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CASE REPORT

Pseudo-Foster Kennedy Syndrome – a case report

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Abstract

Objective: To report a case of Pseudo-Foster Kennedy (PFK) syndrome and describe its clinical and paraclinical particularities, as well as the diagnostic difficulties and established treatment.
Methods: The case of a 60-year-old male patient with sudden, painless visual impairment in the left eye (LE), and a medical history of old optic nerve atrophy in his right eye (RE) was described.
Results: The diagnosis of nonarteritic anterior ischemic optic neuropathy (NAION) was established based on the medical history, local and general clinical and paraclinical examination, and temporal artery biopsy.
Conclusions: Although there is no current generally accepted treatment for NAION, a correct diagnosis and supportive treatment may contribute to the improvement in visual acuity (VA), improvement that in this case remained stable for 6 months after the onset. The patient is still being monitored and no relapses have been noted.
Keywords: Pseudo-Foster Kennedy syndrome, anterior ischemic optic neuropathy, giant cell arteritis, nonarteritic anterior ischemic optic neuropathy

Introduction

Foster Kennedy syndrome represents the presence of a unilateral optic disc swelling with contralateral optic nerve atrophy, due to an intracranial mass. A true Foster Kennedy syndrome is rare, but the presence of these findings, in the absence of an intracranial mass is referred to as pseudo-Foster Kennedy syndrome. The most common causes of PFK syndrome are nonarteritic anterior ischemic optic neuropathy (NAION) and arteritic anterior ischemic optic neuropathy (AAION).

Materials and methods – Case report

A 60-year-old Caucasian man first presented to our clinic in February 2016 with a painless visual impairment in the LE, of sudden onset 3 days earlier, for which he did not undergo any medical treatment. His medical history revealed an old optic nerve atrophy (anterior ischemic optic neuropathy (AION)) in his RE, diagnosed in 2009, and dyslipidemia. There was no relevant family history of ocular or systemic diseases and the only prior medication the patient was receiving was lipid-lowering medication (statins).

On presentation, his best-corrected visual acuity (VA) was 1/ 20 in his RE (which the

patient was aware of), and 2/ 20 on his LE (where it was previously documented at a value of 20/ 20, 2 months earlier). The intraocular pressure measured by applanation tonometry was 16 mmHg in the RE and 15 mmHg in the LE. The visual field testing showed a severe altitudinal visual field defect in the LE (**Fig. 1**). These functional changes were not present 2 months prior to the presentation to our clinic. No significant changes were observed on the visual field examination for the RE.

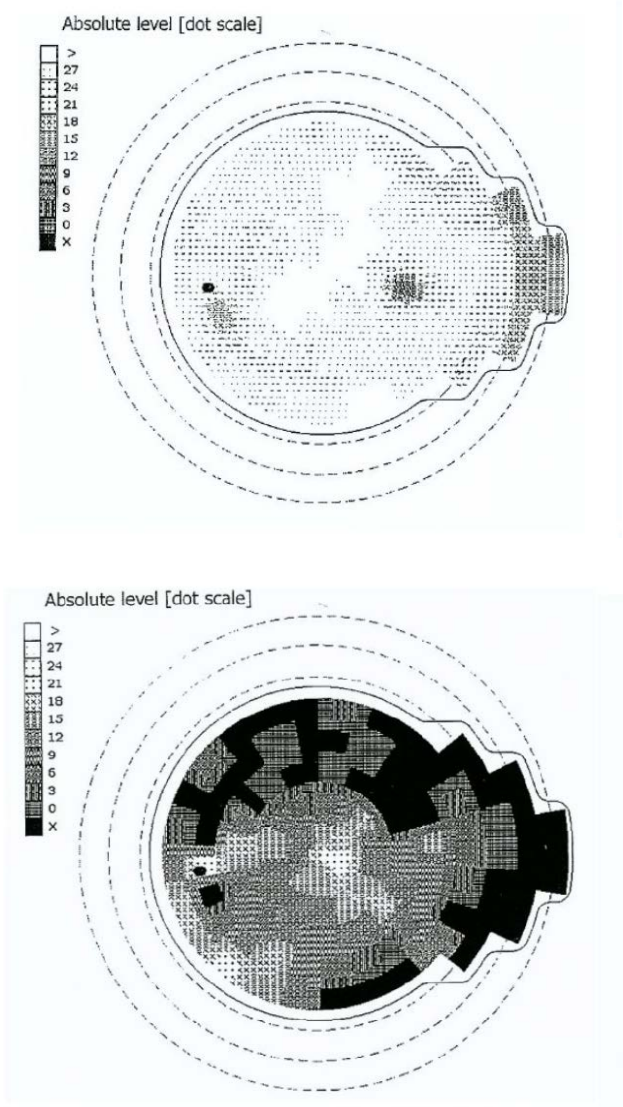


Fig. 1 Perimetric examination of the LE 2 months prior to presentation (left image) and at presentation (right image), showing a new, severe altitudinal field defect

The slit-lamp examination revealed optic nerve atrophy in the RE and hyperemic optic nerve swelling with a "splinter" hemorrhage in the temporal superior sector of the optic nerve head in the LE (**Fig. 2**).

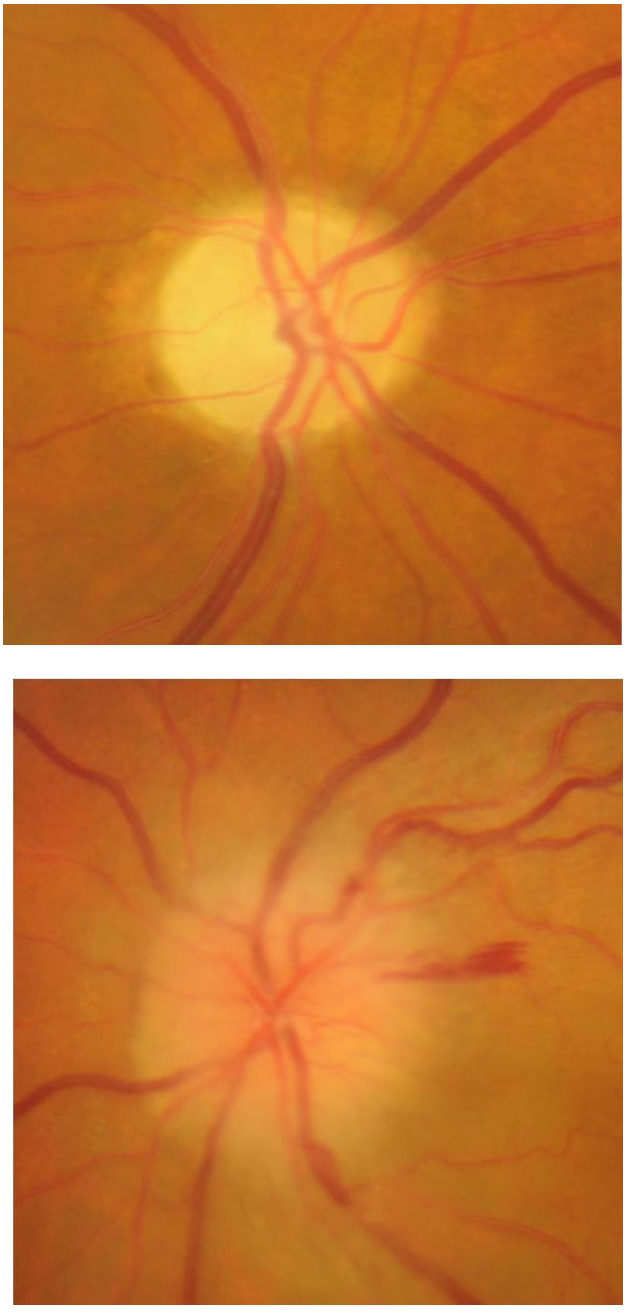


Fig. 2 Fundus color photography showing atrophy of the right optic disc (left image) and hyperemic swelling of the left optic disc, with a "splinter" hemorrhage (right image)

A diffuse RE atrophy in the superior and inferior quadrants was identified on the optical coherence tomography (OCT) examination (**Fig. 3**). Regarding the LE, the elevated aspect of the optic disc led to an artifactual interpretation of the average RNFL thickness, which was falsely registered as thickened.

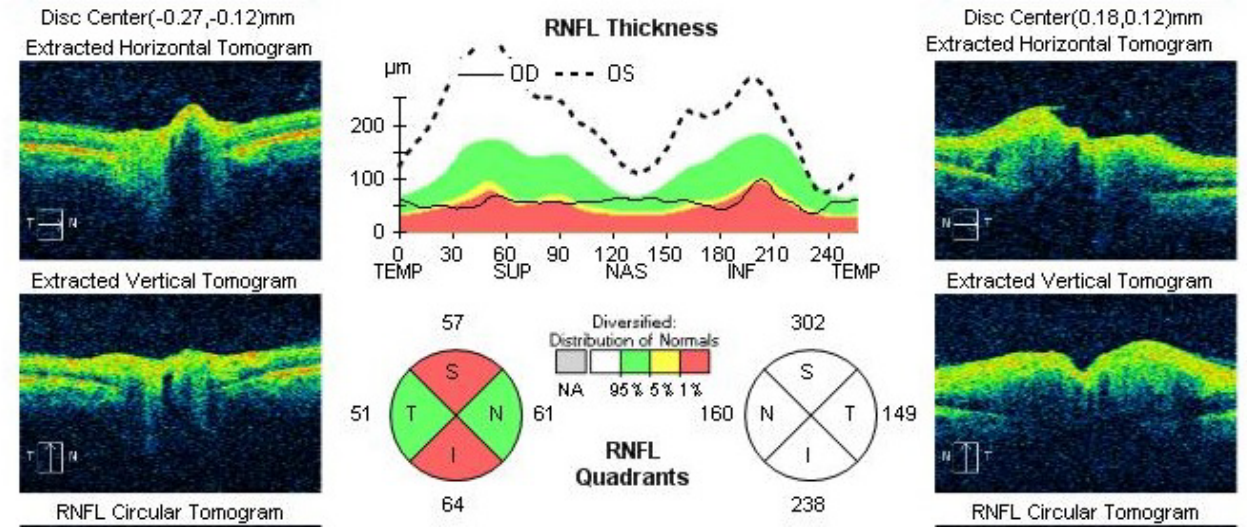


Fig. 3 OCT examination of RE and LE on presentation

We further referred the patient for other investigations in order to establish a diagnosis. His blood cell count showed elevated white blood cells – 14 100/ µl (normal = 4 000 – 10 000/ µl), his lipid panel showed high blood cholesterol – 295 mg/ dl (normal = 140 - 220 mg/ dl), elevated total lipids - 835 mg/ dl (normal = 500 - 800 mg/ dl), and his inflammatory markers revealed a slightly higher erythrocyte sedimentation rate (ESR) – 23 mm. at 1 hour (normal < 20 mm.) and a higher C-reactive protein (CRP) level – 168% (normal = 70 - 130%). The antiphospholipid antibodies and thrombophilia tests found no abnormalities. A neurological examination found the patient clinically normal, and a magnetic resonance imaging (MRI) head scan revealed no expansive intracranial or intraorbital processes, and no demyelinating lesions. A cardiovascular exam diagnosed arterial hypertension (so antihypertensive medication was prescribed) and excluded abnormal heart rhythms. The echocardiogram revealed no regurgitations, no vegetations, and no hemodynamically significant stenosis. A cervical Doppler ultrasound examination found bilateral carotid atheromatosis, but no hemodynamically significant stenosis.

The medical history, local and general clinical examination, and investigative test results corroborated towards the presumptive diagnosis of anterior ischemic optic neuropathy (AION) of yet unestablished etiology (arteritic or nonarteritic).

Furthermore, a temporal artery biopsy was recommended. In the expectation of the histopathological result, a treatment was established with: systemic corticosteroids (methylprednisolone) – intravenous, 1 g/ day for 3 days, followed by an oral dose of 1 mg/kg/day, H2-receptor antagonists (famotidine), cerebral vasodilating agents (nicergoline), antiplatelet drugs (acetylsalicylic acid), peripheral arterial vasodilators (pentoxifylline) and retinal neuroprotective agents.

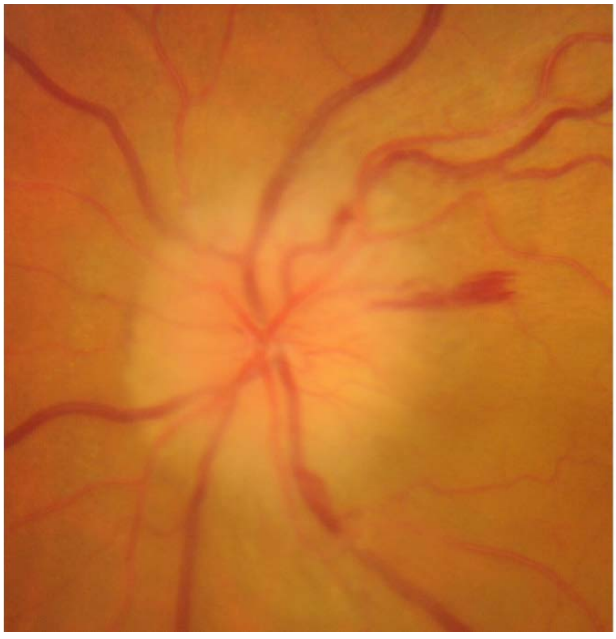
One week after the presumptive diagnosis was established and the treatment started, the histopathological examination result was

received, showing no pathological alterations of the arterial wall structure, no giant cells, no epithelioid cells, and no inflammatory infiltrates.

Thus, because of the negative histopathological examination result for AAION (giant-cell arteritis aka Horton's disease), normal CRP and ESR levels one week after presentation, and lack of a suggestive symptomatology (headaches, jaw pain, scalp tenderness, fever, fatigue), a definitive diagnosis of NAION was established.

Therefore, the treatment with systemic corticosteroids was progressively reduced and then completely stopped. The patient continued taking cerebral vasodilating agents, antiplatelet drugs and retinal neuroprotective agents and was instructed to return for regular check-ups every month.

One month after presentation, no change in VA-RE (which remained stable at a 1/ 20) was found, but a significant improvement in VA-LE (which rose from 2/ 20 to 10/ 20) was found. The intraocular pressure was normal, with a value of 11 mmHg in both eyes. The fundoscopic examination (**Fig. 4**) showed the reduction of the optic disc swelling, now with clear optic nerve margins contour and a resolution of the "splinter" hemorrhage and hyperemic aspect, in the LE. The RE showed no significant modifications. 6 months after the initial presentation, the patient's VA-LE remained stable at 10/ 20.



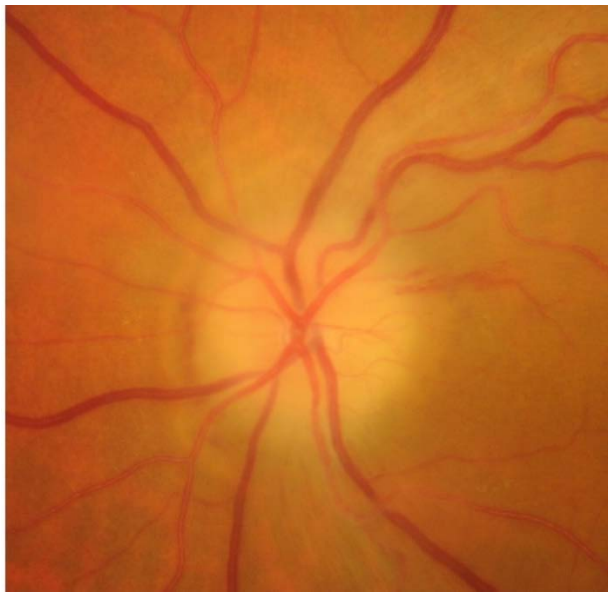


Fig. 4 LE Fundus aspect on presentation (left image) and 1 month later (right image), showing the reduction of optic disc swelling, now with clear optic nerve margins contour and resolution of the "splinter" hemorrhage and hyperemic aspect

Discussion

Because the PFK syndrome is a diagnosis of exclusion, neuroimaging plays an important role in distinguishing between the "true" Foster-Kennedy syndrome and the PFK syndrome. Thus, because of the lack of expansive intracranial or intraorbital processes on the head MRI scan, we had a case of PFK syndrome and papilloedema was deemed unlikely. The most common cause of PFK syndrome is AION, and the differential diagnosis included optic neuritis (unlikely because of the absence of a central scotoma on the visual field testing, atypical age of onset, and lack of any demyelinating lesions on the head MRI) and central retinal vein occlusion (unlikely due to fundoscopic findings, medical history, and RE optic disc aspect).

After establishing the diagnosis of AION, it became necessary to determine whether the patient had the nonarteritic or arteritic form of AION. Even though the most common form of AION (accounting for 90-95% cases) is attributed to NAION [**1**], the mean age of onset for NAION being 60 years (our patient also being 60 years old), and absent suggestive

symptomatology for AAION (headaches, jaw pain, scalp tenderness, fever, fatigue), the presence of elevated ESR and CRP levels on presentation determined us to recommend a superficial temporal artery biopsy, the gold standard for the diagnosis of AAION [**2**].

The immediate initiation of high-dose glucocorticosteroids was recommended and should not have been delayed while awaiting the histopathological confirmation if suspicion for AAION was raised [**3**].

Even though a negative biopsy result does not definitely rule out AAION (due to the presence of skip lesions or suboptimal biopsy in some cases) [**3,4**], the lack of a suggestive symptomatology and the normalization of ESR and CRP values deemed the diagnosis of AAION unlikely. This, together with the presence of systemic hypertension, hyperlipidemia, bilateral carotid atheromatosis, and optic nerve atrophy of the RE, corroborated towards the definitive diagnosis of NAION.

There is no generally accepted treatment for NAION. The Ischemic Optic Neuropathy Decompression Trial failed to show any benefit of surgery in NAION [**5**], but oral corticosteroids have shown some improvement in visual acuity and visual field in treated versus untreated cases [**6**]. Under supportive treatment with cerebral vasodilating agents, antiplatelet drugs and retinal neuroprotective agents, the patient's VA-LE increased (to 10/ 20) and then remained stable. What should be also noted is that without a proper referral to a cardiologist, the patient might have still not been diagnosed with systemic arterial hypertension for which he is now undergoing cardiac treatment.

Financial Disclosures

None of the other authors has any financial or proprietary interests to disclose.

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