Central Retinal Vein Occlusion of the Young: A Profile

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Abstract: <u>Background and objective</u>: In contrast to the central retinal vein occlusion (CVO) seen in the adults, its affection in nondiabetic and non - hypertensive young people has been limited. Are these two different entities or one and the same? <u>Methods</u>: Fifty eyes of 43 non - diabetic and non - hypertensive patients with age less than 40 years who had CVO were examined. <u>Results</u>: There were 36 males (83.72%) and 07 females (16.28%). Mean BCVA at presentation was 6/60 and 6/18 at final follow - up after 60 months.29 patients (67.44%) were less than 35 years of age. Bilateral affection was seen in 7 (16.28%).13 patients (30.23%) had RAPD. Keratic precipitates were seen in 5 eyes (10%). Neovascularisation of the iris was seen in 11 eyes (22%), cystoid macular edema in 32 eyes (64%) and retinal neovascularisations were noted in 25 (50%) eyes Neovascularisation of the disc was noted 14 eyes (28%) but secondary glaucoma was seen only in 11/50 eyes (22%). Systemic associations included active pulmonary tuberculosis (8%), toxoplasmosis (4%), multiple myeloma, Hodgkin's lymphoma and Waldenstrom's macroglobulinemia (2% each). Eyes developing NVD and NVE were subjected to laser photocoagulation. Clinical improvement was noted in 27 eyes with improvement in the final best corrected visual acuity noted in 14 eyes (P<0.05). <u>Conclusion</u>: CVO of the young has a varied different presentation than in adults. A thorough evaluation is a must.

Keywords: Central Retinal Vein Occlusion In Young, scatter photocoagulation, hyperviscosity syndrome, systemic syndrome

1. Introduction

Central retinal vein occlusion is commonly seen in patients above 50 yrs of age with underlying hypertension and or diabetes as the causative agent. CVO occurring in young people without underlying diabetes or hypertension has been termed as CVO of the young (CVOY) and has previously been presumed to be of inflammatory origin leading [^{1]} to terminology such as papillophlebitis and benign retinal vasculitis but now it has been known to have multifactorial etiology - mutation, drugs, blood disorders, systemic diseases etc ^{[2] [3]}.

Objective: Is CRVO is same as CRVY?

2. Materials and Methods

Any case of CVO less than 40 yrs of age was taken up for study. Patients were firstly evaluated for diabetes and hypertension and if any one of them was detected the case was excluded from the study. The patients were also subjected to the following investigations:

- 1) CXR for any evidence of Koch's disease
- 2) Mantoux testing
- 3) ELISA for tuberculosis
- 4) ELISA for toxoplasmosis

Patients with systemic disorders who were found to have CVO were also included in the study if they were less than 40 yrs of age and were non - hypertensive and non - diabetic.

The follow up of the patients included the following:

- 1) BCVA
- 2) Slit lamp Biomicroscopy for detection of keratic precipitates and/or NVI
- 3) Goldman Applanation Tonometry
- 4) Detailed ophthalmoscopy under mydriasis
- 5) Fundus fluorescein angiography when felt appropriate

Eyes developing NVD and NVE were subjected to photocoagulation with either Diode or Frequency doubled Nd YAG laser.

3. Results

Demographics (Table 1)

There were a total of 50 eyes of 43 patients who had CVOY. The ages ranged from 18 years to 40 years. There were 36 males (83.72%) and 07 females (16.28%). Two thirds of the patients [29 patients (67.44%)] were less than 35 years of age. Right eye was affected in 19 patients (44.19%), left eye in 17 (39.53) and in 7 patients (16.28%) it was bilateral.

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Tuble 1. Demographies				
Age range	Male	Female	Total	%
16 - 20	01	01	02	04.65
21 - 25	-	01	01	02.32
26 - 30	14	01	15	34.89
31 - 35	09	02	11	25.58
36 - 40	12	02	14	32.56
Total	36 (83.72%)	07 (16.28%)	43	100

Table 1: Demographics

Clinical features

- 1) Mean BCVA at presentation was 6/60 and 6/18 at final follow up after 60 months.
- About one third of the patients had RAPD [13 patients (30.23%)].
- 3) Keratic precipitates were seen in 5 eyes (10 %).
- 4) One fifth of the eyes had neovascularisation of the iris [11 eyes (22%)].
- 5) Two thirds of the eyes had cystoid macular edema [32 eyes (64%)]
- 6) One fourth of the eyes had retinal neovascularization. [25 (50%) eyes]
- Neovascularisation of the disc was noted 14 eyes (28%) but secondary glaucoma was seen only in 11/50 eyes (22%).
- Systemic associations included active pulmonary tuberculosis four patients (9.3%), toxoplasmosis in 02 patients (4.65%), multiple myeloma, Hodgkin's lymphoma and Waldenstrom's macroglobulinemia in one each (2.32% each).

Eyes developing NVD and NVE were subjected to scatter photocoagulation. Clinical improvement was noted in 27 eyes with improvement in the final best corrected visual acuity noted in 14 eyes (P<0.05).

4. Discussion

The study to be compared is the CVO Study (CVOS) [^{4]}. Over half of the patients in CVOS were more than 65 years of age whereas in this study 2/3 of the patients were less than 35 years of age. It has also to be noted that in the CVOS there were patients with 20 years of age and above [^{4]}. However, they have not been analysed separately. Owing to the rarity of the condition in young but being more susceptible to changes in visual acuity, results in discrepancies in the number reported. This study aims at removing this confounding factor. Moreover, CVO could point towards bimodal distribution in >65 years and <35 years, reason being atherosclerosis and hypertension in former and increase in the manifestations of hyper viscosity syndrome complications in the latter group.

In the CVOS there were slightly more males but in CVOY group the males were far more in number (83.72%). Similar observations were found in studies conducted by Nalcaci et al, Vieira M. J. et al and Koh YY et al ^{[5] [6] [7]}.

This may be due to dehydration by outdoor work in male counterparts, causing progress of manifestations of hyperviscosity syndromes^[8].

The mean BCVA in the CVOS was 6/24 whereas in this study it has been 6/60 suggesting that the base - line visual

acuity is worse in CVOY compared to CVO. This may be due to paracentral acute middle maculopathy (PCAMM) more common in young compared to older age that may have been masked at the time of presentation by hemorrhage and macular edema^[9].

Iris neovascularisation was seen in 16% of the eyes in CVOS which is lesser than 22% seen in this study. The retinal vascular plexus is led by the variation between the mean arterial and venous pressures because as pulse pressure difference is lower in young with CVO, the deep capillary plexus is more vulnerable to hypoperfusion resulting in greater release of pro angiogenic factors with greater risk of neovascularisation in young^[3].

It might have to be indirectly concluded that the incidence of secondary glaucoma was more in CVOY than in CVOS, as the latter study has commented on the regression of iris neovascularisation with laser treatment but not on actual numbers except for the ten uncontrolled cases of secondary glaucoma (1.4%). The CVOS has not studied RAPD. By this comparison it does appear that CVOY has a graver presentation in comparison. The same has been corroborated by AC Fong et al [10] in their study of CVOY. Fong et al had studied their patients for 06 months, had male preponderance (64%), but lesser NVE (18%), NVD (18%) and NVI (19%) than the present study. The final BCVA in the study of Fong et al was also bad at 6/60, whereas it has been at 6/18 in the present study. Could this be ascribed to the institution of steroids in idiopathic cases? Shaikh and Blumenkraz^[11] had treated only two patients and with only transient improvement. In this study the number of treated cases has been 38 and with good results.

It is also to be noted that the systemic disorders are also varied in this group.

These systemic disorders lead to hyperviscosity syndrome which are more common in age <45, thus leading to increased thrombosis and causing venular obstructions. A study conducted by Zhang X et al discussed various disorders leading to increase in risk of thrombosis leading to development of CRVO in young and all these pointed towards systemic syndromes in consistent with our study ^[3].

5. Conclusion

CVO of the young has a varied and diverse presentation than in adults. A thorough evaluation is a must. In the treatment of idiopathic cases, systemic administration of steroids is useful with significant improvement noted in the final BCVA at a mean follow - up of 60 months. Therefore, it concludes active intervention in these cases, would have a better prognosis at follow - up.

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