculitis secondary to systemic disease, patients in the range of 6/6-6/18 increased to 18.18% compared to nil initially. Patients in the range of 6/24-6/60 increased to 50% from initial 29.17%. Patients with visual acuity less than 6/60 reduced to 31.82% compared to 70.83% initially.

2 patients were lost to follow up.

Complications were noted in a few cases with 5 patients developing weight gain following oral corticosteroid administration. But we did not switch over to immunosuppresants because patient was responding well to oral corticosteroids. Patient was advised occlusion therapy .Two patients complaint of gastritis following oral steroid administration.

CONCLUSION

Retinal vasculitis is a challenge for an ophthalmologist both to diagnose and to treat. If left untreated it may lead to total loss of vision. Ophthalmologist also play an important role in finding out systemic diseases in a patient who have no other manifestations apart from ophthalmological signs. That aids in timely treatment of the underlying cause. For each patient treatment has to be individualised based on their findings. A multidisciplinary approach is required in cases with systemic involvement.

In our study, primary vasculitis is the predominant form of vasculitis accounting for 62% of the cases.

Male predilection is noted with 80% cases being males and 20% being females.

Most commonly involved age group is between 18-28 years of age (44 %).

Maximum cases had a bilateral presentation (54%) with defective vision being the chief complaint in most of the cases (88%).

Oral corticosteroids were the main modality of treatment in our study. Patients treated with laser photo-coagulation did not show worsening of signs. Observation was helpful in patients with vitreous hemorrhage. Treatment of associated systemic conditions in addition to oral corticosteroids showed improvement in visual acuity.

Although systemic steroids are efficacious in controlling active disease and easily administered, adverse systemic complications is a matter of concern.

Since the time period of the study was short (1 year), we have kept 3 months as the criteria for assessing the outcome of management. Due to referrals to various departments, patients took time to review with us with all the work up.

More long term studies are required to overcome these shortcomings.

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PROFORMA

1. NAME :

2. AGE/SEX :

3. IP NO :

4. ADDRESS:

5. OCCUPATION:

6. CHIEF COMPLAINTS:

A. OCULAR

	RE	LE	Duration
a.	**Defective Vision (Distant/Near)**		
b.	**Total loss of vision**		
c.	**Floaters**		
d.	**Flashes**		
e.	**Positive scotoma**		
f.	**Macropsia/Micropsia/Metamorphopsia**		
g.	**Field defects**		
h.	**Pain**		
i.	**Redness**		

B.SYSTEMIC- Joint pains/ rashes/fever/ oral ulcers/genital ulcers/ skin lesions/ chest pain cough/ weight loss/ weakness of limbs/others

7. PAST HISTORY:

Diabetes Mellitus/ Hypertension/ Ischemic heart Disease/ Tuberculosis/ Bronchial Asthma/ HIV/Syphilis/ skin diseases like psoriasis

Duration:

8. FAMILY HISTORY:

9. PERSONAL HISTORY:

Smoking/ Alcoholism/ Vegetarian/ Non Vegetarian

10. TREATMENT HISTORY :

Medications **Surgery**

11. EXAMINATION: **RE** **LE**

Visual Acuity

Extra Ocular Examination

Tension

Lids

Conjunctiva

Cornea

Iris

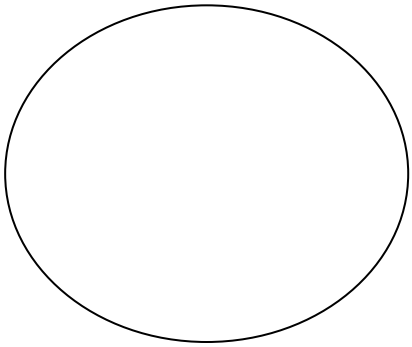
Anterior Chamber

Pupil

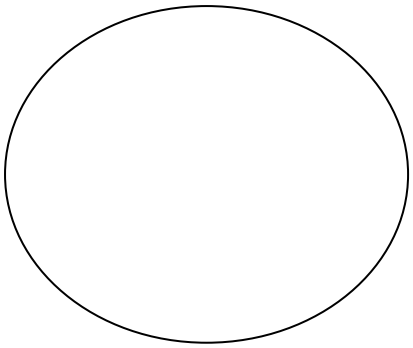
Lens

Vitreous

12. FUNDUS EXAMINATION:



RE



LE

13. CLINICAL DIAGNOSIS

14. INVESTIGATIONS

a. BP-

b. RBS-

c. Hemoglobin d.TC e.DC f. ESR

g. VDRL

h. ELISA for HIV

15. FUNDUS FLUORECEIN ANGIOGRAPHY

16. OPTICAL COHERENCE TOMOGRAPHY

17. B SCAN:

18. OTHER SPECIALITY OPINIONS-

19. TREATMENT:

20. FOLLOW UP:

KEY TO MASTERCHART

Sex

M - Male

F - Female

Laterality

B - Both Eyes

R - Right Eye

L - Left Eye

Symptoms

D - Defective Vision

F - Floaters

R - Redness

P - Photophobia

Associated Systemic Disorder

TB - Tuberculosis

AIDS - Acquired Immunodeficiency Syndrome

SLE - Systemic Lupus Erythematosus

AS - Ankylosing Spondylitis

CMV - Cytomegalovirus

TP - Toxoplasma

TC - Toxocara

Associated Ocular Condition

IU - Intermediate Uveitis

C - Choroiditis

AU - Anterior Uveitis

SIGNS

S - Sheathing

H - Hemorrhage

Visual Acuity

HM - Hand Movements

CFCF - Counting Fingers

Diagnosis

E - Eales Disease

O - Vasculitis secondary to ocular disease

S - Vasculitis Secondary to Systemic Disease

Treatment

S - Oral Corticosteroids

L - Laser photocoagulation

O - Observation

A - Anti tuberculuous therapy

T - Topical Steroids and Cycloplegics

B - Bactrim DS

Z - Albendazole

G - Ganciclovir

Complication

W - Weight Gain

G - Gastritis