

Case Report: Pseudo-Foster Kennedy Syndrome

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Abstract

Pseudo-Foster Kennedy Syndrome is characterized by unilateral optic disc swelling and contralateral optic atrophy without any compressive lesion of optic nerve. The cause is bilateral sequential anterior ischemic optic neuropathy.

Introduction

Foster Kennedy Syndrome is a rare disorder. It characterized by unilateral optic disc swelling with contralateral optic atrophy due to an intracranial lesion commonly a tumor at frontal lobe, sphenoid wing or olfactory groove region such as meningioma. This cause optic atrophy due to optic nerve compression in affected eye and papilledema due to increased intracranial pressure in contralateral eye¹. In some cases may have symptoms such as nausea, vomiting or loss of smell². In the absence of any compressive lesion these may be called Pseudo-Foster Kennedy Syndrome³. This is usually due to bilateral anterior ischemic optic neuropathy consecutively, which has optic atrophy in the previous affected eye and optic disc swelling in the current attack eye.

Case report

A 46-year-old woman presented with a painless sudden visual loss after awakening in the left eye for

1 month. She had a history of sudden visual loss in the right eye for 6 months. She had a history of diabetic mellitus and hypertension for 5 years, but she lost follow up and did not taking medications about several months. She had the symptoms of chronic intermittent headache, nausea and vomiting 1-2 times a month for 1 year.

Ophthalmological examination revealed best corrected visual acuity was 20/200 in the right eye and 20/70 in the left eye. The intraocular pressure was 15 and 16 mmHg. Slit lamp examination revealed the cornea was clear, no anterior chamber cells/flare. There was grade 1 nuclear sclerosis cataract in her both eyes. The pupil was 3 mm react to light both eyes with afferent pupillary defect (RAPD) positive in the right eye. The fundus examination reveal optic atrophy in the right eye and diffuse swollen disc and inferior peripapillary flame shaped hemorrhage in the left eye with some hard exudates at macular region in both eyes (Figure 1). The retinal vessels had mild retinal arterial narrowing and no dilatation or tortuosity of retinal veins. Physical examination revealed blood pressure was 140/80 mmHg. Other neurological examinations are normal. The perimetry showed a central scotoma in the right eye and an enlarged blind spot in the left eye.

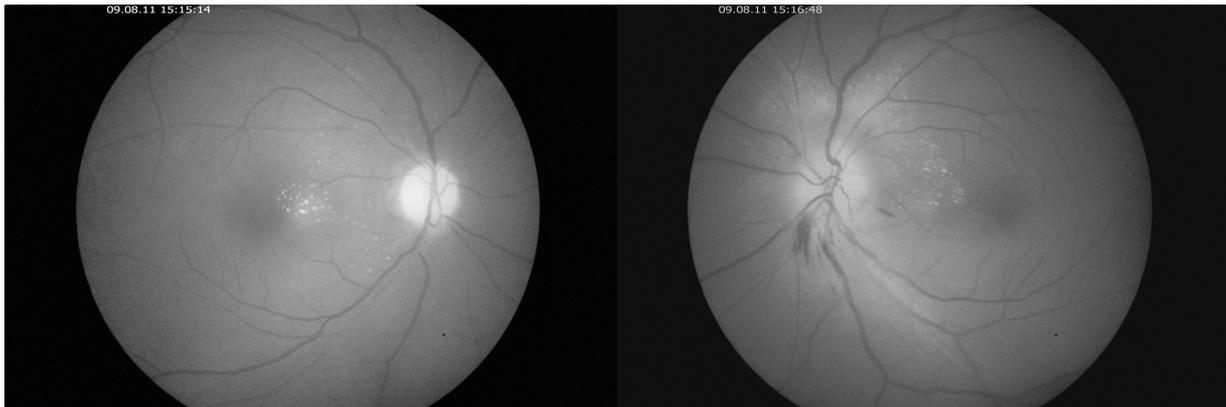


Figure 1 Fundus photographs show optic atrophy in the right eye and swollen disc in the left eye.

Computerized tomography (CT) of brain demonstrated no intracranial lesion. The blood test revealed high fasting blood sugar level and hyperlipidemia. This case is diagnosed Pseudo-Foster Kennedy syndrome due to bilateral nonarteritic anterior ischemic optic neuropathy (NAION). Risk factors for NAION in this case include systemic hypertension, diabetes and hyperlipidemia. Although there is no proven therapy for NAION. The short course of corticosteroid therapy may be used to improved optic disc swelling and visual function in the current attack eye. This case had start oral prednisolone dose 1 mg/kg/day which taper slowly in 4 weeks and tight control hypertension, diabetes and hyperlipidemia by the internist. Follow up periods at 1, 2 and 4 weeks, the patient reported of improved vision in her left eye. The visual acuity was 20/200 in the right eye and recovered to 20/40 in the left eye. The fundus examination revealed optic atrophy in the right eye and improved of sectoral swollen disc in the left eye. On the follow up period at 2 months after treatment, the fundus examination revealed optic atrophy in both eyes with marked pallor disc in the right eye (Figure 2). The final visual acuity still was 20/200 and 20/40. The systemic blood pressure and blood sugar also be good control.

Discussion

Diagnosis of Pseudo-Foster Kennedy Syndrome is based on the clinical findings of unilateral optic disc swelling and contralateral optic atrophy without any compressive lesion of optic nerve. This syndrome must be differentiated from True-Foster Kennedy Syndrome with similar optic disc pictures because it caused by an intracranial lesion such as meningioma at frontal lobe, sphenoid wing or olfactory groove regions. The neuroimaging (CT/MRI) should be performed in all suspected cases. In Pseudo-Foster Kennedy Syndrome is commonly caused by bilateral sequential anterior ischemic optic neuropathy with normal neuroimaging.

Anterior ischemic optic neuropathy (AION) is the common acute optic neuropathy in older patients⁴. The patients will present with painless monocular visual loss over days. An RAPD usually present. In acute stage, the optic disc is hyperemic or pale, edema may be sectoral or diffuse edema. There can be seen peripapillary flame shaped retinal hemorrhage and retinal arteriole narrowing. AION can be classified as arteritic (AAION) or nonarteritic (NAION).

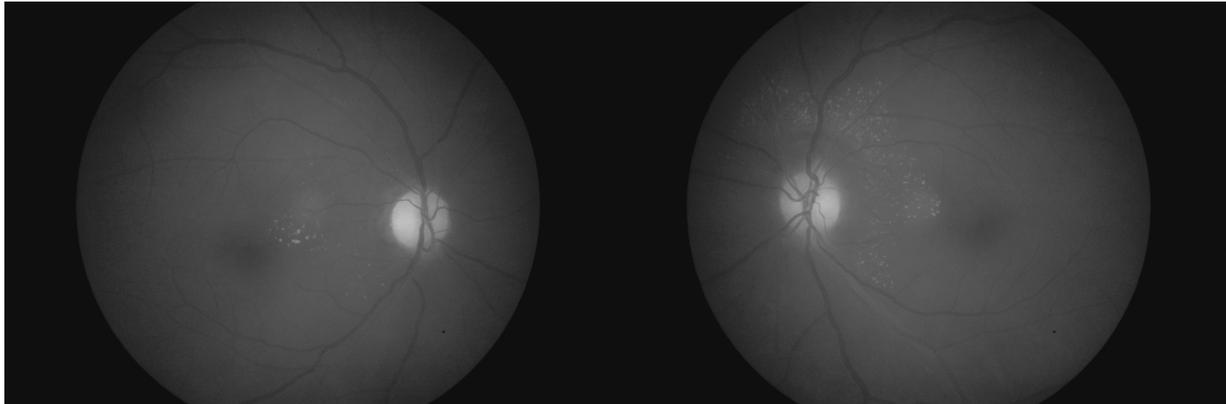


Figure 2 Fundus photographs show optic atrophy in both eyes with marked pallor disc in the right eye

NAION is more common than AAION and occur in patients over 50 years of age. The etiology may be related to compromise of the optic disc circulation affected by microvascular occlusive disease such as hypertension and diabetes⁵. The visual acuity usually over 20/200 in majority of cases. The optic disc may be sectoral or diffuse edema. The optic disc in contralateral eye is typically small or absent physiologic cupping “Disc at risk”. After resolved edema, the optic disc becomes atrophic in 4-8 weeks. The 5 year risk of fellow eye involvement is 12-19%⁶. If the fellow eye involvement occurred, the previous optic disc is atrophy and the current attack eye is swollen disc, which can give clinical pictures of Pseudo-Foster Kennedy Syndrome. The goal of therapy is decreased systemic vascular complications and contralateral visual loss. In AAION, the corticosteroid is the main therapy with early intravenous methylprednisolone 1 g/day for 3-5 days then oral prednisolone 1 mg/kg/day tapered slowly within 3-12 months, but in contrast to NAION which there is no proven therapy. Recent options for NAION include neuroprotective agents, hyperbaric oxygen, optic nerve sheath decompression surgery. Treatment of NAION with short course

corticosteroid is controversial. Visual function may be recovered faster, but the long term outcome may not differ and considering of side effect of corticosteroid must be weighted for patients. There is no role of Aspirin in both affected eyes but there may be a role to reducing the stroke in the high risk case⁷. Furthermore, the tight control of systemic vascular disease remain important in all cases.

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รายงานผู้ป่วย: โรคเส้นประสาทตาขาดเลือดหล่อเลี้ยง ชนิด Pseudo-Foster Kennedy syndrome

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บทคัดย่อ

ผลการศึกษา Foster Kennedy syndrome เป็นภาวะที่มีขั้วประสาทตาบวมข้างหนึ่งร่วมกับตรวจพบขั้วประสาทตาฝ่อในตาอีกข้างซึ่งสาเหตุเกิดจากเนื้องอกสมองบริเวณ frontal lobe, sphenoid wing หรือบริเวณ olfactory groove ซึ่งทำให้ขั้วประสาทตาข้างหนึ่งบวมจากความดันในสมองเพิ่มสูงขึ้นและขั้วประสาทตาอีกข้างฝ่อซีดจากการกดเบียดของเนื้องอก นอกจากนั้นในภาวะขั้วประสาทตาบวมจากการขาดเลือดหล่อเลี้ยง (anterior ischemic optic neuropathy) ที่เป็นสองข้างตามกัน จะตรวจพบขั้วประสาทตาบวมในตาข้างหนึ่งและขั้วประสาทตาฝ่อซีดในตาอีกข้างที่เคยมีขั้วประสาทตาบวมจนภายหลังได้ยุบบวมลงแล้ว เรียกภาวะนี้ว่า Pseudo-Foster Kennedy syndrome ภาวะนี้มีมักพบในผู้ป่วยที่อายุมากกว่า 50 ปี และสัมพันธ์กับโรคประจำ

ตัวเช่น ความดันโลหิตสูง เบาหวาน เป็นต้น รายงานผู้ป่วย 1 รายที่มาด้วยอาการตามัวข้างหนึ่ง และเคยมีการสูญเสียการมองเห็นในตาอีกข้างในลักษณะเดียวกัน จากการตรวจพบมีการบวมขั้วประสาทตาข้างหนึ่งและขั้วประสาทตาฝ่อในตาอีกข้าง จากการตรวจเอกซเรย์คอมพิวเตอร์สมองไม่พบเนื้องอกหรือความผิดปกติอื่น ผู้ป่วยมีโรคประจำตัว ความดันโลหิตสูงและเบาหวานที่ขาดการรักษาต่อเนื่อง ได้รับการวินิจฉัยว่าเป็น Pseudo-Foster Kennedy syndrome จากภาวะ anterior ischemic optic neuropathy ภายหลังการรักษาด้วยการควบคุมความดันโลหิตและเบาหวาน ร่วมกับการให้ยาสเตียรอยด์ในระยะสั้น พบว่าระดับสายตาในตาข้างที่มีอาการภายหลังสามารถกลับฟื้นคืนขึ้นได้

คำสำคัญ: เส้นประสาทตาขาดเลือดหล่อเลี้ยงชนิด

Pseudo-Foster Kennedy syndrome