

Bilateral simultaneous non-arteritic anterior ischemic optic neuropathy with occlusion of unilateral cillioretinal arter: Case report

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ABSTRACT

A 45-year-old female patient was consulted for bilateral visual loss. She was receiving hemodialysis since 15 years. At presentation ophthalmologic examination her visual acuity was hand movements in the right eye and light perception negative in the left. Direct light response was weak in the right eye and absent in left eye. A total afferent pupillary defect was detected in the left eye. Fundoscopy revealed bilateral sectorial pale, sectorial hyperaemi and swollen optic discs. On the left papilla there was splinter haemorrhages. At the right eye there was a pale edema at superior maculopapillary bunch that coherent with occlusion of cillioretinal arter. She was diagnosed as non-arteritic ischemic optic neuropathy based on the clinical and fundoscopic examination. Methylprednisolone intravenous 1000 mg/day for 3 days then oral methylprednisolone 1 mg/kg/day was given for 1 week. Simultaneous acetylsalicylic acid 325 mg/day and hydration with 0.9% 1000cc isotonic solution treatment started. After 2 months visual acuities were not change in both eyes and both optic disks were pale.

Keywords: Cillioretinal arter; hemodialysis; ischemic optic non-arteritic neuropathy.

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Optic nerve ischemia may be in different anatomical locations. Anterior ischemic optic neuropathy (AION) involves the 1mm segment of the optic nerve head. AION can be non-arteritic (NAION) or arteritic (AAION). Most of the cases of AION are NAION (95%) and usually unilateral involvement occurs [1]. NAION is the most common optic neuropathy over 50 years old and average age is 60 [2]. NAION can have a numerous of etiologies including atherosclerosis, diabetes mellitus, hyperlipidemia, hypertension, sudden hypotensive events, cataract surgery, sleep apnea syndrome, hemoconcentration, hemodilution, hypercoagulable states, migraine, collagen vascular disease [3, 4]. AAION is almost always associated with giant cell arteritis (GCA) and average age is 70 [5]. Bilateral involvement is more

often at AAION than NAION. Bilateral simultaneous NAION is extremely rare [6]. Massive haemorrhage, hypotension, chronic hemodialysis, chronic anaemia and some drugs (interferon alfa and sildenafil) can cause optic nerve ischemia. Bilateral simultaneous NAION may occur in young individuals at these situations [7–9]. In this case report, we presented a 45 years old female with bilateral simultaneous NAION with occlusion of righth cillioretinal arter.

CASE REPORT

A 45 years of female patient with chronic renal failure who was receiving hemodialysis three times a week for 15 years was applied to emergency service because of



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bleeding from femoral artery catheter. Two units blood transfused to patient due to decrease in haemoglobin. The patient was operated for bleeding control and hospitalized at intensive care unit. Patient described bilateral vision loss one day after the hospitalization and consulted to ophthalmology clinic.

At presentation, patient described sudden, painless, bilateral vision loss. Visual acuity was hand movements in the right eye and light perception was negative in the left eye. Direct light response was weak in the right eye and absent in left eye. A total afferent pupillary defect was detected in the left eye. Anterior segment examination was unremarkable in both eyes. Intraocular pressure was 19 mm Hg in the right eye and 18 mm Hg in the left eye. Fundoscopy revealed bilateral sectorial pale, sectorial hyperaemia and swollen optic discs. On the left papilla there was splinter haemorrhages. At the right eye there was a pale edema at superior maculopapillary bunch that coherent with occlusion of cillioretinal arter. There were dirty yellow spot accumulations at the parafoveal area and pigmentary changes at the peripheral retina in both eyes. Also retinal veins enlarged compatible with hypertensive retinopathy (Figure 1).

At sistemic examination blood pressure was 100/54 mmHg, haemoglobin was 6.9 gr/dL, urea was 100 mg/dl, serum glucose level was 85 mg/dl, HbA1C was 4.7, low-density lipoprotein (LDL) cholesterol and triglyceride levels were 77 and 174 mg/dl respectively, vitamin B12 was 1000 pg/ml, serum erythrocyte sedimentation rate was 22/mm/h and C-reactive protein was 1 mg/dl (both were in normal limits). Vasculitic markers Antinuclear antibodies (ANA), anti-DNA, cytoplasmic and perinuclear anti-neutrophil cytoplasmic antibodies (c-ANCA and p-ANCA), anti kardiolipin and anti-phospholipid antibodies were all negative. Patient did not have headache, jaw claudication, scalp tenderness and polymyalgia rheumatica that were the most common systemic symptoms of AAION. Both temporal arteries were evaluated as normal on palpation. Colour Doppler ultrasound examination of both temporal arteries showed no significant obstruction and intima thicknesses were measured 1.5 mm in the right eye and 1.4 mm in the left. Rheumatology did not consider temporal arteritis and other vasculitis.

Neurological examination was normal. Remarkable cerebral or orbital pathology explaining bilateral visual loss was not detected in venous MR angiography, contrast-enhanced cranial MR and orbital MR. At cardiac



FIGURE 1. First examination pictures.

examination she had cardiac hypertension for 15 years. She was using beloc 50 mg 2x1 and plavix 75 mg 1x1. Transthoracic echocardiography showed grade one diastolic dysfunction, mild aortic, mitral and tricuspid valve insufficiency. Doppler ultrasonography revealed that internal carotid artery (ICA) peak systolic velocity (PSV) was 78 cm/sec at right and 76 cm/sec at left (within normal ranges). Angiotensin-converting enzyme, chest CT, rapid plasma reagin (RPR), venereal disease research laboratory (VDRL), pathergy test, purified protein derivative (PPD) were within normal range.

She was diagnosed as non-arteritic ischemic optic neuropathy based on the clinical, fundoscopic and systemic examination. Intravenous methylprednisolone 1000 mg/day was started for 3 days then oral methylprednisolone 1 mg/kg/day was given for 1 week. In addition to that acetylsalicylic acid 325 mg/day and 0.9% 1000 cc/day isotonic solution (to control the hypotension) treatment was started. Visual acuities did not change after 1 week. Fundus fluorescein angiography was revealed delay in filling optic disc and normal filling in peripapillar vascularisation at two weeks follow up (Figure 2). Optic disc and maculopapillary bunch edema was started to resolve substantially, both optic discs were mildly pale (Figure 3). After two months visual acuities did not change, direct light response was weak in the right eye and absent in left eye. Maculopapillary bunch edema resolved and both optic discs were pale (Figure 4).

DISCUSSION

The cause of NAION is the infarction of the optic nerve head that supplied by the posterior ciliary arteries. Presentation of NAION is sudden, painless, unilateral visual loss. Vision loss frequently develops in the morning therefore nocturnal hypotension may play an important role in the occurrence of NAION [10]. Optic disc is

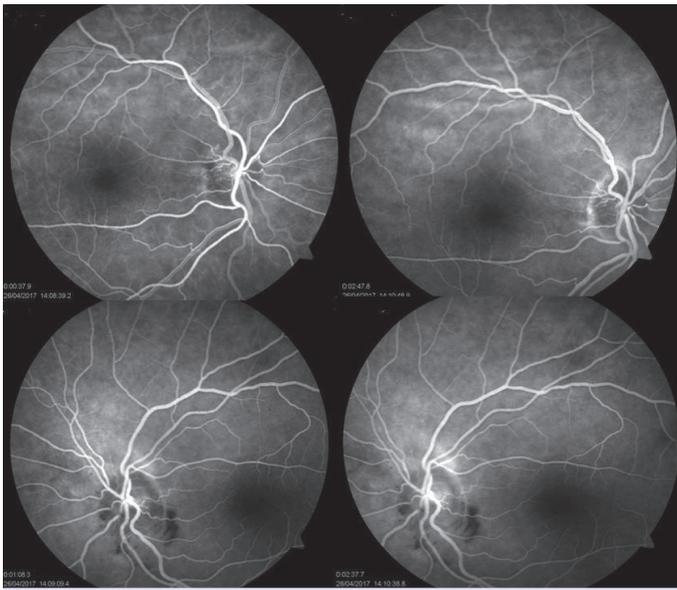


FIGURE 2. Fundus fluorescein angiography at two weeks follow up.



FIGURE 3. At two week follow up.



FIGURE 4. After two months.

small and crowded. There can be diffuse or sectoral, hyperaemic or pale disc swelling with peripapillary splinter haemorrhages [11].

In our case the presence of bilateral vision loss, bilateral disc edema and right eye cilioretinal artery occlusion

suggests an AAION at first glance. However age of the patient, absence of the most common systemic symptoms (headache, jaw claudication, scalp tenderness and polymyalgia rheumatica) and bilateral visual loss when woked up in the morning support the NAION [12]. The factors that were not support the AAION were there was not loss of pulsation of both temporal arteries, ESR and CRP were normal. At NAION and AAION after 4–8 weeks optic atrophy and generalized retinal artery thinning develops. But at AAION optic disc excavation may develop [13]. At our patient optic disc excavation did not develop. At both at FFA disk filling was delayed but at AAION also peripapillary choroidal filling was delayed [14]. Our patient's peripapillary choroidal filling was normal. But we could not do FFA at the first examination because the patient's systemic condition was not appropriate. In our patient there were dirty yellow spot accumulations at the parafoveal area and pigmentary changes at the peripheral retina in both eyes. In the literature there were retina pigment epithelium changes at chronic hemodialysis patients [15, 16].

In patients <50 years of age It is very important to exclude idiopathic optic neuritis, syphilis, sarcoidosis, infiltrative optic neuropathies, anterior orbital lesions that produce optic nerve compression and other autoimmune diseases [17]. As a result of our investigations, we could not find any of these diagnoses.

Our patient was receiving hemodialysis since 15 years and bleeding from femoral artery after right femoral catheter insertion for hemodialysis just before the consultation to us might have disturbed the already inadequate blood supply of the optic nerve and probably due to chronic hemodialysis the patient had a chronic anemia that reducing the optic nerve blood supply. Chronic hemodialysis patients often have risk factors for NAION such as anemia, hypotension, atherosclerosis and uremia [18]. Hypotension is the main cause of decreased perfusion of the microcirculation of the optic disc. Many causes can lead to chronic hypotension among patients on hemodialysis including hypovolemia, the use of anti-hypertensive medications and removal of vasopressors during dialysis [19]. Atherosclerosis causes increased resistance to blood supply. Anemia causes low blood oxygen carrying capacity. Uremic patients often may also have other coexisting pathological factors such as hypotension or hypertension, atherosclerosis, and anemia. These compounded factors decrease the posterior ciliary artery blood supply that predispose them to NAION [20].

Despite the fact that the ciliary artery occlusion was in the right eye, it was observed that the visual defect in the left eye is heavier. The reason of this may be deep ischemia of the left optic nerve.

The first report of anterior ischemic optic neuropathy as a result of hemodialysis-associated hypotension was published in 1986 [21]. Afterwards different publications on this subject were made [22, 23]. Different visual acuities have been reported in patients with NAION on hemodialysis at first examination and after treatment [24–26]. According to other reports, our patient's first and after treatment visual acuities were low. Also there were reports of bilateral simultaneous NAION in the literature [27, 28]. However this is the first report bilateral simultaneous NAION with occlusion of unilateral cilioretinal artery after chronic hemodialysis.

Based on all the findings described above patient was diagnosed as NAION. NAION has no proven effective treatment. Optic nerve sheath decompression surgery has been tried to treat NAION but it was not beneficial and potentially harmful [29]. Hyperbaric oxygen therapy did not produce a significant improvement in visual acuity [30]. Acetylsalicylic acid is effective in reducing systemic vascular events but it does not appear to reduce the risk of involvement of the fellow eye [31]. At Salomon O. and et al study they suggest that acetylsalicylic acid at 325 mg/day may be effective in reducing the frequency of second eye involvement with NAION [32]. Therefore we used the dose of 325 mg/day acetylsalicylic acid. Correction of hypotension ensures the adequate perfusion of the optic nerve. The use of steroids is controversial, but positive effects have been reported in some studies [33]. We started methylprednisolone, acetylsalicylic acid and hydration treatment to our patient but after the treatment the visual acuities were not increased.

Conclusion

Chronic hemodialysis may cause anemia, hypotension and uremia. This case supports chronic hemodialysis is a risk factor for developing NAION. NAION be accompanied with cilioretinal artery occlusion. Both nephrologist and ophthalmologist should be aware of this potentially complication.

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authors.

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