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

Authors
Dr Shashikant Singh[1], Dr Suchita Singh[2], Dr Abhijit Kumar Handique[3]
[1]MBBS MD Medicine 2nd year SR (Academic) DM Cardiology AIIMS Bhubaneswar
[2]MBBS 3rd year JR (Academic) MS Opthalmology Assam Medical College Dibrugarh
[3]Assistant Professor Dept of Opthalmology 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]

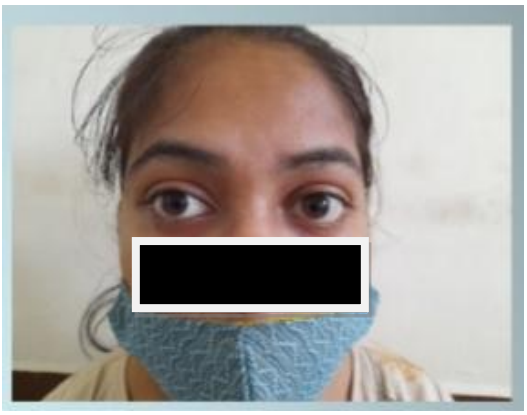

Fig 1

Examination

General examination was within normal limit.

EXAMINATION	OD	OS
BCVA:	6/6, N6	6/60
EYELIDS:	WNL	WNL
CONJUNCTIVA:	WNL	WNL
CORNEA:	Clear	clear
PUPILS:	RRRL	RRRL
IOP (Atn) :	16 mmHg	17mmHg
Slit lamp examination	WNL	WNL
Gonioscopy	Angle open	Angle open

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 tortouus retinal veins.

Fig 3

- **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.

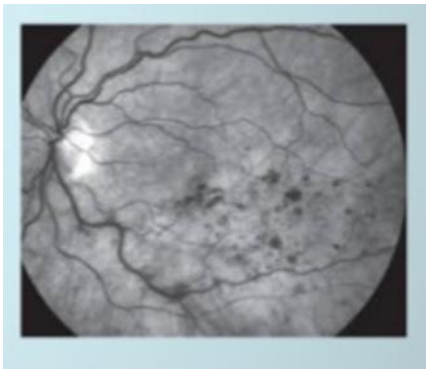

Fig 4

- **OCT Macula**

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

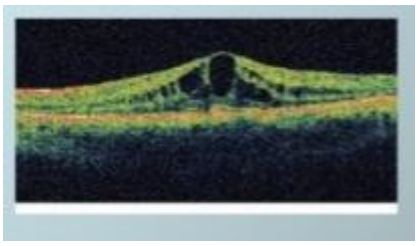

Fig 5

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]

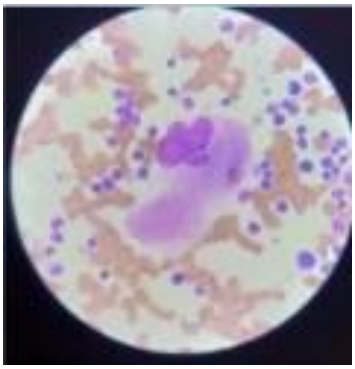

Fig 6

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 complition of treatment VA L/E was 6/6 with normal fundus examination findings.

Discussion

Unusual causes for CRVO in young patients include hypercoagubility 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 myloproliferative 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 rareity of presentation.[10]

The devastating outcome of such vascular occlusions warrants a close follow-up in patients with PV to prevent visual loss.

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