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Review Article

Retinal Vein Occlusion

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Introduction

Retinal vein occlusion is occurring due to blockage of the small veins that is carrying blood away from the retina. It is usually happen by hardening of the arteries and formation of blood clot. It is the 2nd most common retinal vascular disease and is a common cause of decreased in vision in old age patients.

Pathogenesis: - The central retinal vein and artery share a common adventitial sheath at crossing point's posterior to the lamina cribrosa. Typically, in branch retinal vein occlusion arteriosclerotic thickening of a branch retinal arteriole is associated with compression of a vein at an arteriovenous crossing point. This causes secondary changes including venous endothelial cell loss, thrombus formation and eventually vein occlusion. There is stagnation of blood flow which causes hypoxia in the drained area of obstructed vein.

Retinal Blood supply:-

Outer 4 layers of RETINA supplied by Choriocapillaries.

Inner 6 layers of RETINA supplied by Central retinal artery

The fovea is avascular and is mainly supplied by Choriocapillaries.

Cilio retinal artery is present in 20% of eyes

The veins of the RETINA unite to form Central retinal vein at the disc, which follows the corresponding artery

Classification: - Mainly venous occlusion 3 types.

1. Central Retinal vein occlusion :- It is the blockage of the main retinal vein
2. Branch Retinal Vein Occlusion :- It is the blockage of one of the smaller branch vein
3. Hemi Central retinal Vein Occlusion: - It is the blockage of the superior or inferior branch of central retinal vein.

Central Retinal vein Occlusion

Central Retinal vein occlusion (CRVO):- It is an occlusion of the main retinal vein posteriorly to the lamina cribrosa of the optic nerve and caused by thrombosis. Mainly CRVO classified in to two categories non-ischemic and ischemic. In all over the globe Non Ischemic Central retinal vein occlusion is common approx, 70 % of cases are non -ischemic reported. The best corrected visual acuity is better than 6/60. Mild or no pupillary defect along with mild to moderate visual changes can be seen in non-ischemic CRVO. In Ischemic CRVO actually it can be progression of a non-ischemic CRVO. In Ischemic CRVO poor visual prognosis. More than 90 % of patient visual acuity is less than 6/60. Ischemic CRVO has a poor prognosis.

Etiology

In Middle East the primary risk factor for the development of central retinal vein occlusion is age and Hypertension. The 90 % of venous occlusion patients are older than 50 years along with systemic hypertension. There are some cases are diabetic mellitus open angle glaucoma and hyperlipidemia also reported. The other risk factors are smoking, obesity and thrombolytic disorder, any reason of reduced venous outflow, the chances of venous occlusion is high.

The venous occlusion is one of the main common reasons for the painless vision loss in gulf country, approx 5 cases found in 1000.

Physiology

There are 3 main factors which contribute to thrombosis, venous stasis, endothelial damage and hypercoagulability. If it is increase in these can lead a central vein occlusion. Anatomically through the process of atherosclerosis, the compression of the vein by the artery can cause of central retinal vein occlusion.

History

Patient suffering from CRVO will describe the sudden Visual Loss. The vision loss is painless. Visual disturbance depends upon the severity of the venous occlusion.

Evaluation

The evaluation of the central retinal vein occlusion required lots of laboratory examination to determine the cause, like, Blood pressure, Complete blood count, Random blood glaucose, Cholestrol, Plasma protein electrophoresis, Blood urea, Thyroid test, Left ventricular hypertrophy, Erythrocyte etc along with chest X Ray, C reactive protein, Rheumatoid factor, Treponemal serology, carotid doplor.

Risk Factors:- There are Following risk factor in the venous occlusion

- **AGE:** 50% of cases of retinal vein occlusion in more than 65 years.

- **Hypertension** :73% in >50 years ,25% in younger patients. It is most prevalent in patients with BRVO.
- **Hyperlipidemia** is present in one-thirds or more of RVO.
- **Diabetes Mellitus** 15% of cases over the age of 50 years.
- Smoking and obesity is associated with an increase incidence of RVO
- Raised intraocular pressure:-More common in CRVO.
- INFLAMMATORY DISEASES, sarcodosis and behcet diseases Causes periphlebitis.
- Hyperviscosity, Acquired or Inherited Thrombophilic Disorder

Non-Ischaemic CRVO

Non-ischemic CRVO is the most common type, accounting for about 75%.

Clinical Features:-

- Sudden painless unilateral loss of vision.
- Vision Acuity is impaired to a moderate degree.
- RAPD is absent or mild (in contrast to ischaemic CRVO)

- Most acute signs resolve over 6–12 months.
- The Main cause of poor vision is chronic macular edema and Secondary atrophy.
- Disc collaterals are common following CRVO, appearing as a small vascular loop on the optic nerve head.
- Conversion to ischaemic CRVO occurs in 15% of cases within 4 Months and 34% within 3 years

Fundus

There is Tortuosity and dilatation of all the branches

Dot/blot and flame-shaped haemorrhages can be seen throughout all Quadrants

Cotton wool spots are rare.

Mild Optic Disc and macular edema are common.

Investigations

Flourescein angiography shows delayed A-V transit time, blockage by hemorrhages, good retinal capillary perfusion and late leakage.

OCT is useful in the assessment of Macular edema.

Follow up

Monthly for the first 6 months, followed by 3 monthly for the next 24 months.

Ischaemic CRVO

Ischemic CRVO is characterized by substantially decreased retinal perfusion with capillary closure and retinal hypoxia. Macular ischemia and NVG are the major cause of visual morbidity.

Clinical features

- Sudden and severe painless unilateral loss of vision occasionally can present with pain, redness or photophobia due to neovascular glaucoma, a prior reduction in vision having passed unnoticed or been ignored.
- Vision acuity is 6/60 or counting fingers
- RAPD is present.
- Anterior segment findings: - NVI, Rubeiosis Iridis developes in about 50% of eyes, usually between 2 and 4 months.
- Most acute signs resolve over 9–12 months.

Fundus

- Tortuosity and engorgement of all branches of the central retinal vein.
- Extensive deep dot/blot and flame-shaped hemorrhages involving the peripheral and posterior retina.
- Cotton wool spots are typically prominent, optic disc swelling usually present.
- FA shows a marked delay in arteriovenous transit time, masking by retinal haemorrhages, extensive areas of capillary non-perfusion and vessel wall staining and leakage.
- OCT enables quantification of CMO. OCT shows macular edema.
- Electroretinogram is depressed and the extent of this has sometimes been used to assess neovascular risk.
- Retinal Neovascularization occur in about 5% of eyes.
- Optic Disc collaterals are common

Systemic Assessments:- In systemic assessment Need to check Blood pressure, ESR, CBC, RBS, Lipid Profie, Urea Creatinine, electrolytes, ECG, ECHO. In some cases Chest X ray in sarcoidosis patient, CRP. Plasma homocysteline level, Plasma protein electrophoresis, Autoantibodies, ANA, ANCA, antiphospholipid antibody can be done.

Management:-

There is no effective medical treatment is available. The main treatment is Intravitreal Injection Anti-VEGF drug to decrease macular edema and blood vessels growth and swelling. Sometimes there are surgical and laser treatment required to improve the vision.

Treatment of Macular Oedema:- Intravitreal anti-VEGF agents: Ranibizumab/Aflibercept intravitreal injections showed a significant visual benefit when used for CMO. Intravitreal Dexamethasone implant is used when the Anti VEGF not effective.

Indication for Intra vitreal injection is if vision is less than 6/9 and significant central macular thickening showing in OCT.

Treatment of Neovascularization:- Intravitreal anti-VEGF agents and PRP in eyes with NVI or NVD OR NVE: application of 1500–3000 burns (0.5–0.1 second, spaced one burn width apart.

Features	Non ischemic	Ischemic
Incidence	More Common	Less Frequent
Visual Loss	Painless, slight or moderate, unilateral	Painless, marked, Unilateral
Pupil	Normal	RAPD Present
Fundus venous dilation	Mild to moderate	Well marked
Retinal hemorrhage	Mild to moderate	Severe hemorrhage involving mostly posterior pole
Cotton wool spot	Rare	Common
Macular chamges	May be or May not	Early and marked
Disc Swelling	Mild	Moderate to severe
Fundus Fluorescien Angiography	Good capillary perfusion, Venous staining	Extensive Cappilary non perfusion
Complication	Vision can be improved, 50 % can get normal vision	Visual loss due to Glaucoma neovascularization, Maculopathy and Vitreous hemorrhage.

Table of Non Ischemic and ischemic retinal vein occlusion.

Differential Diagnosis:-

The differential diagnosis can be:-

Branch retinal vein occlusion.

Proliferative Diabetic retinopathy.

Ocular Ischemic syndrome.

Prognosis:-

The prognosis of Non Ischemic central retinal vein occlusion is good. The visual acuity returns to normal in 50 percent cases. In Non Ischemic CRVO Macular edema is the main reason for decreased vision. Prognosis depends in the most cases with the initial vision. If Vision in early 6/12, the vision is likely to the same, If the vision is 6/24 or less the clinical course can be different , it can improve, same or may be worsen. If The vision is less than 6/60 the improvement is very less. Ischemia CRVO has a poorer prognosis due to macular ischemia. The chance of Neovascular glaucoma is very high due to rubeosis iridis. Generally within 2 to 4 months retinal neovascularization occurs in 5 % cases.

Complication:-

The main complications are macular ischemia, vitreous hemorrhage, Neovascular glaucoma and macular edema. Macular edema is main reason for the decreased vision. Neovascularization develops in two third of cases in ischemic CRVO.

Branch Retinal Vein Occlusion

In branch retinal vein occlusion, Macular BRVO involving only a macular branch. Peripheral BRVO is not involving the macular circulation.

Etiology:

Branch retinal Vein Occlusion can have multiple causes like Hypertension, Diabetes mellitus and advanced age. It can be devided in to two part peripheral BRVO and Macular BRVO. It refers to the obstruction of a branch of the retinal vein at an arteriovenous crossing. The most affected part of the retina is superotemporal quadrant in almost 65% of the eye.

Pathophysiology:-

The main reason of decreased vision in BRVO is macular edema which can have multiple pathophysiological mechanisms. The thickening of the arterial wall which can compress the retinal vein at a point where they cross resulting in turbulent blood flow and potential thrombus formation.

History:-

Branch retinal vein occlusion sometimes asymptomatic however it can be sudden onset of painless decreased vision. It is very important to see the history of hypertension, smoking, glaucoma, diabetes or other systemic diseases and risk factors.

Evaluation:-

A complete eye examination including dilated fundus examination

Clinical Features:-

- Sudden painless loss of vision.
- Peripheral occlusion may be asymptomatic, VA is variable, Historically 50% of untreated eyes retain 6/12 or better vision in the long term, but about a quarter only achieve 6/60/or worse.
- NVI and NVG are much less common than CRVO (2-3% at 3 years)
- Chronic macular edema is the most common cause of persistent decreased visual acuity after BRVO.

Fundus

In fundus Tortuosity with dot or blot and flam shaped hemorrhage can be seen.Cotton wool spots and retinal edema is present. Supero temporal quadrant is most commonly affected. The site of occlusion may be identifiable at an arteriovenous crossing point. The acute features usually resolve within 6–12 months. Later venous sheathing and sclerosis develop.

Management / Treatment

Systemic assessment should carried out with appropriate specialist referral. Observation without intervention if VA is 6/9 or better.

Macular edema-VA worse than 6/9 and significant central macular thickening on OCT.

Intravitreal anti-VEGF agents: Ranibizumab/aflibercept injections. It is first line of treatment.It causes significant improvemet in macular edema & visual acuity

If macular edema is not responding, Intravitreal Dexamethasone implant can be given. It can raise Intra ocular pressure or can cause cataract.

NVE/NVI: Sectoral photocoagulation 200-500 µm diameter for 0.05 sec duration and spaced one burn width apart are applied to ischemic area.

Hemi Retinal Vein Occlusion

Hemiretinal vein occlusion is generally regarded as a variant of CRVO and may be ischaemic or non-ischaemic. It is less common than CRVO and BRVO. It can involving the superior or inferior branch of Central retinal vein.

Clinical features

- Sudden onset altitudinal visual field defect.
- Vision reduction is variable.
- NVI more common than BRVO , but less than CRVO

- Fundus shows the features of BRVO, involving the superior or inferior hemisphere , NVD more common
- FA shows blocked fluorescence due to masking by haemorrhages , hyperfluorescence due to leakage and variable capillary non perfusion

Treatment:-

It depends on the severity of retinal ischaemia. Extensive retinal ischaemia carries the risk of neovascular glaucoma and should be managed in the same way as ischaemic CRVO.

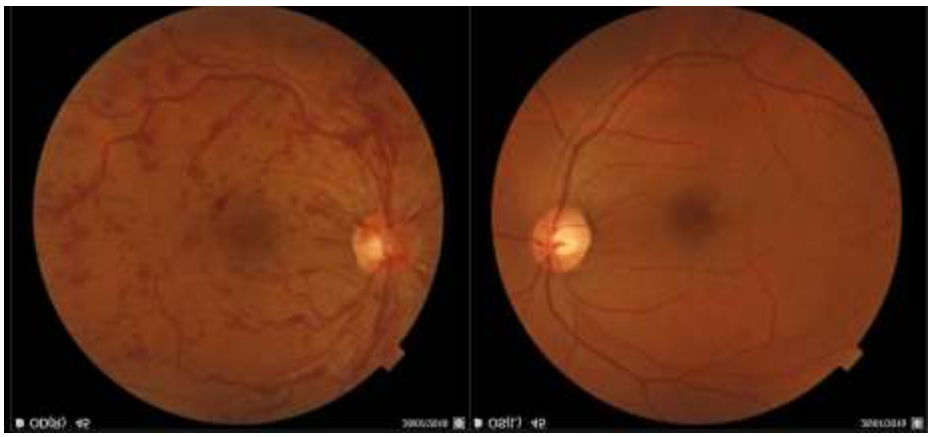
Case Presentation

A 48 year old male patient presented with complaints of sudden painless decreased vision with 5 days duration in the right eye. In Medical history Patient was recently diagnosed hypertensive. On Ocular examination found Vision Right eye 0.4 and left eye 1.0, Mild RAPD Present. Anterior segment and intraocular pressure found both eye were normal. On fundus examination of right eye dilated and tortuosity of the blood vessels along with flame shaped hemorrhage, dot blot, retinal edema and cotton wool spots is upper quadrant involving macula and left eye fundus was normal.

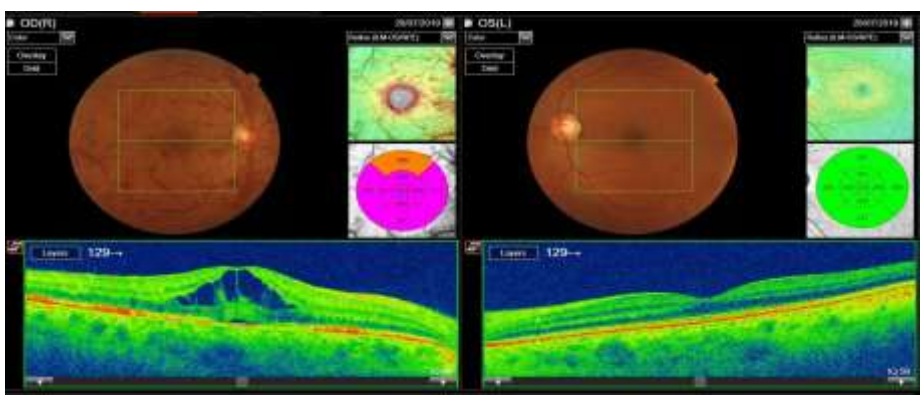
Investigations:- CBC, Blood Glucose (R), CRP, ESR, , lipid profile, plasma homocystein, Plasma Protein,cloting and bleeding time , Serum Creatinine , ECG.

Treatment:- Intra vitreal Lucentis (ranibizumab) injection 0.05ml was given to the patient and Monthly Follow up was done to check the recurrence of macular edema and Development of any Neovascularization.

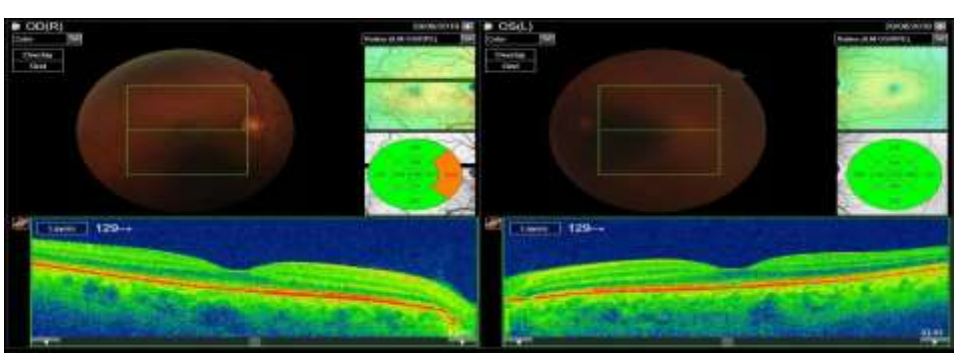
Fundus Photo Of Central retinal Vein Occlusion



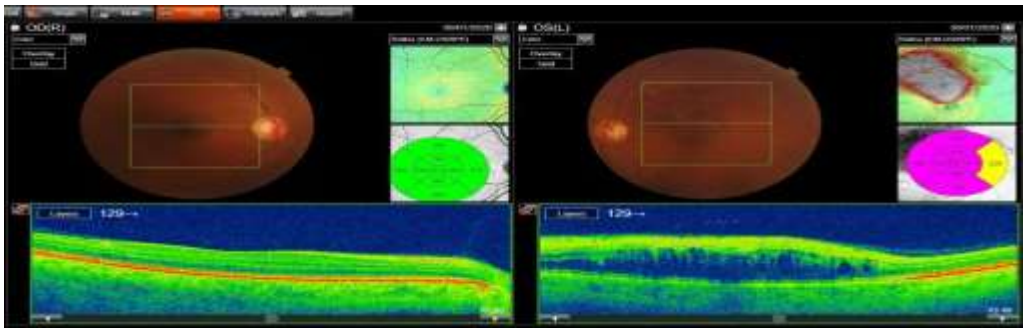
OCT Picture Of Central retinal Vein Occlusion before Intra vitreal Injection



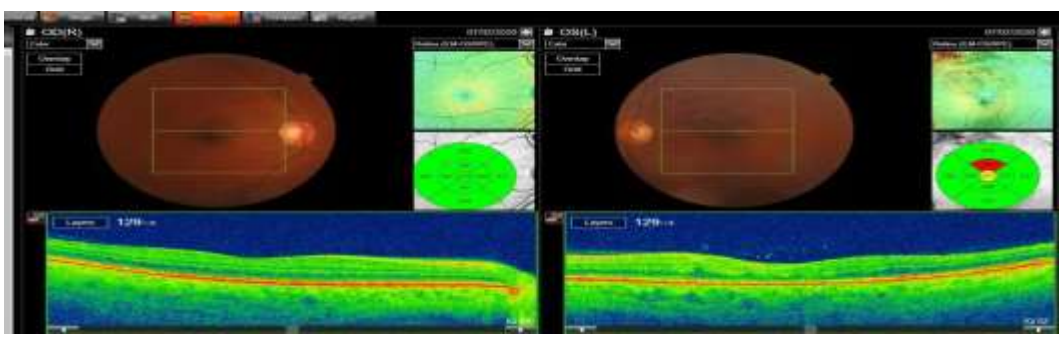
OCT Picture Of Central retinal Vein Occlusion after 1 Month of Intra vitreal Injection



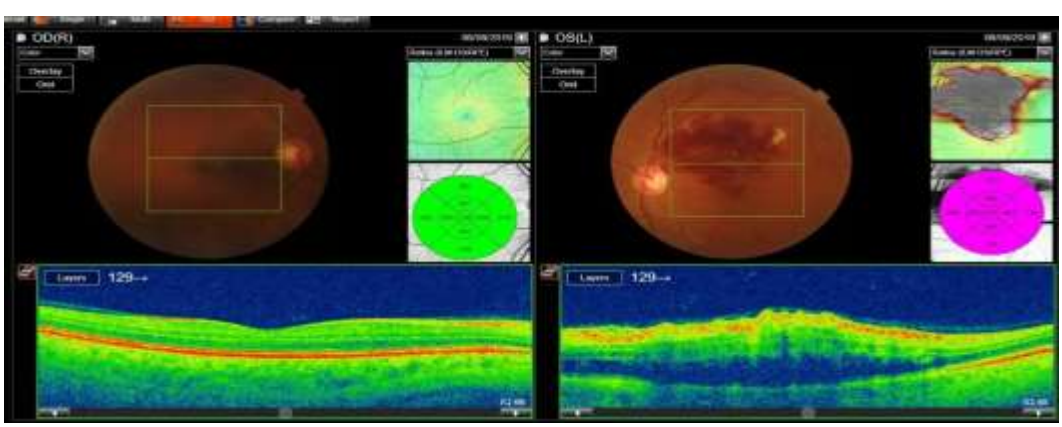
Another OCT Picture of venous occlusion Before Intra Vitreal Injection



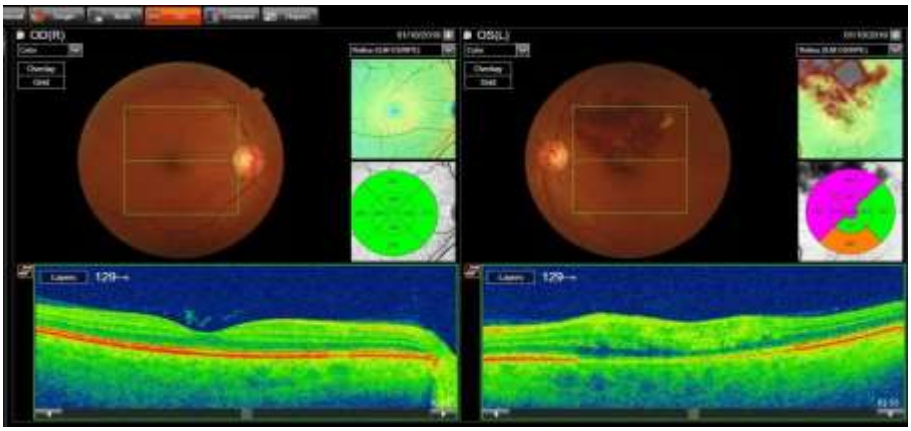
Another OCT Picture of venous occlusion After Intra Vitreal Injection



Another OCT Picture of venous occlusion Before Intra Vitreal Injection



Another OCT Picture of venous occlusion After Intra Vitreal Injection



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