

# UNIQUE CASE OF DEVELOPMENT OF CENTRAL RETINAL VEIN OCCLUSION IN DIABETIC PAPILOPATHY

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## ABSTRACT

Diabetic papillopathy (DP) is a rare ocular complication of diabetes presenting as acute disc edema. It is characterized by minimal or no visual symptoms and has tendency to resolve spontaneously in most of the patients with no visual sequel. Visual debility, if present, is attributable to diabetic macular edema (DME), advanced diabetic retinopathy or conversion to ischemic optic neuropathy. We report a case of 33 years old female who presented with bilateral papillopathy with best corrected visual acuity (BCVA) of 6/6 in both eyes. She had associated severe non proliferative diabetic retinopathy but no DME. On follow up, the patient had good metabolic control and clinical improvement in DP, but she developed sudden diminution of vision in left eye as a result of non ischemic central retinal vein occlusion (CRVO) which was characterized by BCVA of 6/36, dilated tortuous veins and macular edema. No specific cause of non-ischaemic CRVO was revealed on detailed work up. Structural and functional improvement was seen on monthly intravitreal anti vascular endothelial growth factor (VEGF) administration. Her BCVA post three injections stood at 6/18. This is to report the possibility of development of CRVO in DP and highlight the need for close follow-up of such patients even in presence of strict diabetic control.

## KEY WORDS

Central retinal vein occlusion, diabetic retinopathy, diabetic macular edema, ischaemic optic neuropathy, vascular endothelial growth factor



## INTRODUCTION

Diabetes mellitus (DM) is responsible for a myriad of ocular morbidities of which posterior segment involvement is seen in diabetic retinopathy (DR) and diabetic papillopathy (DP). While DR is more common and has high propensity of causing visual impairment, DP is rare and has a benign course. It is transient, acute onset disc edema in a known diabetic with absence of substantial optic nerve dysfunction. Initial investigators reported it to occur bilaterally in young patients with type 1 DM, but later it was reported in older patients with type 2 DM and was known to have unilateral presentation too.<sup>1,2</sup> Sudden blood sugar control and small cup disc ratio are known risk factors for its occurrence. DP is a diagnosis of exclusion where other infectious, inflammatory and infiltrative causes of disc edema as well as increased intracranial tension have been ruled out. These patients have no or minimal visual symptoms comprising of painless mild blurring or distortion of vision. It has an excellent prognosis with spontaneous resolution in 2-10 months.<sup>2-4</sup> Visual impairment if associated is because of coexisting macular edema or proliferative diabetic retinopathy.<sup>5</sup> Poor outcome is seen in cases who convert to non arteritic ischemic optic neuropathy.<sup>6</sup> We report a case of DP which converted to central retinal vein occlusion (CRVO) on follow up with acute deterioration of vision. To the best of our knowledge, there is no such association reported in literature till date.

## CASE DESCRIPTION

A 33-year old diabetic (type 2 DM) female presented with blurring of vision in both eyes (OU) for days. She was non hypertensive and had history of diabetes for 3 years. Her blood sugar was raised to 396mg% which was aggressively lowered down to 160mg% with insulin initiation 20 days back. She had normal weight (BMI-25.3) and had no other systemic co-morbidities. There was no feature of increased intracranial pressure like nausea, vomiting or headache. Her best corrected visual acuity (BCVA) was 6/6 OU. No relative afferent pupillary defect (RAPD) was detected. Color vision and contrast sensitivity were normal. Fundus examination showed hyperaemic disc swelling with superficial, radially oriented, telangiectatic vessels and features of severe non proliferative diabetic retinopathy (NPDR) which was confirmed by fluorescein angiography (FFA). OCT macula and visual field were normal. MRI brain and orbit was normal with no intracranial space occupying lesion (SOL) or features of pseudotumor cerebri or multiple sclerosis. Infective and inflammatory causes of bilateral disc edema were also ruled out by battery of tests. Apart from post prandial blood sugar which was 202 mg/dl, all her tests were within normal limits. The patient was diagnosed as a case of both eyes severe NPDR with DP and strict systemic control was planned.

On two months follow up, BCVA OU was 6/6, disc remained hyperemic, with resolving disc edema. Her blood sugar was in control on continued insulin therapy. She visited a month

later with sudden diminution of vision left eye (OS). Her BCVA was 6/36 in OS. Her pupillary reaction, color vision, contrast sensitivity and visual field remained normal. She had developed non ischaemic CRVO with macular edema measuring 837 $\mu$  (central macular thickness [CMT] by OCT) in left eye. She underwent thorough investigations to rule out causes of CRVO in young which included kidney and liver function test, bleeding and clotting time, prothrombin and activated plasma thromboplastin time, Protein C, Protein S, Antithrombin 3 and homocysteine. Connective tissue markers were checked and a thorough cardiac evaluation was done. All her reports were normal. Obstructive sleep apnea syndrome (OSAS) was excluded by absence of nocturnal snoring, mid night awakening or day time somnolence.

Monthly intravitreal anti Vegf were given in left eye and CMT reduced from 837 $\mu$  to 293  $\mu$  with no features of macular atrophy. Her BCVA improved not better than 6/18, the cause of which could not be explained.

## DISCUSSION

CRVO in young is uncommon. It is usually non ischemic and needs detailed work up. However, it is found to have good prognosis.<sup>7</sup> Diabetes, hypertension, hypercholesterolemia are known traditional risk factors in older patients. Younger patients would not consistently exhibit these risk factors and instead harbour many non-traditional causes for CRVO like obstructive sleep apnoea, hypercoagulability, homocysteinemia, connective tissue disease or cardiac disease.<sup>8</sup> Our case is diabetic but proper work up was done to exclude these non-traditional causes and head injury was also ruled out.

Our case had onset of DP in both eyes 3 months back following aggressive blood sugar control which is a known risk factor for development of DP.<sup>2</sup> Diagnosis of DP was established by excluding other causes of bilateral disc edema like optic neuritis, NAION, papilloedema due to SOL or malignant hypertension and benign intracranial hypertension. Optic neuritis and NAION were excluded by absence of clinical signs of optic nerve dysfunction with normal visual field and MRI. Absence of features of increased intracranial pressure and normal MRI excluded papilloedema. BIH was also ruled out clinically and by normal BMI and MRI. Refusal of lumbar puncture by our patient is a limitation of this study. Papillophlebitis can also be ruled out as it was bilateral and was resolving spontaneously before progressing to CRVO.

Several optic nerve head (ONH) conditions like ONH drusen, ONH glioma, pseudotumor cerebri, optic neuritis and AION give rise to CRVO as secondary phenomenon.<sup>9,10</sup> Duker reported 5 cases of ONH edema in optic neuritis leading to secondary venous stasis giving rise to CRVO due to mechanical compression.<sup>11</sup> There has been reports of tumor infiltration of disc in ONH glioma and several reports of ONH drusen causing CRVO by direct pressure effect. Doesschate et al



histopathologically proved ONH edema, CRVO and optic atrophy occurring in ONH drusen.<sup>9</sup> Hayreh in his experimental model of induced optic neuritis in rhesus monkey found vaso-occlusive retinopathy occur in 5 of 12 cases. This experiment proved ONH edema under certain circumstances does result in secondary CRVO.<sup>12</sup>

As per our best knowledge CRVO in case of DP has yet not been reported or discussed. Association of NAION with CRVO has been reported in two cases by Ahmed et al. and one case by Kim et al.<sup>13,14</sup> All these 3 cases had hypercoagulable state. We discuss this because DP may convert to NAION in few patients. Infact, few investigators believe DP to be a part of NAION spectrum but this still remains controversial in light of difference in the pathology of two entity.<sup>15</sup> Even if DP is considered as an incipient NAION these 3 cases had one or other cause of hypercoagulability which may have resulted

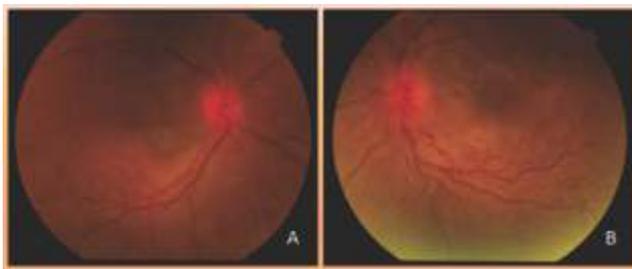
in CRVO. In addition to this, hypercoagulability itself is a risk factor for development of NAION. So these cases showed the role hypercoagulable state causing NAION which then in unison resulted in CRVO.

## CONCLUSION

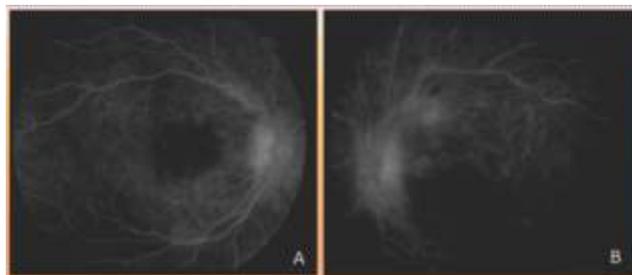
Our report describes an unusual association between CRVO and DP. DP in young even in absence of other risk factors may lead to CRVO. We believe that this is because diabetes itself is risk factor for CRVO which is complemented in DP by direct compression of central retinal vein by swollen disc. So the need of close follow up of all patients of DP even if they are under good metabolic control cannot be over emphasised in view of risk of developing CRVO.

## CONFLICTS OF INTEREST

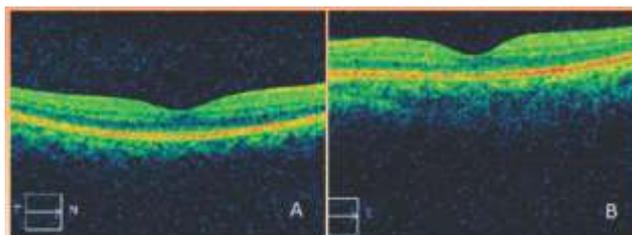
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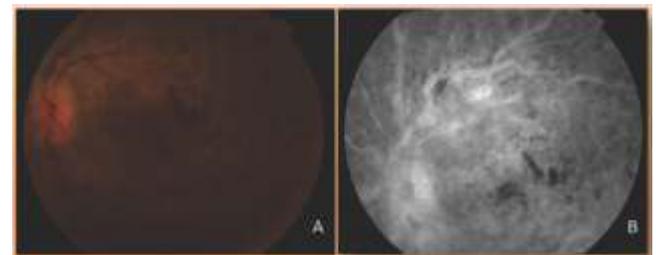
**Figure 1:** A, B : Colour fundus photograph at presentation showing bilateral diabetic papillopathy and severe Non Proliferative Diabetic Retinopathy (NPDR)



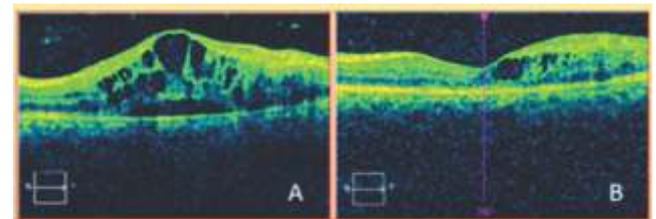
**Figure 2:** A, B - Fluorescein Angiography (FFA) of right and left eye showing disc hyper fluorescence and leakage, blocked fluorescence due to dot blot haemorrhage and nerve fibre layer haemorrhage in all quadrants; suggestive of diabetic papillopathy and Severe NPDR



**Figure 3:** Optical coherence tomography (OCT) both eye at presentation showing absence of macular edema



**Figure 4:** A : Colour fundus photo at 3 month follow up showing dilated tortuous veins with disc edema and dot blot as well nerve fibre layer haemorrhage in all quadrant. B : FFA left eye at 3 month follow up showed markedly dilated, tortuous veins with delayed AV transit. Late film had vessel wall staining and perivascular leakage suggestive of central retinal vein occlusion



**Figure 5:** A – OCT left eye at 3 month follow up showing central subfield thickness (CST) of 837 $\mu$   
B – OCT left eye after 3 intravitreal Antivegf injections showing remarkable decrease in macular edema to CST of 297 $\mu$  with few remaining cystic spaces.

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