Case Report

Variable presentation of diabetic papillopathy and treatment dilemma- A case series

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Abstract

Introduction: Diabetic papillopathy (DP) is a diagnosis of exclusion in type 1 and type 2 diabetics with transient disc edema. It was initially described in young patients with type1 diabetes mellitus (DM) as a bilateral disease with minimal visual symptoms which resolved spontaneously. Lately, DP has been a focus of controversy because of its wide clinical spectrum.

Cases: We describe three variable cases of DP. These are unilateral DP with Proliferative Diabetic Retinopathy (PDR) with macular edema (ME), unilateral DP with severe Non Proliferative Diabetic Retinopathy (NPDR) with ischemic maculopathy and a case of bilateral DP with Moderate NPDR with ME. We also discuss viable treatment for the variable presentation.

DP has been reported in this case series in moderate NPDR, severe NPDR as well as PDR. Macular involvement in the form of macular edema as well as ischemia has been demonstrated to result in diminution of vision. It shows both unilateral and bilateral presentation. Remarkable visual loss seen, in these cases, call for intervention.

Conclusions: DP has a wide spectrum of presentation and its knowledge is eminent to make a complete diagnosis. Individualisation of treatment has to be done for variable presentation and realistic outcomes should be explained to the patients.

Key word: Diabetic Papillopathy, Moderate Non Proliferative Diabetic Retinopathy, Severe Non Proliferative Diabetic Retinopathy, Proliferative Diabetic Retinopathy.

Introduction

Diabetic papillopathy (DP) is a rare ocular disease in diabetic patients (Bayrktar Z, 2002). According to Appen (1980) diagnostic criteria of DP was classically described as - confirmed case of diabetes with disc edema and absence of substantial optic nerve dysfunction, evidence of ocular inflammation or elevated intraocular pressure (Appen RE, 1980). DP presents with acute onset transient disc edema with telangiectatic vessels which shows early disc hyperfluorescence and late leakage in Fundus Fluorescein Angiography (FFA). However, lately its wide clinical spectrum has

Financial Interest: Nil
Conflict of Interest: Nil
Received: 07.10.2019 Accepted: 01.02.2020

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been observed. There is no treatment guideline available depending on varied presentations. We present a series of 3 cases with variable presentation of DP.

Case details

Case 1

A 49-year male presented with diminution of vision in his left eye (LE) for 3weeks. He was a type 2 diabetic and hypertensive, well controlled on oral medication. His best corrected visual acuity (BCVA) at presentation was 20/30 in right eye (RE) and 20/120 in LE. Slit lamp examination revealed no ocular inflammation. Pupillary reflex examination revealed absence of relative afferent pupillary defect (RAPD). Colour vision and contrast was normal in RE, these tests however could not be done in LE due to poor VA. Patient failed to cooperate for visual field (VF) evaluation. RE had moderate Non Proliferative Diabetic Retinopathy (NPDR). LE showed disc edema and features of Proliferative Diabetic Retinopathy (PDR). Neurological examination and computed tomography (CT) of head was normal. FFA of left eye revealed blurred disc margin and telangiectatic vessels over the disc with early disc hyper fluorescence and late leakage suggestive of disc edema, absence of neovascularization of disc (NVD), presence of neovascularization along infero-temporal arcade (NVE) with leakage at macula. Disc leakage and filling defect was absent. Central subfield thickness (CMT) measured by Optical coherence tomography (OCT) was 504 µ. Patient was then diagnosed as Right eye moderate NPDR and left eye DP with Right Proliferative Diabetic Retinopathy with macular edema (ME) (Figure 1). Left eye was treated with anti-vascular endothelial growth factor (anti-VEGF) intravitreal injection and Pan-retinal photocoagulation in view of PDR. DP resolved in 3 months. Neovascularization regressed and BCVA improved to 20 /30.

Case 2

A 45-year-old non hypertensive, type 2 diabetic male on insulin therapy (9 years) presented with diminution of vision in his right eye for 7 months. His BCVA was 20/400 and 20/30 in RE and LE respectively. Colour vision, contrast sensitivity and visual field of the left eye were normal, whereas these tests could not be done in RE due to poor VA. RAPD was present. Fundus examination revealed disc edema in right eye and severe NPDR in both eyes. CT scan head was normal. FFA confirmed right eye DP and ischemic macula (suggested by irregular and enlarged foveal avascular zone). OCT revealed flattening of foveal contour and loss of ellipsoid zone in the same eye. This case was then diagnosed as both eye severe NPDR and right eye DP with ischemic maculopathy (Figure 1). Patient was kept on observation with regular follow up. DP resolved in 4 months but there was no functional improvement due to pre-existing macular ischemia.

Case 3

A 38-year-old type 2 diabetic female on oral hypoglycemic agent (OHA) for 12 years had diminution of vision of both eyes for 2weeks. BCVA was 20/40 and 20/80 in RE and LE respectively. She was non hypertensive but had a history of sudden blood sugar (BS) control with insulin therapy started 3 weeks prior to the development of the symptoms. Colour vision and contrast was normal in both eyes. No RAPD was detected. VF was normal. Both eyes had disc edema with moderate NPDR with ME. Neurological evaluation and computed tomography of the head was normal. Work up for connective tissue disorder, blood counts and sedimentation rate was normal. On complete evaluation, the patient was diagnosed as bilateral DP with Moderate NPDR and ME (Figure 2). Bilateral ME prompted us to treat her with anti-VEGF for 3 consecutive months which resulted in dry macula, resolution of DP and 20/40 VA in each eye. The patient lost to follow-up thereafter.

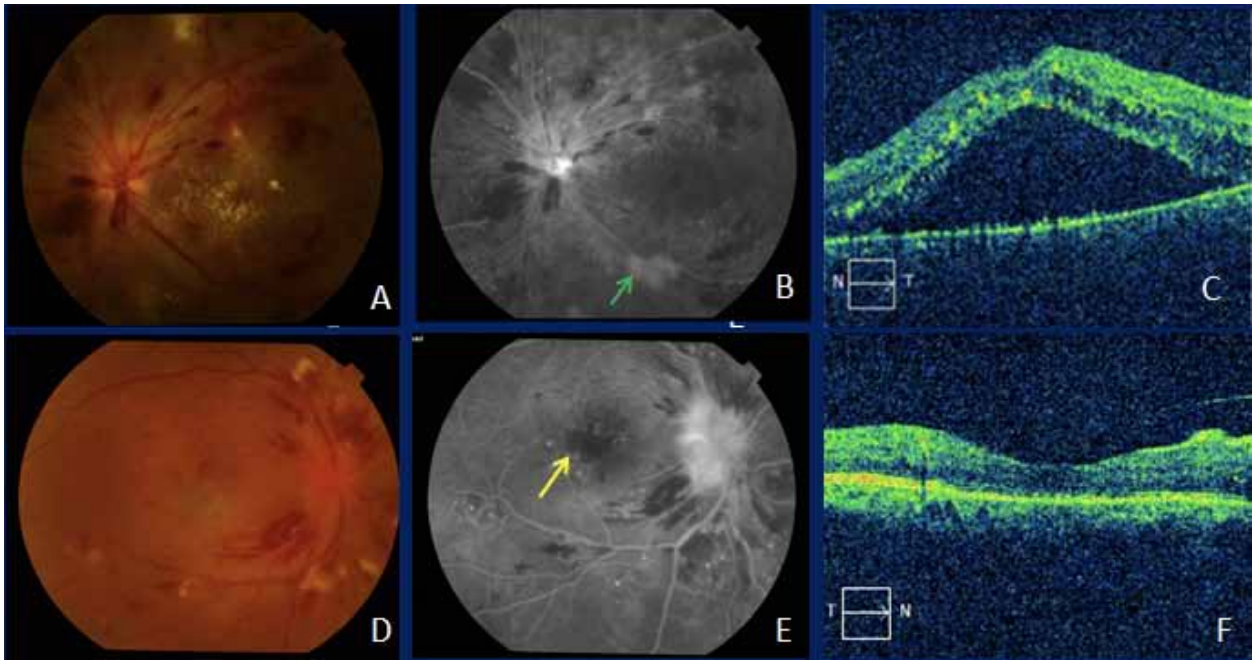

Figure 1: A,B,C: A case of 49 year, male with left eye DP with PDR and ME

A: Colour fundus photo showing disc edema with haemorrhage in all quadrant and hard exudate in macula

B: FFA showing DP characterised by telengectatic vessels and early disc hyperfluorescence. NVE shown by green arrow along inferotemporal arcade suggestive of PDR

C: OCT shows macular edema. Subretinal fluid is present with CMT 504µ

D,E,F: A case of 45year, male with right eye DP with ischemic maculopathy

D: Colour fundus photo showing disc edema with extensive peripapillary haemorrhage and cotton wool spots in all quadrant suggestive of severe NPDR

E: FFA is suggestive of DP. Enlarged foveal avascular zone is pointed with yellow arrow implying ischemic macula

F: OCT macula shows foveal thinning with absence of subfoveal ellipsoid zone as sign of ischemic maculopathy

Abbreviations - DP: Diabetic papillopathy, PDR: Proliferative diabetic retinopathy, ME: Macular edema, FFA; Fundus fluorescein angiography, NVE: Neovascularization elsewhere, OCT: Optical coherence tomography, CMT: Central macular thickness, SRF: Subretinal fluid

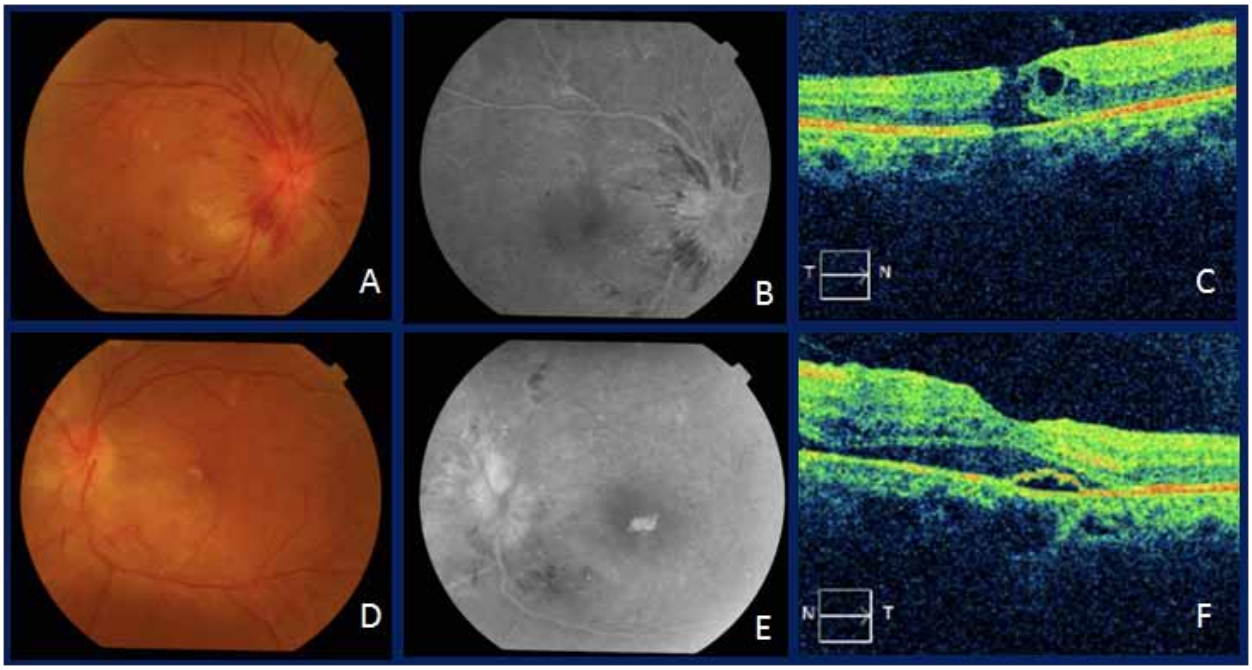

Figure 2: A case of 38 year, female with both eye DP with moderate NPDR with ME

A, D: Colour fundus photo showing disc edema right eye more than left eye with features of moderate NPDR

B, E: FFA showing DP characterised by telangiectatic vessels and early disc hyperfluorescence.

C, F: OCT macula shows SRF and intraretinal fluid in both eyes. CMT right eye 429µ and left eye 402µ

Abbreviations- DP: Diabetic papillopathy, PDR: Proliferative diabetic retinopathy, ME: Macular edema, FFA; Fundus fluorescein angiography, NVE: Neovascularization elsewhere, OCT: Optical coherence tomography, CMT: Central macular thickness, SRF: Subretinal fluid

Discussion

Diabetic papillopathy as an entity has been mentioned in literature since 1980, although similar cases were described as early as 1971. The prototype was described first by Appen et al (1980). This initial description has been edited widely thereafter by different investigators. DP was initially described in juvenile young diabetic type 1, with bilateral presentation, usually having no or little visual symptoms (Appen RE, 1980). Lately it has also been reported in type 2 diabetes mellitus (DM), elderly DM patients and is known to be unilateral in 50% cases (Bayrktar Z, 2002) (Regillo CD, 1995) (Pavan PR, 1980). All cases in the present series were of type 2 DM, involvement in 2 cases was unilateral and the 3rd one was bilateral.

Our series of DP had one PDR and 2 NPDR. Earlier literatures have suggested minimal fundus finding associated with DP, but now its association with different stages of DR has been established and is reported to be 54% in NPDR and 9% in PDR (Bayrktar Z, 2002) (Regillo CD, 1995) (Eric K. Chin, 2015).

Small cup disc ratio and sudden aggressive control of diabetes are known risk factors for DP (Appen RE, 1980). Similarly, in our series, one patient had a history of aggressive blood sugar control.

DP was earlier believed to have no visual sequel but now we know that it may have poor visual acuity due to worsening of DR (17%) or ME (Bayrktar Z, 2002). All cases in our series had poor visual acuity, 1st and 3rd case due to associated macular edema (ME) and 2nd case had macular ischemia. Bayraktar et al (2002) described that 25% of type 2 DM patients may have ME with DP (Bayrktar Z, 2002). We could not find any case of DP associated with macular ischemia in literature.

In light of the wide clinical spectrum of DP, the biggest challenge is its management in the absence of a definite treatment protocol. DP is considered to be self-resolving in 2-10 months (Barr CC, 1980) (Pavan PR, 1980). Scholars practiced serial examination with strict systemic control as the only treatment for years. Recently, intravitreal and periocular steroids and various anti Vascular Endothelial Growth Factors (anti-VEGF) intravitreal injections have been used in the treatment of DP (Pavan PR, 1980) (Eric K. Chin, 2015) (Barr CC, 1980) (Al-Haddad CE, 2004) (Ornek K, 2010) (Kim M, 2013). The aim of treatment is to hasten visual recovery in bilateral cases with poor VA and one eyed patients. In the present series, apart from blood sugar control, case 1 and 3 received ocular treatment whereas case 2 was simply kept on observation. DP resolved in all the cases. There was one report of severe NPDR with DP treated with Anti-VEGF and PRP causing faster resolution of DP (Lovestam-Adrian M, 2003). However, the role of intervention has been strongly implicated in patients where DP is associated with ME or PDR which itself warrants treatment.

Conclusion

It is critical for the caregiver to frame a complete diagnosis of DP with respect to history, unilateral or bilateral involvement, extent of visual impairment, stage of DR and involvement of macula. Individualisation of treatment has to be done for variable presentation, and realistic outcomes should be explained to the patients.

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