



Clinical and ultrasonographic features in anterior ischemic optic neuropathy

Klinička i ultrasonografska obeležja prednje ishemičke optičke neuropatije

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Abstract

Background/Aim. Anterior ischemic optic neuropathy (AION) represent a segmental infarction of the optic nerve head which is supplied by the posterior ciliary arteries. There are two types of AION: non-arteritic (NA-AION) and arteritic (A-AION), due to giant cell arteritis (GCA). The aim of this work was to investigate the clinical features and ultrasound characteristics of the orbital vessels and superficial temporal and carotid arteries, in patients with unilateral acute AION in order to help differentiate newly diagnosed NA-AION from A-AION. **Methods.** In this prospective comparative, observational study, 62 consecutive patients with clinical suspicion of unilateral acute AION were examined at admission and in the first two months of evolution, following a protocol including color Doppler imaging (CDI) of the orbital vessels. **Results.** We found 12 patients with A-AION, all of them with biopsy-confirmed disease, and 50 patients with NA-AION. A-AION patients presented a combination of a history of amaurosis fugax before acute, painless, and severe vision loss in the affected eye, and a diffuse pale optic disc edema. In these patients, CDI of the orbital vessels indicated high resistance index (RI), with severe diminished blood flow velocities in all orbital vessels, in both orbits. In the NA-AION patients, none of these clinical symptoms were found and blood velocities and RI in posterior ciliary arteries were preserved. Typical sonographic feature in temporal arteritis as part of GCA was the „dark halo” sign. **Conclusions.** The ultrasound investigations enable prompt differentiation between NA-AION and A-AION.

Key words:

optic neuropathy, ischemic; risk factors; giant cell arteritis; diagnosis differential; ultrasonography, doppler, color.

Apstrakt

Uvod/Cilj. Prednja ishemička optička neuropatija (AION) predstavlja segmentni infarkt papile optičkog nerva vaskularizovanog preko posteriorne ciliarne arterije. Postoje dva tipa AION: arterijska (A-AION), koja je gotovo bez izuzetka, posledica arteritisa dzinovskih ćelija (GCA) i nearterijska (NA-AION). Cilj rada je bio da se istraže klinička i ultrazvučna obeležja orbitalnih krvnih sudova, temporalne superficijalne arterije i karotidnih arterija, koja bi bila od pomoći u diferencijalnoj dijagnostici akutne AION. **Metode.** Prospektivnom, uporednom opservacionom studijom obuhvacena su 62 uzastopna bolesnika sa sumnjom na jednostranu akutnu AION, koji su bili ispitani prilikom prijema i tokom prva dva meseca evolucije bolesti prema protokolu koji je uključivao kolor dopler snimanje orbitalnih krvnih sudova. **Rezultati.** Kod 12 bolesnika je ustanovljena i biopsijom potvrđena A-AION, dok je kod 50 bolesnika ustanovljena NA-AION. Kod bolesnika sa A-AION bolest se manifestovala prisustvom *amaurosis fugax* pre pojave akutnog, bezbolnog gubitka vida teškog stepena u zahvaćenom oku i difuznim bledim edemom optičkog diska. Kolor doplerom orbitalnih krvnih sudova ustanovljen je ozbiljno smanjen protok krvi u posteriornim ciliarnim arterijama izraženiji na zahvaćenoj strani i praćen visokim indeksom rezistencije (IR) u svim retrobulbarnim krvnim sudovima obe orbite. Kod bolesnika sa NA-AION kolor doplerom nisu registrovane opisane promene. Tipično ultrazvučno obeležje u temporalnom arteritisu, kao delu GCA, bio je „dark halo” znak. **Zaključak.** Ultrazvučno ispitivanje omogućava postavljanje brze diferencijalne dijagnoze između A-AION /NA-AION kod bolesnika sa akutnom AION.

Ključne reči:

neuropatija, optička, ishemička; faktori rizika; arteritis, temporalni; dijagnoza, diferencijalna; ultrasonografija, dopler, kolor.

Introduction

Anterior ischemic optic neuropathy (AION) represent an acute ischemic disorder involving the anterior part of the optic nerve, also called the optic nerve head (ONH), supplied by the posterior ciliary arteries (PCAs) – nasals and temporals¹⁻⁷. Blood supply blockage of the PCAs can occur with or without arterial inflammation. There are two types of AIONs: arteritic (A-AION), caused by a vasculitic mechanism due to giant cell arteritis (GCA), and non-arteritic (NA-AION)¹⁻⁷.

GCA is a primary vasculitis that concerns predominantly extracranial medium-sized arteries, especially branches of the external carotid artery (ECA), including the superficial temporal arteries (TAs)¹⁻⁸. The typically predominant extracranial vascular involvement (including the orbital vessels) is explained by the affinity of inflammation to the elastic fibers in the media; as intracranial arteries have less elastic fibers in the media, they are seldom involved³⁻⁸. The diagnosis of GCA requires, according to the criteria established by the American College of Rheumatology, age more than 50 years at clinical disease onset, new headache in the temporal area, temporal artery tenderness, and/or reduced pulse, jaw claudication, systemic symptoms, erythrocyte sedimentation rate (ESR) exceeding 50 mm/h, elevated C-reactive protein (CRP), and typical histologic findings (granulomatous involvement) in temporal artery biopsy (TAB)¹⁻⁸. Approximately 20% of patients with GCA have ophthalmologic complications, including visual loss secondary to A-AION, or central retinal artery occlusion¹⁻⁸. These are generally early manifestations due to the vasculitic involvement of retrobulbar (orbital) vessels deriving from the ophthalmic artery (OA), more precisely the PCAs and the central retinal artery (CRA)¹⁻⁸.

NA-AION is a multifactorial disease that results in hypoperfusion and ischemia of the ONH, with multiple risk factors that contribute to its development (nocturnal arterial hypotension, etc)¹⁻⁷. According to Biousse and Newman¹, the most important contributing factor to NA-AION is represented by congenital and physiologic small cups. They mentioned that the size of the ONH (optic disc) and the physiological cup depend on the size of the scleral canal; a small scleral canal will result in a small cup (with a crowded ONH and a small cup-to-disc ratio)^{1,2,9}. The presence of a “disk at risk”, with structural crowding of nerve fibers (crowded disk) and reduction of the vascular supply will impair perfusion of the ONH to a critical degree^{1-6,9}.

The main purpose of our study is to show the essential role of color Doppler imaging (CDI) of orbital vessels in order to quickly differentiate the mechanism of AION (arteritic, versus non-arteritic); the former should be treated promptly with systemic corticosteroids to prevent further visual loss of the fellow eye.

Methods

In this prospective, comparative and observational study, we included 62 consecutive patients who presented, in our ophthalmology and neurology departments from June

2012 through February 2015, with clinical suspicion of acute unilateral AION.

The study was approved by the “Victor Babes” University of Medicine and Pharmacy Local Research Ethics Committee. All patients gave informed consent and were examined following a complex protocol, including:

1 – a complete history of all previous or current systemic diseases;

2 – an ophthalmological examination, (conducted by an ophthalmologist at the presentation, at 2 weeks, at 1 month and at 2 months), including visual acuity with the Snellen visual acuity chart, visual fields with a Goldmann perimeter, relative afferent pupillary defect, intraocular pressure, slit-lamp examination of the anterior segment, lens and vitreous, direct ophthalmoscopy and color fundus photography (both methods were used at the presentation and repeated at 2 weeks after the onset of visual loss), and fluorescein fundus angiography which was performed during the first 2 days after the presentation (acute stage of AION);

3 – a physical examination (including possible contralateral neurologic signs such as hemiparesis, the inspection and palpation of the TAs) was performed at the presentation by a neurologist and an internist in order to detect an eventual stroke or a temporal arteritis (as part of GCA);

4 – a CDI of orbital vessels was realized with a 10 MHz linear probe for detecting (by color Doppler sonography) and measuring (by spectral analysis pulsed Doppler sonography) the blood flow in the orbital vessels: the OAs, the CRAs, the PCAs (nasal and temporal), and the superior ophthalmic veins. Blood flow towards the transducer was depicted as red and flow away from the transducer was colored blue: a) the CRA was identified just below the optic disc (< 1 cm) and had a forward red-coded blood flow; b) the nasal and temporal trunks of PCA were identified along both sides of the optic nerve. The arteries had a forward red-coded blood flow; c) the OA was identified deeper in the orbit usually before crossing the optic nerve. It had a forward red-coded blood flow. The Doppler sample gate placed on the detected vessel was 15 mm. When the orbital vessels were not parallel to the ultrasound beam, we performed an angle correction between 0–60°. Also, a spectral velocity analysis was performed. The peak systolic velocity (PSV) and end-diastolic velocity (EDV) were calculated for each vessel. The resistance index (RI) was automatically calculated according to the following equation: $RI = (PSV - EDV) / PSV$. Absent signals not corresponding to ipsilateral internal carotid occlusive disease were classified as Doppler sonographic findings typical of GCA of the orbital arteries (occlusion of an orbital artery). Serial CDI examinations of the orbital blood vessels were performed at the presentation, at 1 week, and at 1 month on all AIONs patients.

5 – Extracranial arteries were examined with a 7.5–10 MHz linear array transducer, combining B mode and color coded Doppler/pulsed-wave Doppler ultrasound duplex sonography (EDS), looking for an internal carotid artery's (ICA) source of emboli and with a 10 MHz linear probe for the examination of ECA branches, especially temporal arteries (TAs). Color box steering and beam steering were maximal and the color covered the artery lumen exactly be-

cause using these machine adjustments, sensitivities and specificities of the temporal arteritis diagnosis are higher. We examined both common superficial TAs and their frontal and parietal rami in longitudinal and transverse planes. Concentric hypoechogenic mural thickening (also called halo) was considered as an ultrasonographic finding typical of GCA.

Arterial segment was considered stenosed when PSV was more than twice than in the pre-stenotic segment with wave forms demonstrating turbulence and reduced velocity beyond the stenosis. Acute occlusion was considered if the ultrasound (US) showed hypoechoic material in the artery lumen with absence of color signals.

The first two authors of the study performed both CDI of orbital vessels and EDS. The first investigator, who was blinded to the patients ophthalmological status, looked only for detecting and measuring orbital and extracranial vessels blood flow. Discrepancies were resolved by consensus.

EDS was performed at the presentation, 2 weeks and at 1 month on all A-AIONs patients, and at the presentation on all NA-AION patients.

6 – Electrocardiogram (ECG) in all patients, and transthoracic echocardiography (TTE) in selected cases were performed in the first 2 days after the presentation to detect an eventual cardiac source of emboli in selected cases (atrial fibrillation, etc.) were done during the first 2 days after the presentation.

7 – Cranial computed tomography (CT) scanning was performed at the presentation in all AION patients in order to identify a concomitant stroke.

8 – CT-angiography (CT-A) was done at the presentation and after EDS, only in selected cases (it allowed the assessment of the arterial wall and the endoluminal part of the aorta and its branches in selected cases of ipsilateral severe ICA stenosis/occlusion).

9 – a laboratory workup, including ESR, CRP, factor V Leiden, glycemia, etc., was done during the first 2 days after the presentation.

10 – a temporal artery biopsy (TAB) was selectively done when GCA was suspected following the criteria of the American College of Rheumatology. Because of unilateral clinical ocular involvement in all cases, we took a biopsy from the ipsilateral TA representing 2.5 cm of the tender, swollen segment of the affected artery or from the TA's site targeted by the color coded Doppler (TAB was guided by EDS because of segmental/discontinuous TA's involvement in GCA: skip lesions). All TABs were performed on the second day after the presentation.

Data analysis

The evaluation of the duplex color coded Doppler of the orbital vessels quality in order to foresee the A-AION diagnostic was completed by the SPSS v.17 program by using the calculated RI for the clinically affected eye for all orbital arteries. Starting from the receiver operating characteristics (ROC) curve coordinates for each investigated artery, the best threshold value was identified in order to achieve the minimum cost of the test (maximum of the sensitivity – Se + specificity – Sp). Using Microsoft Excel, the classification quality assessment was performed for the following parameters: sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV).

Results

We found 12 patients with unilateral acute A-AION, all of them with biopsy-confirmed disease (TAB positive), and 50 patients with unilateral acute NA-AION (no TAB).

We obtained the two groups of AION patients, using both clinical features, laboratory data, and results of ultrasound exams.

The comparison of major features of unilateral acute A-AION and NA-AION patients are presented in Table 1.

Table 1

Comparison of major features of arteritic anterior ischemic optic neuropathies (A-AION) and non-arteritic anterior ischemic optic neuropathies (NA-AION) patients

Feature	A-AION	NA-AION
Age (years), mean \pm SD	72.3 \pm 7.5	59.2 \pm 11.9
Sex ratio	Female > male (8 : 4 patients)	Female = male (26 : 24 patients)
Associated symptoms	New temporal headache, scalp tenderness, abnormal TAs on palpation, jaw claudication (11 patients)	Pain occasionally noted (3 patients)
	Amaurosis fugax (4 patients)	No amaurosis fugax
Visual acuity	83.3% (10 patients) < 20/200	60% (30 patients) > 20/200
Optic disc	Pale > hyperemic edema (11 patients)	Hyperemic > pale edema (48 patients)
	Cup normal (all 12 patients)	Cup small (38 patients)
ESR (mm/h)	> 50 (10 patients)	< 50 (47 patients)
CRP (mg/L)	> 5 (11 patients)	< 5 (48 patients)
Temporal artery biopsy (TAB)	Granulomatous inflammation of the media layer (all 12 patients)	No TAB
Color coded Duplex sonography of the retrobulbar (orbital) vessels	Severe diminished blood flow velocities in the PCAs, especially on the affected side, and high RI in all retrobulbar vessels, in both orbits (all 12 patients)	Blood flow velocities and RI in PCAs were preserved (all 50 patients)
Fluorescein fundus angiography	Disc and choroid filling delay (all 12 patients)	Disc filling delay (all 50 patients)
Treatment	Corticosteroids (all 12 patients)	None proved (all 50 patients)

TAs – temporal arteries; ESR – erythrocyte sedimentation rate; CRP – C-reactive protein; PCAs – posterior ciliary arteries; RI – resistance index.

All 12 A-AION patients presented with GCA. The NA-AION patients presented with: a) systemic associations: 20 (40%) patients with arterial hypertension, 13 (26%) patients with diabetes mellitus, 10 (20%) patients with nocturnal arterial hypotension, 4 (8%) patients with ipsilateral ICA's disease, and 3 (6%) patients with neoplasms, and/or; b). ocular and ONH risk factors: 38 (76%) patients with small cup in the optic disc and 2 (4%) patients optic disc drusen. On the other hand, all 12 A-AION patients presented large cups. Anterior segment examination of both eyes was normal in all 62 patients. Amaurosis fugax was an important early visual symptom in 4 (33.3%) A-AION patients and preceded permanent visual loss. The 8 other A-AION cases developed permanent visual loss without any warning. However, amaurosis fugax was never found in NA-AION patients.

Visual fields

The most common visual field defect in NA-AIONs cases was an inferior nasal sectorial defect found in 32 (64%) patients which was relative or absolute. The next most common visual field defect was relative or absolute inferior altitudinal in 12 (24%) patients.

Color coded duplex Doppler of the retrobulbar (orbital) vessels features

1) Spectral Doppler analysis of the retrobulbar vessels in 12 A-AION patients: in the first week of evolution found an undetectable or severe diminished of blood flow velocities in the PCAs on the affected side (orbit), with an increased RI in all other retrobulbar vessels in both orbits (including the PCAs in the contralateral orbit) (Figure 1A and B), despite the treatment with high-dose corticosteroids was found.

In month one, CDI examinations of orbital blood vessels revealed practically the same aspects.

2) Spectral Doppler analysis of the retrobulbar vessels in 50 NA-AION patients: by contrast, in all NA-AIONs cases, blood flow velocities and RI in PCAs were generally preserved. In the first week of evolution, this analysis revealed only a slight decrease of PSV in PCAs (nasal and temporal) in the affected eye, compared to the unaffected eye and a very slight decrease of PSV in CRA of the affected eye due to papillary edema. In OAs, PSV were variable – normal to decreased, according to ipsilateral ICA's status. In 4 patients severe ICA stenosis (>70% of vessel diameter) com-

bined with an insufficient Willis polygon led to decreased PSVs in the ipsilateral OAs.

In month 1, CDI examinations of orbital vessels revealed that blood flow normalization was reached. The only exceptions were the 4 cases with ipsilateral severe ICAs stenosis.

Analysis of the data (Table 2) revealed that a threshold value of 0.71 for the IR of the temporal PCAs in the clinically affected eye, provided the best combination of Se (86%), Sp (96%), PPV (88%) and NPV (96%), respectively, for the detection of A-AION. A threshold value of 0.68 for the IR of the nasal PCAs in the clinically affected eye, provided the best combination of Se (86%), Sp (93%), PPV (76%), and NPV (96%), respectively, for the detection of A-AION.

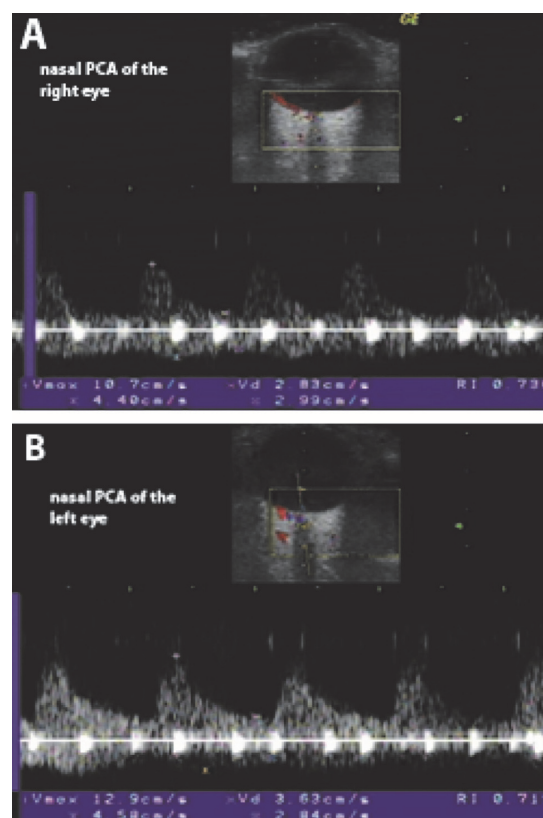


Fig. 1 – Color Doppler imaging (CDI) of the posterior ciliary arteries (PCAs) in arteritic anterior ischemic optic neuropathies (A-AION): decreased blood flow velocities (especially enddiastolic velocity – EDV) in the nasal PCAs: A) of the clinically affected right eye, and B) of the clinically unaffected left eye.

Table 2

The threshold values of resistance index (RI) in the orbital vessels and the corresponding values of sensitivity (Se), specificity (Sn), positive predictive value (PPV) and negative predictive value (NPV)

Arteries	CRA	PCA t	PCA n	OA
Cut- off point	0.67	0.71	0.68	0.81
Se	0.76	0.86	0.86	1
Sp	0.81	0.96	0.93	0.96
PPV	0.51	0.88	0.76	0.89
NPV	0.92	0.96	0.96	1

CRA – central retinal artery; PCA t – temporalu posterior ciliar artery; PCA n – nasal posterior ciliar artery; OA – ophthalmic artery.

Extracranial Duplex sonography (EDS)

1) EDS in all 12 A-AION patients: EDS demonstrated segmental inflammation of TAs in 12 patients with A-AION. At the presentation, we identified: a) "dark halo" sign in 11 patients (Figure 2); b) stenoses in 5 cases, and c) acute occlusions in 2 patients. Similar ultrasound patterns were discovered in other branches of the ECAs, including the facial, internal maxillary and lingual arteries. In week 2 and in month 1, the "halo" revealed by TAs ultrasound disappeared, because of the treatment with corticosteroids.

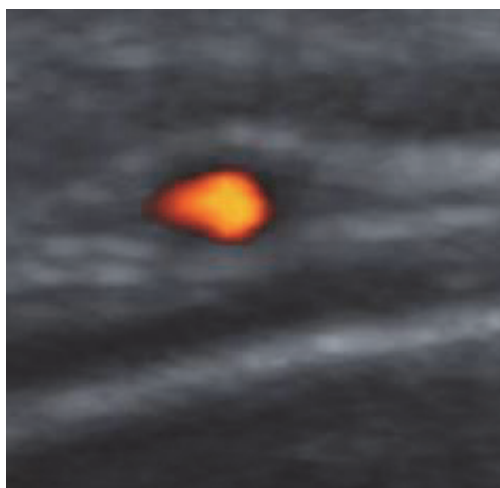


Fig. 2 – Extracranial duplex sonography (EDS) of the temporal ramus of the right temporal artery (TA) – "dark halo" sign.

2) EDS in 50 NA-AION patients: at the presentation we identified 4 cases with ipsilateral severe ICA stenosis and consecutive NA-AION;

CT and CT-A

CT-scanning excluded strokes in all 62 patients. CT-A done in 4 patients confirmed severe ICA stenosis and consecutive ipsilateral NA-AION in their cases.

ECG and TTE

These examinations excluded an eventual cardiac embolic source of NA-AION.

Treatment and evolution of A-AION patients

In all 12 GCA patients with A-AION, the treatment was initiated before TAB with intravenous methyl-prednisolone, 1g/day for 3 consecutive days after the presentation, followed by oral prednisone 60 mg daily for one month. The daily dose was then reduced by 5 mg weekly in the next month of follow-up. All 12 patients with A-AION presented stationary ophthalmologic evolution in 2 months (unilateral visual loss) without classic clinical symptoms of GCA (temporal headache, etc.) and any systemic manifestations (fever, malaise, etc.).

Discussion

In the patients with A-AION due to GCA, transient visual loss caused by optic-nerve or choroidal ischemia (amaurosis fugax) often precedes permanent visual loss by days to weeks (like in 4 of our 12 cases with A-AION). This symptom is unusual in NA-AIONs cases^{1, 3-7, 10-14} and we did not come across them in our NA-AION patients.

Biousse and Newman¹ mentioned that the degree of visual loss is often more severe in A-AION than in NA-AION. In one study, 54% of the patients with A-AION had an initial visual loss degree ranging from counting fingers to no light perception, as compared to 26% in the NA-AION group⁴. This result shows that acute, painless, severe permanent loss of vision is extremely suggestive of A-AION, as in 10 out of 12 our cases with A-AION. Different authors noted that once an untreated patient with GCA lost vision in one eye, the risk of GCA – related visual loss in the second eye is highest in the following hours to weeks (in at least 50% of cases)^{1, 15-18}; we did not come across with this situation in our A-AION patients because they were treated with high doses of corticosteroids.

We noted that inferior nasal field defect was the most common diagnostic visual field defect in our NA-AION patients, such as it was found reported in literature^{3-7, 10}.

According to Biousse and Newman¹, NA-AION is manifested as isolated, sudden, painless, monocular vision loss with edema of the optic disc. Ophthalmoscopy indicated that optic disc edema was associated more frequently with hyperemia in our NA-AION patients and with pallor (a chalky white color) in our A-AION patients, like in other studies^{3-7, 10-14}.

We found a small, crowded ONH with a small physiological cup in 38 of our 50 patients with NA-AION. The small cup-to-disc ratio defines a "disc at risk"^{1, 9}. Although this is difficult to observe during the acute phase of NA-AION, when the optic disc is swollen, examination of the clinically normal fellow eye should show a "disc at risk"¹. According to different authors, the absence of a crowded optic disc in the second eye of a patient with AION should increase the probability of A-AION^{1, 9}; we found only large cups in our 12 A-AION patients.

A-AION results from entire PCAs trunk vasculitis and the consecutive ONH infarction. Human autopsy studies of acute A-AION demonstrated ischemic necrosis of the ONH and infiltration of the PCAs by chronic inflammatory cells. In some of the cases reported in these studies, segments of PCAs were occluded by inflammatory thickening and thrombi³⁻⁷. Severe diminished blood flow velocities in the PCAs, especially on the affected side, and high RI in all retrolbulbar vessels in both orbits, represented characteristic signs of the CDI of the orbital vessels in A-AION in our study^{10-14, 19-21}. In NA-AION, blood velocities and RI in PCAs were preserved. Similar results were obtained in other studies^{10-14, 19-21}. The small number of A-AION cases could influence the calculated values (both the threshold values and the classifier quality). In spite of this, high PPV and NPV values in cases of temporal and nasal PCAs of the clinically affected eye, suggest that color coded duplex Doppler of orbital vessels may be a valid tool in the diagnosis of A-AION¹⁰.

Extremely delayed or absent filling of the choroid, which was identified in all our 12 A-AION fluorescein-angiograms, was suggested in other studies as a fluorescein-angiograms characteristic of A-AION^{3-7, 10-14}.

In our study, Duplex Doppler of retrobulbar vessels and fluorescein angiogram data supported the histopathological evidence from other studies³⁻⁷ of the involvement of the entire PCA trunk in A-AION (impaired both ONH and chorioidal perfusion in these patients)^{10-14, 19-21}. In contrast, in NA-AION cases, impaired flow to the ONH is distal to the PCA trunk, usually at the level of the paraoptic branches^{10-14, 19-21}. These branches directly supply the ONH with only one-third of the flow from the PCAs (impaired optic disc perfusion, with relatively preserved chorioidal perfusion in NA-AION patients)^{10-14, 19-21}.

While color coded Doppler sonography of orbital blood vessels does not eliminate the need for intravenous fluorescein angiography, it does, however, enhance the precision and reliability of the diagnostic evaluation for these patients, because it accurately, reproducibly and safely assesses the vascular supply of the optic nerve and retina¹⁰.

There were certain cases in our study where the differential diagnosis between A-AION and NA-AION was difficult: a) three patients with NA-AION had high ESR levels due to an associated neoplasm; b) two patients with GCA had a normal ESR; c) one case with GCA without systemic/clinical symptoms, even without a swollen TA (occult GCA)⁷.

Biousse and Newman¹ asserted that systemic symptoms of GCA (malaise, fever, temporal headache) may precede visual loss by months; however, about 25% of patients with positive TAB for GCA presented isolated A-AION without any systemic symptoms (so-called occult GCA)^{7, 16-18, 22-24}.

We believe that when duplex Doppler does not show evidence for A-AION, the patient should not receive high dose of corticosteroids until a TAB is performed, even if the ESR is elevated. On the other hand, patients with clinical evidence of A-AION, who have typical signs on Doppler of retrobulbar vessels, should be treated with corticosteroids before TAB in order to protect the fellow eye from going blind^{10, 16-19, 23-25}.

Ultrasonography of the TAs in temporal arteritis is very important for the GCA diagnosis^{15, 16}. Schmidt et al.²⁶ and Schmidt²⁷ asserted that the most specific (almost 100% Sp) and sensitive (73% Se) sign for GCA was a concentric hypoechogenic mural thickening "halo" which was interpreted as vessel wall edema. Other positive findings for GCA are the presence of occlusion and stenoses. We detected these signs in 11 out of 12 our patients in our A-AION group with GCA. The 12th patient presented an occult GCA. Ultrasound investigation of the TAs needs to be performed before corticosteroid treatment or within the first 7 days of treatment because the "halo" revealed by TAs ultrasound disappears within 2 weeks of cortico-therapy, like in our 11 GCA cases with temporal arteritis^{7, 22, 26, 27}. Schmidt et al.²⁶ and Schmidt²⁷ compared the results of TAs ultrasound examinations with the occurrence of visual ischemic complications in 222 consecutive patients with newly diagnosed, active GCA. However, findings of the TAs ultrasound examinations did not correlate with eye complications. For this reason, color coded duplex sonography of the retrobulbar vessels is of critical importance to identify A-AION^{10, 14}.

Conclusion

A history of transient visual loss (amaurosis fugax) associated with an acute, painless, and severe visual loss of the involved eye, with concomitant diffuse pale optic disc edema were characteristics of A-AION. On the other hand, none of these symptoms and signs were found in the NA-AION patients. The presence of a disc at risk in the fellow eye in a patient with unilateral AION increased the probability of NA-AION. Color coded duplex sonography of the orbital vessels identified the alterations in orbital blood flow, especially in the PCAs, which corresponded with the clinical features of A-AION, and enabled prompt differentiation between NA-AION and A-AION.

Conflict of interests

The authors declare no conflict of interest.

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