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Presumed ischemic optic neuropathy

Nonarteritic anterior ischemic optic neuropathy (NAION) is a relatively common cause of acute, painless vision loss in patients typically older than 50 years. Presentation is often associated with predisposing vasculopathic risk factors, but has also been reported as a complication of pregnancy. We describe a young, healthy female with disc drusen who suffered bilateral NAION due to postpartum blood loss. To our knowledge this is the first such case to be reported in the English-language ophthalmic literature. Clinicians should be aware that patients with disc drusen may be at additional risk of developing NAION, including postpartum NAION.

A 31-year-old, pregnant Norwegian female presented with acute, painless, bilateral, simultaneous vision loss after a complicated pregnancy and delivery. Her medical history was unremarkable, and she had no prior anemia, deep venous thrombosis, pulmonary embolus, or other vasculopathic risk factors. Her body mass index was 20.55 kg/m^2. Her surgical history was significant for adenoidectomy and tonsillectomy. She was not taking any medications. She had no known medical allergies. Her obstetric history was unremarkable, and she had no prior pregnancies or miscarriages. She immigrated from Norway 1 year earlier and was working as a nurse anaesthetist. The patient was married, used alcohol socially, and denied a history of smoking and illicit drug use. Her mother had optic disk drusen, but no other significant family history was noted. Her review of symptoms was unremarkable. Her ocular history was significant for normal eye examinations except for bilateral optic disc drusen (ODD), which were asymptomatic.

The patient's labor and delivery took place in Norway. According to her transferred medical records, the patient had an uneventful pregnancy and there was no proteinuria, eclampsia, or preeclampsia noted. She underwent epidural anaesthesia and delivered via spontaneous vaginal delivery. The delivery was complicated by a difficult, protracted labor, and her blood pressure rose to a maximum of 150/85 mm Hg. Uterine atony and postpartum hemorrhage were noted. She experienced 2 litres of blood loss, and her hemoglobin was low, at 8.2 g/dL. No blood transfusion was administered. Her baby weighed 4 kg and required a forceps delivery due to cephalopelvic disproportion. In the recovery room, the patient immediately noted new onset blurred vision in both eyes.

Magnetic resonance imaging (MRI) of the head was normal except for some T2 signal change consistent with ischemic optic neuropathy. No posterior nerve ischemia was noted. There was no evidence for stroke or pituitary apoplexy. A diagnosis of presumed NAION was made but no treatment was initiated. Testing for a hypercoagulable state was negative.

One year after the visual loss, the patient was referred to our clinic. Neuro-ophthalmic examination revealed best-corrected visual acuity of 20/20 OU. Ishihara color plates were correctly identified in 14/14 plates OU. Humphrey visual field testing showed bilateral arcuate defects superiorly and inferiorly with a mean deviation of −9.73 dB OD and −20.23 dB OS. The pupils measured 3 mm in the dark and 2 mm in the light bilaterally, and a relative afferent pupillary defect was noted OS. Motility examination was full. Intraocular pressure measured 11 mm Hg OD and 13 mm Hg OS. External and slit-lamp examinations were unremarkable OU. Fundus examination (Fig. 1) showed ODD and superimposed optic

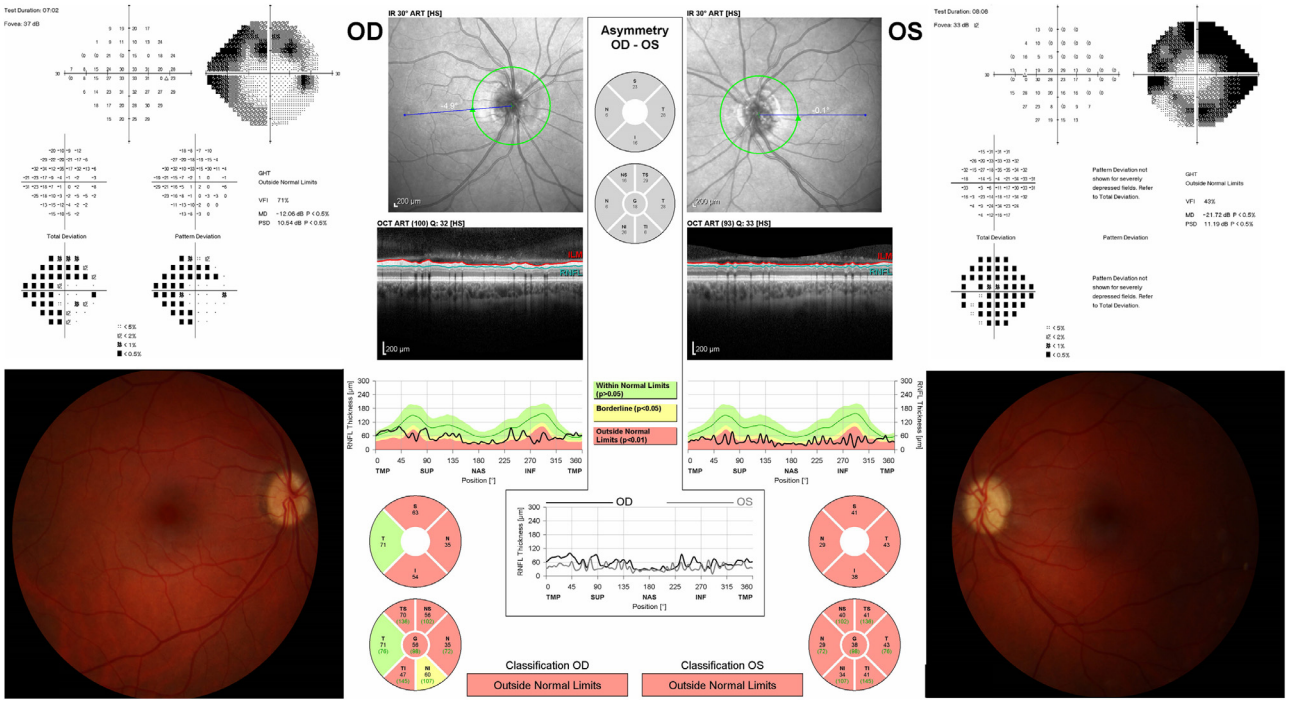

Fig. 1—Fundus examination showed optic disc drusen and superimposed optic atrophy OU. Optical coherence tomography demonstrated global retinal nerve fibre layer loss at 56 μm OD and 39 μm OS. Humphrey visual field testing showed bilateral arcuate defects superiorly and inferiorly with a mean deviation of −9.73 dB OD and −20.23 dB OS.

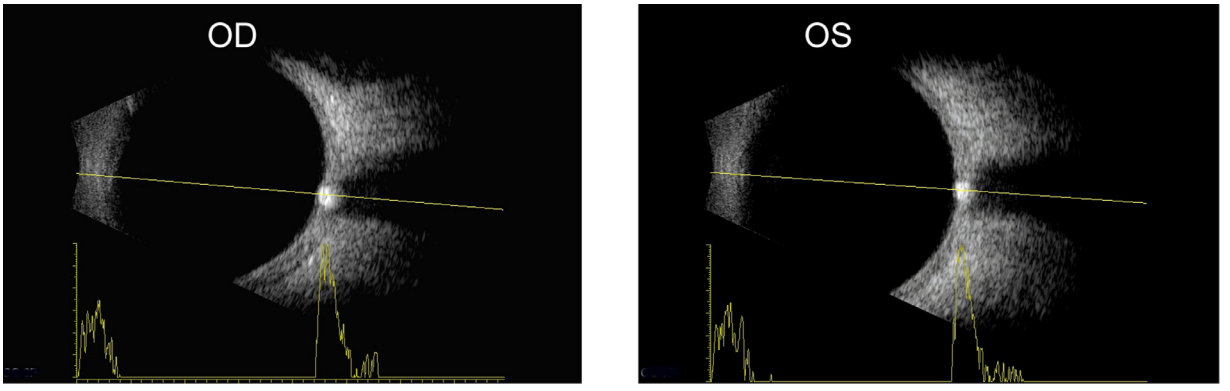

Fig. 2—Ultrasonography of the left and right orbits shows calcified optic disk drusen bilaterally.

atrophy OU. Optical coherence tomography demonstrated global retinal nerve fibre layer loss at 56 μm OD and 39 μm OS. Orbital ultrasonography confirmed calcified optic disk drusen bilaterally (Fig. 2). A repeat MRI of the head (Fig. 3) revealed no acute ischemia or abnormal post-contrast enhancement. A stable, linear focus of T2-weighted-fluid-attenuated inversion recovery (T2 FLAIR) hyperintensity in the left parietal, periventricular white matter was noted, likely due to chronic insult of unclear etiology. No abnormal signals or masses were noted in the optic nerves, intraorbital contents, cavernous sinuses, optic chiasm, or sellar and supersellar regions. Both optic nerves were noted as slightly smaller in size, compatible with the given clinical history of optic atrophy. No serial change in optic nerve intensity was noted.

We did not see this patient acutely, and so the possibility of both anterior (disc edema) and posterior levels of ischemia cannot be excluded. However, based on the available patient history and our findings, NAION remains the presumed diagnosis. NAION has been associated with a variety of complications, including hypercholesterolemia, diabetes mellitus, hypertension, coagulation disorders,[1] nocturnal systemic hypotension, migraine, and acute blood loss.[2] Our patient's case of NAION is notable due to the

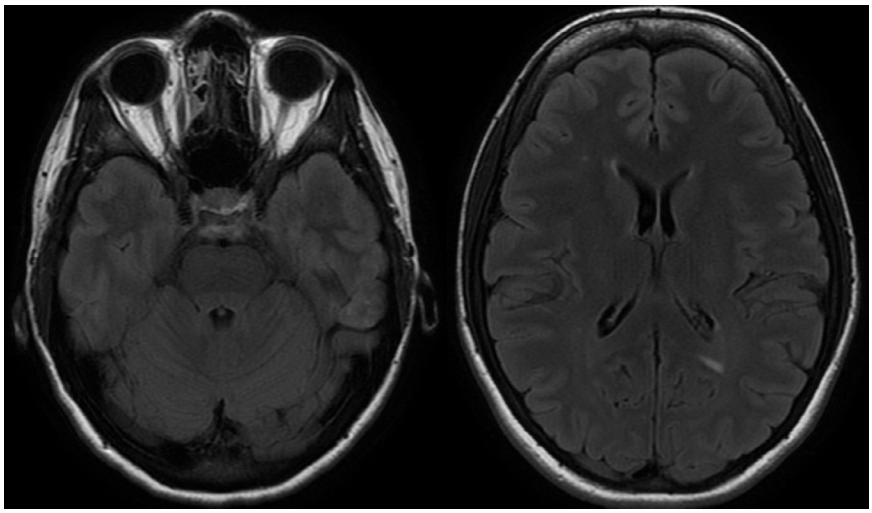

Fig. 3—A stable, linear focus of T2 FLAIR hyperintensity is visible in the left parietal, periventricular white matter. Both optic nerves were noted as slightly smaller in size, compatible with the given clinical history of optic atrophy.

patient's young age, the presence of ODD, and the rarity of NAION from delivery complications.

ODD, a congenital disc abnormality involving crowded and cup-less nerve heads, has been associated with NAION. In ODD, an abnormal nerve head may exacerbate the small cup-disc ratio often seen in NAION, increasing the probability of decreased perfusion pressure of the optic nerve head and NAION-causing ischemia.[3] An earlier mean diagnosis age, better visual acuity, and higher likelihood of preceding, transient visual obscuration have been associated with patients with NAION and ODD compared to NAION alone, although the visual field loss topography patterns present similarly in both cohorts.[3,4] This age discrepancy may be explained by drusen bodies increasing the risk of NAION earlier in life by directly causing vessel infarction, or by nerve fibre layer thinning typical of ODD decreasing infarction risk later in life through a reduced crowding in the optic disc over time.[3] Prevalence of vascular risk factors has been noted as both insignificant[3] and significant[4] by separate studies. Our patient with ODD presented at an atypically young age without preceding episodes of transient visual obscuration and without prior vascular risk factors.

While literature often cites 50 years and older as the age threshold for NAION presentation, rare cases of NAION in patients less than 50 years old have been reported,[5] often associated with an underlying risk factor or abnormality, such as primary antiphospholipid syndrome,[6] hemodialysis accompanying end-stage renal disease,[7] hypercoagulable state,[5] hepatitis C treatment,[8] diabetes mellitus,[9] hyperhomocysteinemia,[10] menorrhagia,[11] and abortion.[12] In contrast, our patient's history at presentation and past medical records indicated no significant risk factors other than ODD.

NAION reported in the context of pregnancy is rare. Presentation has been reported both predelivery[13] and postdelivery,[2,14] with both vaginal[14] and Caesarian section[2,13] delivery. Although some cases report pre-existing risk factors such as preeclampsia,[13,14] other cases report no risk factors before NAION presentation. Prior cases have reported ODD[15] as a

Table 1—Prior cases of pregnancy-related nonartertitic anterior ischemic optic neuropathy (NAION)

	This Study	Giridhar and Freedman[14]	Gupta et al.[2]	Beck et al.[13]	Mehta et al.[15]
NAION timing	Postdelivery	Predelivery	Postdelivery	Predelivery	Predelivery
Disc edema	None	Unilateral	Unilateral	Bilateral	Unilateral
Vision loss	Bilateral	Unilateral	Unilateral	Bilateral	Unilateral
Vasculopathic risk factors	None	Preeclampsia, gestational diabetes mellitus	None noted	Preeclampsia	None noted
Drusen	Yes	Unknown	None	Unknown	Yes
Delivery method	Vaginal	Vaginal	Caesarean section	Caesarean section	Unknown
Blood loss	2000 mL	Unknown	500 mL	Unknown	Unknown
Delivery complications	Uterine atony, postpartum hemorrhage, protracted labor	Unknown	Hypotensive episode, possible dural puncture during epidural anaesthesia	Unknown	Unknown
Treatment	None	None	None	Unknown	Unknown
Response	No vision improvement	Partial vision improvement at 6-month follow-up	No vision improvement	Vision improvement at 3-day follow-up	Unknown

possible risk factor, although in different circumstances compared to our patient (Table 1).

There is no proven, effective treatment for NAION,[16] and the topic remains heavily contested: some studies have concluded that the risks of steroid treatment outweigh the benefits,[17] whereas other literature has documented steroid treatment for patients, with the rationale that early treatment may improve or prevent disk edema.[18] Our patient was not treated with steroids, and her vision has not improved 1 year after onset. Our patient intends to pursue a second pregnancy. Possible countermeasures to prevent or mitigate postpartum hemorrhage were discussed, including hospital delivery, treatment of pre-existing anemia, iron supplementation including blood-typing, and cross-matching before delivery in preparation for autologous blood transfusion, if necessary.

In summary, we report a case of a young, healthy female with ODD presenting with bilateral NAION from postpartum blood loss. Clinicians should be aware of the risk for NAION in patients with ODD, especially pregnant and postpartum patients.

Patient Consent: This report does not contain any personal information that could lead to the identification of the patient. Thus, consent to publish this case was not obtained.

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A case of left congenital homonymous hemianopia associated with right occipital porencephaly

Congenital homonymous hemianopia is a rare entity, often missed during childhood and incidentally diagnosed during adulthood, given that the patients are usually unaware of their visual field defect (VFD).[1] The causes of congenital homonymous hemianopia include occipital porencephaly, arteriovenous malformation, and Sturge Weber syndrome.[2] The hallmark of congenital homonymous hemianopia is the homonymous pattern of transsynaptic retinal nerve fibre layer (RNFL) degeneration as seen on fundoscopy and optic nerve optical coherence tomography (OCT).[2] Here, we report a patient with left congenital homonymous hemianopia, and an interesting constellation of multiple ophthalmic and neuro-ophthalmic