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Brief Communication

Neurovascular Compression in the Anterior Visual Pathway: A Case Series

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ABSTRACT: A retrospective review of 29 patients with neurovascular compression syndrome (NVCS) involving the anterior visual pathway was conducted. Various patterns of NVCS and visual defects were identified, most commonly involving the optic nerve and internal carotid artery. Most patients were stable, except one with progressive visual field defects. Although mostly asymptomatic, NVCS can rarely cause compressive optic neuropathy. NVCS should be kept in the differential diagnosis of normal tension glaucoma, especially with progressive visual loss despite treatment. Patients with progressive visual loss may require decompression surgery. Non-contrast computed tomography scan may miss NVCS, and magnetic resonance imaging is diagnostic.

RÉSUMÉ : Compression neurovasculaire dans la voie visuelle de la région antérieure : une série de cas. Nous avons effectué une étude rétrospective portant sur 29 patients atteints d'un syndrome de compression neurovasculaire (SCNV) impliquant la voie visuelle de la région antérieure. De nombreux types de SCNV et d'anomalies visuelles ont ainsi été identifiés ; ils impliquaient le plus souvent le nerf optique et l'artère carotide interne. À l'exception d'un patient souffrant d'anomalies progressives du champ visuel, la plupart des autres patients ont donné à voir un état stable. Bien que le plus souvent asymptomatiques, les SCNV peuvent rarement entraîner une neuropathie optique compressive. Qui plus est, les SCNV doivent être maintenus dans le diagnostic différentiel du glaucome à pression normale (GPN), surtout en cas de perte visuelle progressive malgré un traitement prodigué. Il est aussi à noter que les patients présentant une perte de vision progressive peuvent nécessiter une chirurgie de décompression et qu'un examen de tomodensitométrie sans contraste pourrait ne pas détecter un SCNV, un examen par IRM s'imposant alors pour obtenir un diagnostic.

Keywords: Neurovascular compression syndrome; Visual field defect; Neuro-ophthalmology; Neuroimaging

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Neurovascular compression syndrome (NVCS) is an abnormal intersection between a cranial nerve and blood vessel.[1] NVCS can be asymptomatic or symptomatic depending on cranial nerve involvement. Examples include trigeminal neuralgia, hemifacial spasm, vestibular paroxysmia, and glossopharyngeal neuralgia.[1] This series details the presentations of uncommon NVCS, specifically involving the anterior visual pathway. The optic nerve carries sensory information from the ganglion cells of the retina along the optic chiasm and tract before synapsing in the lateral geniculate nucleus. An artery with normal caliber or dolichoectasia (artery with dilatation or tortuous component) can cause a compression. Although often incidentally found, NVCS can lead to symptomatic vision loss in various patterns. Diagnosis is based on clinical and radiological evaluation. For any suspected nerve compression syndrome, magnetic resonance imaging (MRI) of the brain is diagnostic.[2] Thorough investigation is required to rule out other causes of neurological deficits. The purpose of this case series is to study the effect of neurovascular compression syndrome involving the anterior visual pathway.

Individuals with NVCS involving the anterior visual pathway on diagnostic imaging were identified by one author (ANES) in their outpatient neuro-ophthalmology clinic between 2014 and 2020. Patients were included if a NVC was identified on imaging. Patients referred for atypical glaucoma were also followed by a glaucoma specialist. This case series is a retrospective analysis of their charts, including symptoms, visual acuities, relative afferent pupillary defects (RAPD); optic disk appearances including neuroretinal rims and cup-to-disk ratios, optical coherence tomography (OCT) images of the retinal nerve fiber layer (RNFL), Humphrey visual fields (HVF), and diagnostic images (MRI) if available. OCT images were collected in a subset

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Table 1: Summary of patient demographics and clinical information

ID	Age	Sex	Vessel	Nerve	Visual Acuity		Visual Defect	RAPD	Cup-Disk-Ratio and ON		Incidental	Referral Details
					R	L			R	L		
1	53	F	R Supraclinoid ICA	R pre-chiasmatic ON R OC	20/20	20/20	None	No	0.3	0.3	Yes	Glaucoma suspect
2	62	M	R ICA dolichoectasia	R pre-chiasmatic ON	20/20	20/20	R nasal step, asymptomatic	No	0.7, temporal pallor	0.6	No	Normal tension glaucoma
3	82	F	L supraclinoid ICA dolichoectasia	L OC L Pre-chiasmatic ON	20/20	20/25	Incomplete R homonymous hemianopia vs. partial junctional scotoma	L	0.6, pallor	0.6, pallor	No	Progressive visual field defect OS (10 years)
4	97	F	R A1 of ACA	R pre-chiasmatic ON	20/25	20/25	None	No	0.4	0.4	Yes	Amaurosis fugax OD
5	67	M	L supraclinoid ICA	L OC	20/25	20/25	R inferior arcuate	No	0.4	0.3	Yes	Known Glaucoma OD
6	57	M	R ACA dolichoectasia	R OT	20/20	20/20	None	No	0.4	0.4	Yes	Diabetic 3rd nerve palsy
7	59	M	R supraclinoid ICA	R ON	20/25	20/20	R inferior depression, asymptomatic	R	0.6, diffuse pallor	0.5	No	Pallor OD. Previous retinopexy OS
8	64	M	R PCA	R OT	20/20	20/20	Incongruous L superior homonymous quadrantanopia, denser R nasally	R	0.6, temporal pallor	0.5, temporal pallor	No	Glaucoma suspect, visual defect gradually worsened since 2005
9	71	M	R supraclinoid ICA dolichoectasia	R OC R OT	20/30	20/400	Depressed bilaterally, asymptomatic (L due to macular hole)	R	0.8	0.6	No	Macular hole OS, high myope with severe myopic degeneration
10	73	F	Duplicated R A1 of ACA	R pre-chiasmatic ON	20/400	20/40	R superior altitudinal	R	0.8, pallor	0.6	No	Progressive right optic atrophy
11	57	M	L A1 of ACA	L ON	20/25	20/30	Bilateral biarcuate (glaucoma)	No	0.4	0.4	Yes	Known Glaucoma. MRI for vertigo and facial numbness
12	79	M	L A1 of ACA	L OC	20/40	20/60	Cecocentral scotoma	No	0.3, temporal pallor	0.3, temporal pallor	Yes	Bilateral central scotomas
13	79	M	L supraclinoid ICA L ACA	L ON L OC	20/20	20/20	L biarcuate	None	0.7, temporal pallor	0.9	No	Visual decline OS since 2006. Left optic neuropathy.
14	66	M	R ICA	R pre-chiasmatic ON	20/20	20/30	L inferior depression	L	0.4	0.4	Yes	Remote left NAION
15	62	F	R ICA L ICA	R pre-chiasmatic ON L pre-chiasmatic ON	20/20	20/20	None	No	0.5	0.5	Yes	Referred for nonspecific blurred vision OU
16	28	F	R supraclinoid ICA	R pre-chiasmatic ON	20/20	20/20	None	No	0.3	0.3	Yes	Benign unilateral episodic mydriasis
17	79	F	R A1 of ACA	R pre-chiasmatic ON	20/25	20/25	Bitemporal pseudohemianopia	No	0.3	0.3	Yes	High myope with bitemporal pseudohemianopia
18	71	F	R A1 of ACA	R pre-chiasmatic ON	20/30	20/25	R central scotoma	R	0.3	0.3	No	Referred after normal retinal exam. Central vision loss OD, progressed over 2 years
19	46	F	R ICA	R OC	20/25	20/20	None	No	0.4	0.4	Yes	Right abducens palsy

(*Continued*)

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Table 1: (*Continued*)

ID	Age	Sex	Vessel	Nerve	Visual Acuity		Visual Defect	RAPD	Cup-Disk-Ratio and ON		Incidental	Referral Details
					R	L			R	L		
20	70	M	L ICA	L pre-chiasmatic ON	20/25	20/25	None	No	0.5	0.5, temporal pallor	No	Incidental temporal pallor OS. Preserved visual function
21	73	M	L PCA	L OT	20/20	20/20	None	No	0.3	0.3	Yes	Downbeat nystagmus
22	46	F	L supraclinoid ICA	L pre-chiasmatic ON	20/20	20/20	None	No	0.3	0.3	Yes	Referred for bilateral nonspecific blurred vision
23	69	F	L ophthalmic A	Intra-orbital ON	20/40	CF	L severely depressed (NAION)	L	0.1	atrophy	Yes	Referred for NAION OS
24	68	M	Dolichoectatic basilar A	R OT R OC	20/25	20/30	L incongruous homonymous hemianopia	No	temporal pallor	temporal pallor	No	Slowly progressing left visual loss. Normal color vision
25	62	M	R ICA	R pre-chiasmatic ON	20/20	20/20	None	No	0.4	0.4	Yes	Pituitary microadenoma
26	53	M	L PCA	L OT	HM	20/20	Normal OS	R	atrophy, crowded	0.1	Yes	Right optic atrophy. Painless R visual loss (silent NAION)
27	79	M	R supraclinoid ICA L supraclinoid ICA	R pre-chiasmatic ON L pre-chiasmatic ON	20/30	20/30	L partial superior arcuate defect R inferior nasal step	R	0.7	0.6	Yes	Progressive visual decline OD, treated for glaucoma.
28	78	M	L supraclinoid ICA	L pre-chiasmatic ON	20/50	20/50	Nonspecific changes	No	0.2	0.2	Yes	Diabetic 3rd nerve palsy
29	58	F	L ACA	L pre-chiasmatic ON	20/25	20/25	Subtle L superior arcuate defect	L	0.4	0.6	Yes	Facial numbness

A = Artery; ACA = Anterior cerebral artery; CDR = Cup-to-disk ratio; HVF = Humphrey visual field; ICA = Internal cerebral artery; L = Left; LCA = Long circumferential artery; N = Nerve; NAION = Non-arteritic anterior ischemic optic neuropathy; NVC = Neurovascular conflict; OC = Optic chiasm; OD = Right eye; ON = Optic nerve; OS = Left eye; OT = Optic tract; PCA = Posterior cerebral artery; R = Right; RAPD = Relative afferent pupillary defect; VA = Visual acuity.

of cases, and most patients were monitored with serial HVFs. Ganglion cell complex analysis was completed in some patients in the last three years.

Twenty-nine patients (17 male, 12 female) with a mean age of 65.8 (standard deviation of 13.5) demonstrated various combinations of neurovascular compression syndrome (Table 1). Seven patients had multiple neurovascular compressions (24.1%) creating a total of 36 compressions (cases 1, 3, 9, 13, 15, 24, and 27). Both anterior and posterior circulations were involved, as well as all components of the anterior visual pathway. Specific neurovascular compressions are detailed in Table 1, (see column 4 for vessel and column 5 for nerve involved). Dolichoectasia was seen in five of the 29 patients (17.2%).

The most common NVCS (5/36, 13.8%) included the prechiasmatic optic nerve and supraclinoid internal cerebral artery. NVCS was an incidental finding in 19 patients (65.5%). These patients were imaged for other purposes such as oculomotor nerve palsy, nonarteritic anterior ischemic optic neuropathy, and downbeat nystagmus. They had no RAPD, optic disk changes, or visual defects secondary to the NVCS. 10/29 cases (34.5%) presented with visual field defects in variable patterns which localized to the NVCS site (cases 2, 3, 7–10, 13, 18, 20, and 24). OCTs were collected when available (72.4%, 21/29) (Table 2). When OCTs were obtained, RNFL atrophy was evident in many symptomatic cases of NVCS. Visual defects varied based on the site of the compression, which included retinal nerve fiber layer defects (such as nasal step, arcuate defects, and central scotomas), diffuse loss, incongruous homonymous hemianopia, and homonymous quadrantanopia. Eleven (37.9%) patients had normal visual fields. 31.0% (9/29) of patients (cases 1–3, 5, 8, 10, 11, 13, and 27) were referred to rule out non-glaucomatous pathology by glaucoma specialists. 55.5% (5/9) of these (cases 2, 3, 8, 10, and 13) patients were found to have visual field defects matching the NVCS, rather than normal tension glaucoma (NTG).

96.5% (28/29) of patients remained stable from the visual perspective during follow-up visits. Figure 1 details HVFs for patients 8, 13, and 24. Figure 2 includes details on T2-weighted MRI images for these patients. Two among the five patients with optic tract compression showed matching homonymous visual field defects (cases 9 and 24). Case 24 presented with significant compression on optic tract and optic chiasm by dolichoectatic basilar artery and showed progression of homonymous quadrantanopia to incongruous homonymous hemianopia. All other patients have remained stable without worsening of vision or visual fields.

This case series is a clinical summary of 29 patients with NVCS involving the anterior visual pathway. Although uncommon, NVCS can be asymptomatic, and discovered incidentally on routine imaging, or cause progressive visual loss

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Table 2: Summary of most recent available OCT results

	Mean RNFL Thickness (μm)		Atrophy Pattern and GCC (when obtained)	
ID	R	L	R	L
2	58	70	Superior and inferior RNFL atrophy	Temporal RNFL atrophy
5	71	80	Superior RNFL atrophy, superior GCC hemiatrophy (66 μm)	Normal, GCC normal (86 μm)
7	57	62	Superior, temporal, and inferior RNFL atrophy, circumferential GCC atrophy (50 μu)	Superior and inferior RNFL atrophy, circumferential GCC atrophy (62 μu)
8	73	77	Superior and inferior RNFL atrophy, mild thinning	Superior and inferior RNFL atrophy, mild thinning
9	69	65	Superior and inferior RNFL atrophy, GCC 76 μu	Superior and inferior RNFL atrophy, circumferential GCC atrophy (42 μu)
11	57	55	Superior and inferior RNFL atrophy	Superior and inferior RNFL atrophy
12	77	78	Normal RNFL, circumferential GCC atrophy (59 μu)	Circumferential GCC atrophy (62 μu)
13	77	56	Mild superior RNFL thinning	Superior and inferior RNFL atrophy
14	79	64	Normal	Superior, temporal, and inferior RNFL atrophy
16	95	92	Normal	Normal
17	109	100	Normal, GCC 80 μm	Normal, GCC 79 μu
18	90	93	Normal	Normal
20	73	64	Superior and inferior RNFL atrophy, circumferential GCC atrophy (58 μu)	Temporal, superior, and inferior RNFL atrophy, circumferential GCC atrophy (56 μu)
21	101	98	Normal, GCC 74 μm	Normal, GCC 78 μm
22	97	92	Normal, GCC 76 μm	Normal, GCC 77 μm
23	68	59	Superior and inferior RNFL atrophy	Superior and inferior RNFL atrophy
24	62	53	Superior, temporal, and inferior RNFL atrophy, circumferential GCC atrophy (51 μu)	Superior, temporal, and inferior RNFL atrophy, circumferential GCC atrophy (51 μu)
25	89	95	Normal, GCC 76 μm	Normal, GCC 78 μm
26	57	136	Superior, temporal, and inferior RNFL atrophy	Inferior RNFL atrophy and elevations due to NAION
27	80	81	Superior and inferior RNFL atrophy	Superior and inferior RNFL atrophy
29	76	66	Superior RNFL atrophy, GCC temporal atrophy (68 μm)	Superior and inferior RNFL atrophy, circumferential GCC atrophy (66 μu)

GCC = Ganglion cell complex; L = Left; R = Right; RNFL = Retinal nerve fiber layer.

evident on HVF.[3,4] Because of the nature of visual defects and the number of patients referred for glaucoma, it is important to consider NVCS in the differential diagnosis of NTG,[3] While NTG results in four characteristic patterns, defects in NVCS have not been well characterized.[5] This is particularly important when the field changes are progressive despite adequate diurnal intraocular pressure control, or when the changes are asymmetric in the setting of similar bilateral intraocular pressures. Visual loss occurs from direct compression by the vessel, although vascular pulsations have also been hypothesized to be an etiological factor.[4,6]

Decompression surgery is usually reserved for patients with progressive visual loss. There have been a few isolated case reports and case series with successful decompression surgeries and visual improvement.[4,6–9] but there are no randomized studies given the rarity of this condition. Currently, the role of surgical decompression is not as well established as in other types of NVCS such as trigeminal neuralgia. Failure of early diagnosis has been postulated for unsatisfactory postoperative visual outcome.[9]

A case series published by Jacobson (1994) discussed the clinical features of 18 patients with optic nerve compression by the supraclinoid carotid artery.[3] All patients had optic neuropathy (6 bilateral, 12 unilateral). Similar to our cases, they reported a variety of visual field defects with abnormal disks (pallor, atrophy, and cupping). In a previous case series of 10 cases, only one patient had progressive visual loss that required decompression.[10] Compared to these previous reports, ours is the largest cohort studied.

In patients with trigeminal neuralgia who have similar pathomechanism resulting in compressive neuropathy, it has been noted that a neurovascular contact alone is not enough for diagnosis of NVCS.[11] It is important to radiologically identify nerve thinning, arterial imprint or grooving, or distortion of the course of the nerve, which may lead to the symptomatic presentations.[11] Arterial imprint or grooving is focal thinning of the nerve at the neurovascular compression site. Intraoperative histopathological study in patients with trigeminal neuralgia secondary to NVCS has shown demyelination and axonal loss.[12] A combination of high-resolution T2-weighted images, MR

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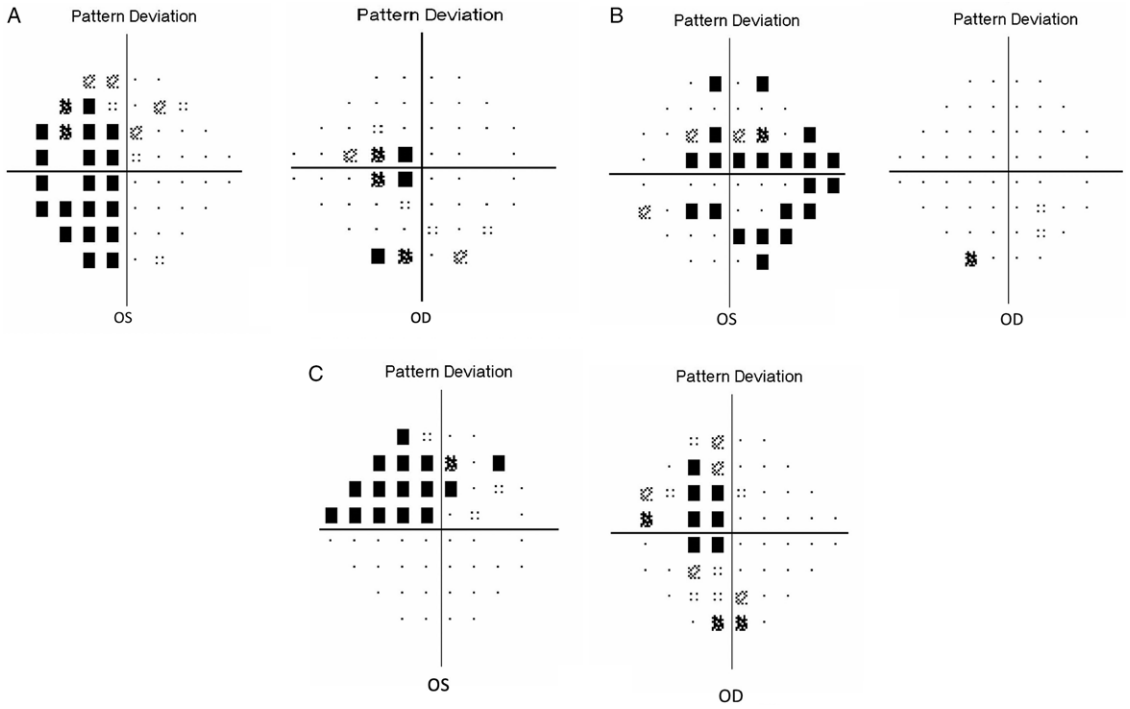

Figure 1: 24–2 SITA Humphrey visual fields. (A) Patient # 24. Incongruous left homonymous hemianopia. Neurovascular compression: Right optic tract and dolichoectatic basilar artery. (B) Patient # 13. Left biarcuate defect. Neurovascular compression: Left pre-chiasmatic optic nerve and left ACA. (C) Patient #8. Incongruous left homonymous quadrantanopia. Neurovascular compression: Right optic tract and right ACA.

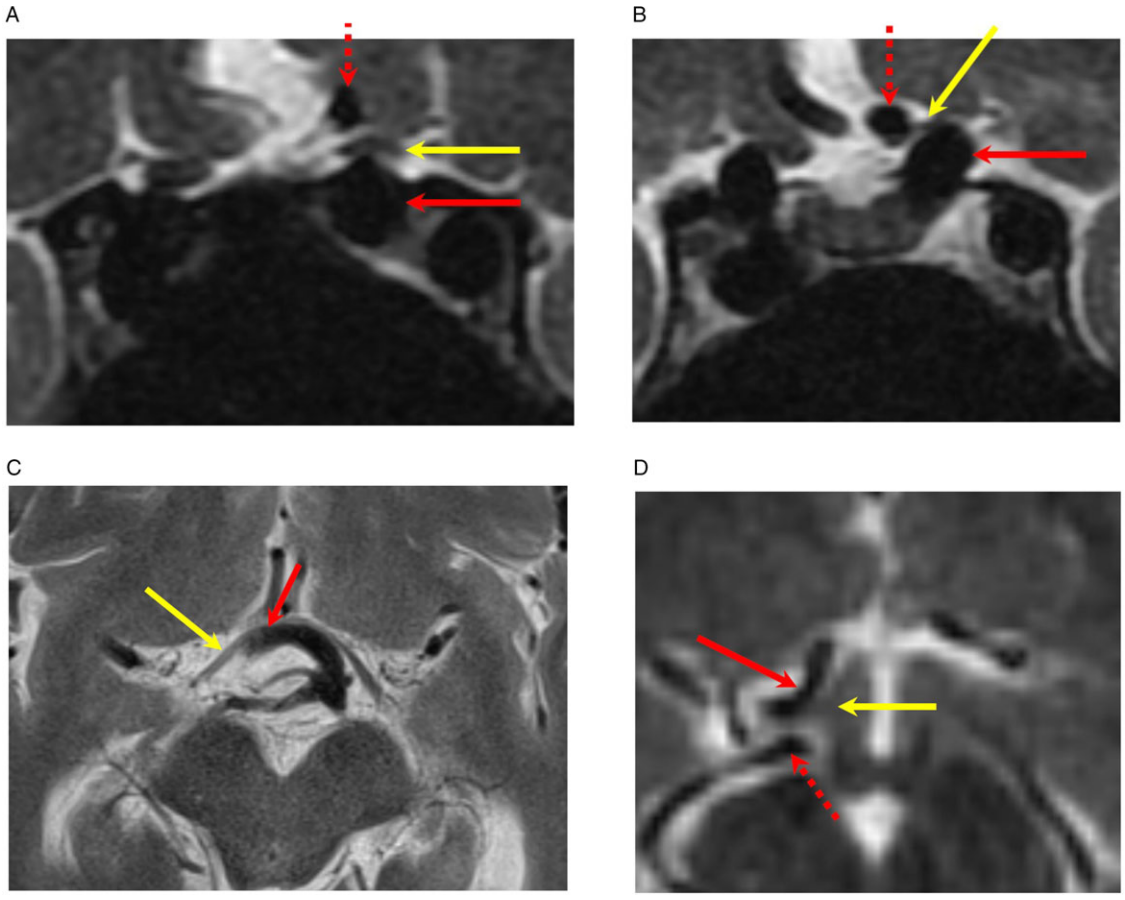

Figure 2: MR images. (A and B) Coronal T2-weighted images demonstrate compression of pre-chiasmatic left optic nerve (yellow arrow) by distal cavernous and paraclinoid left internal carotid artery (solid red arrow). The dashed red arrow is the left anterior cerebral artery (patient 13). (C) Axial T2-weighted image demonstrates compression of the right optic tract (yellow arrow) by the dolichoectatic basilar artery (red arrow) (patient 24). (D) Axial T2-weighted image demonstrates compression of the right optic tract (yellow arrow) by the right anterior cerebral artery (solid red arrow). The dashed red arrow is the right posterior cerebral artery. (Patient 8).

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angiogram, and volumetric gadolinium-based contrast-enhanced T1-weighted image is recommended for detection of NVCS.[1] In terms of NVCS of optic pathways, the culprit arteries are large enough that they can be easily visualized as vascular flow voids on high-resolution T2-weighted images, and therefore, there may not be a need for contrast-enhanced MR angiogram in these cases.

Although mostly asymptomatic, NVCS can cause compressive optic neuropathy leading to significant visual disturbances. The majority NVCS is found incidentally and do not progress to this point. Decompression has been successful in isolated cases of progressive visual loss; however, there are no controlled studies. Patients are diagnosed using MRI imaging. It is important to include neurovascular compression syndrome in the differential diagnosis for normal tension glaucoma, especially with progressive visual field loss despite treatment. Patients with progressive visual field loss may require decompression surgery.

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