

# Bilateral simultaneous non-arteritic anterior ischemic optic neuropathy with occlusion of unilateral cilioretinal artery: A case report

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## ABSTRACT

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

*Keywords:* Cilioretinal 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 the average age is 60 [2]. NAION may have many etiologies, including atherosclerosis, diabetes mellitus, hyperlipidemia, hypertension, sudden hypotensive events, cataract surgery, sleep apnea syndrome, hemoconcentration, hemodilution, hypercoagulable states, migraine and collagen vascular disease [3,

4]. AAION is almost always associated with giant cell arteritis (GCA), and the 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) may cause optic nerve ischemia. Bilateral simultaneous NAION may occur in young individuals at the situations reported above [7–9]. In this case report, we presented a 45 years old female patient with bilateral simultaneous NAION with occlusion of the right cilioretinal artery.



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## CASE REPORT

A 45-year-old female patient with chronic renal failure who was receiving hemodialysis three times a week for 15 years was applied to our emergency service at the hospital because of bleeding from femoral artery catheter. Two units of blood transfused to the patient due to a decrease in haemoglobin. The patient was operated for bleeding control and hospitalized at an intensive care unit. The patient described bilateral vision loss one day after the hospitalization and consulted ophthalmology clinic.

At presentation, the 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. The direct light response was weak in the right eye and absent in the 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 were splinter haemorrhages. At the right eye, there was pale edema at superior maculo-papillary bunch that coherent with occlusion of the cilioretinal artery. 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 (Fig. 1).

At systemic examination blood pressure was 100/54 mmHg, haemoglobin was 6.9 gr/dL, urea was 100mg/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-cardiolipin and antiphospholipid antibodies were all negative. The patient did not have a headache, jaw claudication, scalp tenderness and polymyalgia rheumatica that were the most common systemic symptoms of AAIION. 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

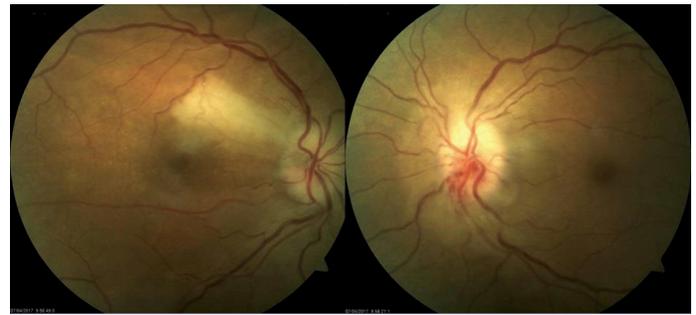


FIGURE 1. Pictures of the first examination.

mm in the left. Rheumatology did not consider temporal arteritis and other vasculitis.

Neurological examination was normal. Remarkable cerebral or orbital pathology that may account for the bilateral visual loss was not detected in venous MR angiography, contrast-enhanced cranial MR and orbital MR. At the cardiac examination, she had cardiac hypertension for 15 years. She was using Beloc 50 mg 2×1 and Plavix 75 mg 1×1. 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, and purified protein derivative (PPD) were within a normal range.

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



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



**FIGURE 3.** At two weeks follow-up.



**FIGURE 4.** After two months.

## DISCUSSION

The cause of NAION is the infarction of the optic nerve head that was supplied by the posterior ciliary arteries. Presentation of NAION is a sudden, painless, unilateral

visual loss. Vision loss frequently develops in the morning. Thus, nocturnal hypotension may play an important role in the occurrence of NAION [10]. The optic disc is small and crowded. There may 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 suggest an AAION at first glance. However, age of the patient, absence of the most common systemic symptoms (e.g. headache, jaw claudication, scalp tenderness and polymyalgia rheumatica) and bilateral visual loss when woke up in the morning support the NAION [12]. The factors that did 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 develop. However, at AAION optic disc excavation may develop [13]. At our patient optic disc excavation did not develop. At both at FFA disk, the filling was delayed, and at AAION, parapapillary choroidal filling delayed [14]. Our patient's parapapillary choroidal filling was normal. However, we could not carry out 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 retinal 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, might have disturbed the already inadequate blood supply of the optic nerve and probably because chronic hemodialysis the patient had chronic anemia that was 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 may lead to chronic hypotension among patients on hemodialysis, including hypovolemia,

the use of anti-hypertensive medications and removal of vasopressors during dialysis [19]. Atherosclerosis gives rise to increased resistance to the 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 predisposes them to NAION [20].

Although the ciliary artery occlusion was in the right eye, it was observed that the visual defect in the left eye was heavier. The reason for this may be the 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 studies were on this subject [22, 23]. Different visual acuities were reported in patients with NAION on hemodialysis at first examination and after treatment [24-26]. According to other reports, our patient's visual acuities were low at first and after the treatment. Also, there were reports of bilateral simultaneous NAION in the relevant literature [27, 28]. However, to our knowledge, this study is the first report bilateral simultaneous NAION with occlusion of the unilateral cilioretinal artery after chronic hemodialysis.

Based on all the findings described above, the patient was diagnosed as NAION. NAION has no proven effective treatment. Optic nerve sheath decompression surgery was 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]. In Salomon et al.'s study, they suggest that acetylsalicylic acid at 325 mg/day might be effective in reducing the frequency of second eye involvement with NAION [32]. Therefore, we used a 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 for our patient, but after the treatment, the visual acuities did not increase.

## Conclusion

Chronic hemodialysis may cause anemia, hypotension and uremia. This case supports chronic hemodialysis

is a risk factor for developing NAION. NAION can be accompanied by cilioretinal artery occlusion. Both nephrologist and ophthalmologist should be aware of this potential complication.

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