CASE REPORT

A Rare Case of Diabetic Papillopathy Presenting with Pseudo-Foster Kennedy Syndrome

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ABSTRACT

Pseudo-Foster Kennedy syndrome (PFKS) is defined as unilateral disc oedema with contralateral optic nerve pallor, in the absence of intracranial pathologies. Diabetic papillopathy is a rare ocular manifestation of diabetes mellitus (DM). We report a rare case of PFKS secondary to diabetic papillopathy in a young patient with type 2 DM who had poor glucose control. There was optic disc swelling over the right eye and optic disc pallor over the left eye. His visual field assessment showed right inferior field defect and general depression in the left. Optical coherence tomography retinal nerve fibre layer showed normal thickness over the right eye and generalized thinning over the left eye. Neuroimaging and other laboratory investigations were unremarkable. With good glycemic control, the optic disc swelling over the right eye resolved. Visual field defect remained the same but retinal nerve fiber layer showed thinning in areas where the edema had resolved.

INTRODUCTION

Pseudo-Foster Kennedy syndrome (PFKS) is defined as optic disc pallor in one eye and optic disc oedema in the contralateral eye in the absence of intracranial mass.[1] It is commonly caused by bilateral sequential optic neuritis or ischemic optic neuropathy.[2] Other causes which have been reported in the literature include benign intracranial hypertension and optic nerve hypoplasia.[3] To date, there are only a handful of cases that have reported diabetic papillopathy presenting as pseudo-Foster Kennedy. We report here on a case of pseudo-Foster Kennedy secondary to diabetic papillopathy and to describe the findings on visual field assessment and optical coherence tomography of the retinal nerve fibre layers (OCT RNFL) of this patient, which has not been discussed in previous literature.

CASE REPORT

A 31-year old Malay gentleman, with underlying Type 2 Diabetes Mellitus (DM), presented to the ophthalmology outpatient clinic with both eyes blurring of vision. His symptoms started in the left eye and had developed similar complaints in the fellow eye 6 weeks later. Otherwise, there were no symptoms of raised intracranial pressure and there were no neurological deficits. His past ocular history was unremarkable. He had Type 2 DM for two years, and his sugar control was not optimum. Upon examination, best-corrected vision in both eyes were 6/12. There was relative afferent pupillary defect in the left eye. Examination of the anterior segment examination was unremarkable. Fundus examination of the right eye showed a markedly swollen optic disc and it was associated with superficial, radially oriented and dilated telangiectatic vessels. The left optic disc was pale and atrophied. (Figure 1) There were no diabetic retinopathy changes seen on both fundi.

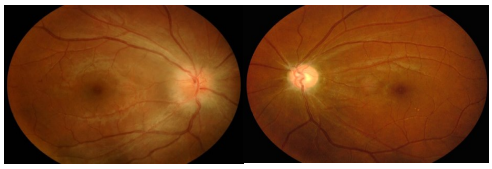

Figure 1: Right eye optic disc swelling and left eye optic disc pallor

The central nervous system examination showed no abnormalities. Colour vision was normal in both eyes. Visual field assessment over the right eye showed an inferior field defect, while the left eye showed a generalized depression. (Figure 2)

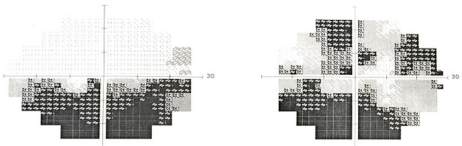

Figure 2: Right eye inferior field defect and left eye generalized depression

OCT RNFL of the right eye showed normal thickness but left eye showed generalized retinal nerve fibre layer thinning in all four quadrants. (Figure 3)

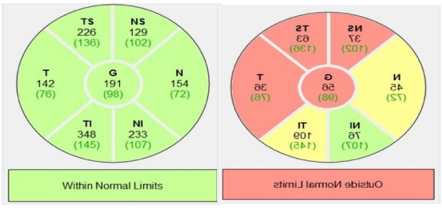

Figure 3: OCT RNFL of both eyes with left eye showing thinning in all quadrants

Computed tomography and magnetic resonance imaging of the brain and orbit showed no intracranial or intraorbital abnormalities. His random blood sugar was 12.3 mmol/L, HbA1C was 11.3%, fasting blood sugar 11mmol/L. Blood pressure during presentation was 139/97. Other blood investigations including full blood count, renal profile and ESR were within normal limits. Syphilis and TB workup were unrevealing. He was diagnosed with pseudo-Foster Kennedy syndrome (PFKS) secondary to diabetic papillopathy.

He was co-managed with the general physician in his glycemic control. He was started on a second oral hypoglycemic agent, with glucose levels targeted between 5 to 7mmol/L. During subsequent follow up at one month and three months, his vision remained the same. Disc oedema over his right eye had mostly resolved except the inferior temporal quadrant, which remained swollen at 3 months. The optic disc of the left eye remains pale. Visual field defects had remained the same. However, OCT RNFL showed thinning in areas where disc edema had resolved in the right eye. (Figure 4)

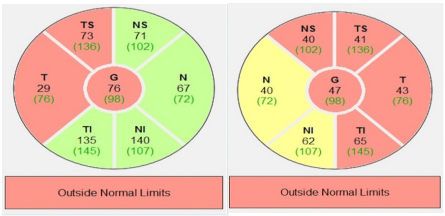

Figure 4: OCT RNFL after three months with right eye thinning in temporal quadrants

DISCUSSION

Diabetic papillopathy is characterized by unilateral or bilateral optic disc edema with array of visual loss seen in patients with diabetes. It is associated with both type 1 and type 2 DM and may occur regardless of metabolic control or severity of diabetic retinopathy.[4] Its exact incidence Is unknown as it is rarely reported in the literature. The pathophysiology of diabetic papillopathy is thought to be caused by a disruption of interstitial fluid dynamics and perfusion of capillary membranes resulting in edema, which will subsequently lead to ischemia, compression or toxic effects on the optic nerve head.[4] Diabetic papillopathy commonly occurs in younger patients and they present with normal pupil function, unilateral or bilateral disc swelling with associated dilated telangiectatic vessels and enlarged blind spot.[4] Bilateral involvement in a sequential manner resulting in a pseudo-Foster Kennedy clinical picture as seen in our patient is rare.

Diagnosis of diabetic papillopathy can be made when there is a confirmed diagnosis of diabetes with presence of optic disc edema without a substantial optic nerve dysfunction. There must not be raised intracranial pressure, and secondary causes such as infection or infiltration to the optic nerve.[5] Non-arteritic ischaemic optic neuropathy (NAION) is a differential diagnosis which may present in a similar manner, particularly a sequential NAION, whereby there may be optic disc swelling accompanied with optic atrophy in the fellow eye. However, NAION typically affects older patients, symptoms are acute in onset and there is usually a more marked visual impairment. Moreover, blood pressure in this patient was also within the normal range during presentation.

An array of ocular investigations can be done to aid diagnosis. Fundus fluorescein angiography of patients with diabetic papillopathy will show disc hyperfluorescence with late leakage and absence of choroidal hypoperfusion. However, there are no reports regarding the visual field or OCT RNFL findings in these patients. The visual field changes in our patient which showed an inferior field defect in the eye with optic disc swelling and generalized depression over the eye with optic atrophy was consistent

with findings in other reported cases of pseudo-Foster Kennedy syndrome PFKS which were secondary to other causes, such as NAION and vitamin B12 deficiency.[6,7] However, Trikey et al reported an enlarged blind spot with peripheral field constriction over the eye with optic disc swelling and generalized depression in the eye with optic atrophy.[8] OCT RNFL of our patient showed generalized thinning in the eye with optic atrophy and normal thickness in the eye with swollen optic disc. This was also consistent with the above mentioned patients with NAION and vitamin B12 deficiency. There was no progression of the visual field defect in our patient over both eyes. However, OCT RNFL showed thinning at areas where the optic disc swelling had resolved, most likely indicating optic disc atrophy occurred in those areas.

Diabetic papillopathy generally has a good prognosis and usually does not require any treatment. Strict glycemic control has shown variable outcomes, with some case reports showing resolution of the disc edema while some reporting worsening of the diabetic papillopathy.[4,9] Periocular steroids and intravitreal Bevacizumab have been reported to show promising outcomes.[10]

CONCLUSION

Though rare, diabetic papillopathy may present as PKFS. As both pseudo-Foster Kennedy and diabetic papillopathy are diagnosis of exclusion, neuroimaging and laboratory investigations are imperative to rule out other life-threatening causes of true Foster-Kennedy syndrome and optic disc swelling. Ocular investigations including visual field assessment and OCT RNFL are helpful in monitoring the structural and functional changes of the optic nerve in these conditions.

CONFLICT OF INTEREST

There is no conflict of interest

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REFERENCES

1. Bansal S, Dabbs T, Long V. Pseudo-Foster Kennedy Syndrome due to unilateral optic nerve hypoplasia: a case report. J Med Case Rep. 2008;2:86.
2. Micieli JA, Al-Obthani M, Sundaram AN. Pseudo-Foster Kennedy syndrome due to idiopathic intracranial hypertension. Can J Ophthalmol. 2014;49 (4):e99−e102.
3. Bhatnagar KR, Rauji C, Kumar R, et al. Pseudo Foster Kennedy Syndrome secondary to uncontrolled hypertension and diabetes mellitus: A case report. Med J DY Patil Univ. 2014:7-385-7. DOI: 10.4103/0975-2870.128999
4. Mallika PS, Aziz S, Asok T, et al. Severe diabetic papillopathy mimicking non-arteritic anterior ischemic optic neuropathy (NAION) in a young patient. Med J Malaysia. 2012;67(2):228−30.
5. Arnold AC. Ischemic opthic neuropathy. In: Miller NR, Newman NJ, eds. Walsh and Hoyt's Clinical Neuro-Ophthalmology, 6th ed. Philadelphia: Lippincott Williams &Wilkins; 2005: 377-8.
6. David C, Suvac, E, Tăbăcaru B, et al. Pseudo-Foster Kennedy Syndrome - a case report. Romanian journal of ophthalmology. 2016: 60(4), 270–4.
7. Tirkey ER, Chandravanshi SL, Abdulrahman CY, et al. Pseudo-Foster Kennedy Syndrome due to idoppathic intracranial hypertension associated with empty sella syndrome and hyperprolactinoma: A Rare Case Report. DJO 2018;2018:48-50
8. Vignesh AP, Srinivasan R. Pseudo-foster kennedy syndrome due to diabetic papillopathy. Adv Ophthalmol Vis Syst. 2015;2(4):112-3. DOI: 10.15406/aovs.2015.02.00050
9. Mansour AM, El-Dairi MA, Shehab MA, et al. eriocular corticosteroids in diabetic papillopathy. Eye (Lond). 2005 Jan; 19(1): 45-51.
10. Al-Dhibi H, Khan AO. Response of diabetic papillopathy to intravitreal bevacizumab. Middle East Afr J Ophthalmol. 2011 Jul; 18(3): 243-5.