A Rare Cause of Unilateral Central Retinal Vein Occlusion in a Young Patient with Polycythemia Vera

Authors
Dr Shashikant Singh¹, Dr Suchita Singh², Dr Abhijit Kumar Handique³
¹MBBS MD Medicine 2nd year SR (Academic) DM Cardiology AIIMS Bhubaneswar
²MBBS 3rd year JR (Academic) MS Ophthalmology Assam Medical College Dibrugarh
³Assistant Professor Dept of Ophthalmology Assam Medical College Dibrugarh

Introduction
Retinal vein occlusion is a common cause of vision loss in older individual, and the second most common retinal vascular disease after diabetic retinopathy[1]. In CRVO, the occlusion is at or proximal to proximal to the lamina cribrosa[2]. Age is the most important risk factor, since over 90% of cases occur in patients over the age of 55 years[3]. A minority of patients with CRVO are young adults under the age of 40 years and in most of cases CRVO occurs without the presence of the typical risk factors[4].

Case Report
A 21 year old female patient presented with sudden onset painless diminution of vision L/E for 2 days [fig 1]. It was not associated with any headache or any discharge. There was no history of similar episode in same or other Eye in the past, no history of trauma to eye.No history of any systemic illness or any medication intake. Family history was not significant. [fig 1]

Fig 1

Examination
General examination was within normal limit.

Fig 2
- **Fundus Examination**: [fig 2]
  - Blurred disc margin i.e disc edema
  - CDR=0.4:1
  - multiple superficial flame shaped haemorrhages with cottonwool spots
  - dull foveal reflex
  - Dilated and tortuous retinal veins.

- **FFA**: [fig 3]
  - Delay in arteriovenous transit time
  - Blocked fluorescence by retinal hemorrhages.
  - Vessel wall staining.
  - Few small area of retinal capillary obliteration.
  - Areas of non-perfusion.

- **OCT Macula**
  Subretinal fluid with small cystic changes within neurosensory retina i.e Macular edema. [fig 4]

---

**Fig 3**

**Fig 4**

**Lab Investigation**: R/E blood revealed R.B.C : 7.8 million/mm3, Hb : 19gm/ dl, HCT : 55.6%, MCV: 123.5 fl, MCH: 42.2 pg , MCHC: 34.2% , TLC: 14,200/ mm3, platelet count :673000/ mm3 and serum erythropoietin level was decreased. Other investigations were found to be normal.

**Bone Marrow Biopsy**: Showed hypercellularity for age with trilineage growth. [fig 5]

**Fig 5**

**Fig 6**

**Final Diagnosis**
Polycythemia Vera with Unilateral Nonischemic Central Retinal Vein Occlusion with Macular Edema Left Eye.

**Treatment**
For polycythemia vera, patient was referred to higher centre in Guwahati.
For crvo, patient was treated with three doses of intravitreal Bevacizumab each 1 month apart for 3 consecutive months

**Follow – Up**
After 12 weeks of completion of treatment VA L/E was 6/6 with normal fundus examination findings.
Discussion
Unusual causes for CRVO in young patients include hypercoaguibility states, collagen vascular disease, lymphoproliferative disorders, malignant hypertension, medication (ocp, tretinoin) and trauma.\[5\]

A recent study has demonstrated that hyperlipidemia and hyperhomocysteinemia are the most significant risk factors for crvo in patients under 40 years.\[6\]

Polycythemia vera (PV) is a rare myeloproliferative neoplasm associated with an increased production of red blood cells, white blood cells and platelets. Vascular thrombosis is the most frequent cause of death in PV patients.\[7\]

Ocular complications are secondary to hyperviscosity and thrombosis.\[8\]
Thrombotic events are present in 20-50% of patients with PV at diagnosis and involve major vessels and microcirculation.\[9\]
Isolated monocular blindness as the presenting feature of PV has been presented only four times indicating rarity of presentation.\[10\]
The devastating outcome of such vascular occlusions warrants a close follow-up in patients with PV to prevent visual loss.

References