GIANT CELL ARTERITIS WITH EYE INVOLVEMENT – COLOR DOPPLER IMAGING (CDI) OF RETROBULBAR VESSELS FINDINGS

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ABSTRACT

Background Giant cell arteritis is a primary vasculitis, that affects medium-sized arteries, especially branches of the ECA.

Purpose The main objective was to assess the role of CDI of retrobulbar vessels in the study of two patients with Horton's disease with eye involvement.

Methods We have used a sonographer with 8-15 MHz linear probe for CDI of orbital vessels.

Results Both patients presented malaise, temporal headache, tender temporal arteries and laboratory signs of inflammation. The first patient had a left central retinal artery obstruction, and the second one had a left ischaemic optic neuropathy. They presented a painless, severe loss of vision of the left eye. Temporal artery histology was positive in both cases. Ultrasound investigation was performed before corticosteroid treatement. CDI of retrobulbar vessels detected low blood velocities, especially end-diastolic velocities and high resistance index in all retrobulbar vessels, in both orbits, for both patients (especially on the affected side). Typical sonographic features in temporal arteritis were „dark halo sign" (vessel wall thickening), associated with stenoses or occlusions of branches of ECA. Interestingly, in both cases, the CCA and the ICA were, also, affected.

Conclusions Ultrasound technique is a valuable diagnostic tool to investigate giant cell arteritis.

Key words: Giant cell arteritis, temporal headache, central retinal artery obstruction, ischaemic optic neuropathy.

1. INTRODUCTION AND PURPOSE

Giant cell arteritis is a primary vasculitis, that affects medium-sized and large arteries, especially branches of the external carotid artery (ECA). The diagnosis of Horton's disease requires age more than 50 years at 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/hr, and typical histologic findings in temporal artery biopsy. 40-50% of these patients have ophthalmologic complications, consisting in visual loss (secondary to ischemic optic neuropathy, or central retinal artery occlusion) or homonymous hemianopsia or cortical blindness (unilateral or bilateral occipital infarction).(3-6)

The main purpose of our study was to assess the role of Color Doppler Imaging (CDI) of retrobulbar vessels in the study of two patients with temporal arteritis with eyes involvement.

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2. METHODS

We have used a sonographer (Logic 500, General Electrics) with 8-15 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 ophthalmic artery-OA; the central retinal artery-CRA and vein-CRV, posterior ciliary arteries-PCA, and the superior ophthalmic vein-SOV).The assessement of extracranial arteries was performed with a sonographer (My Lab 50, Esaote Italy) with a 7.5-Mhz linear-array transducer, combining B mode and Color-Doppler/ pulsed-wave Doppler ultrasound. (7-10)

3. RESULTS

3.1. Clinical features

A 62-year-old man (MM), and a 72-year-old woman (CE) presented temporal headache, especially common at night, scalp tenderness, with swollen, pulseless temporal arteries, and jaw claudication, for the 2 weeks prior to admission. Systemic symptoms included fever, fatigue, and malaise. They had not arthralgias or psychiatric symptoms. They didn't presented transient ischemic attacks, or stroke. (3-6)

3.2. Laboratory findings

Laboratory findings revealed in both cases increased ESR (70 mm/hr, respectively 60 mm/hr), and an elevated C-reactive protein: CRP (28 mg/L, and 52 mg/L, respectively).(3-6)

3.3. Extracranial Duplex sonography

Ultrasound investigation was performed before corticosteroid treatement. Extracranial Duplex sonography investigated almost completely the whole length of the common superficial temporal arteries, including the frontal and parietal branches, and found that inflammation was segmental (discontinous arterial involvement).

Three findings were important for the ultrasound diagnosis of temporal arteritis: (7-9)

a) „dark halo" sign – a typically hypoechoic, circumferential wall thickening around the lumen of an inflamed temporal artery – which represents vessel wall edema. (Figure 1)

b) stenosis were documented by blood-flow velocities, which were more than twice the rate recorded in the area of stenosis compared with the area before the stenosis, with wave forms demonstrating turbulence and reduced velocities behind the area of stenosis. (Figure 2. a, b)

c) acute occlusions, in which the ultrasound image was similar to that of acute embolism in other vessels, showing hypoechoic material in the former artery lumen with absence of color signals.

Similar ultrasound patterns were found in other arteries, such were the facial, the internal maxilary, the lingual, the distal subclavian, and axilary arteries. (Figure 3)

Interestingly, in both cases, the common carotid and the internal carotid arteries were, also, involved. (Figure 4, Figure 5) Both patients presented large-vessel giant cell arteritis. (8)

Wall swelling, stenoses, or occlusions of larger arteries remained for months, despite corticosteroid therapy, in both cases.(7-9)

3.4. The histopathologic picture

The temporal arteries biopsy was guided by the Doppler Ultrasonography, because of discontinuous arterial involvement in Horton's disease ("skip lesions").We observed intimal thickening, internal limiting lamina fragmentation, and chronic inflammatory infiltrate with giant cells.(3-6) (Figure 6. a, b)

3.5. Computed Tomography (CT) and CT-Angiography (CT-A)

Cerebral CT on the third day of admission were normal in both cases. CT-A in the first case, (patient MM) confirmed the occlusion of the left CCA, ECA, and ICA. (Figure 7)

3.6. Ophthalmologic complications and Color Doppler Imaging (CDI) of retrobulbar vessels

Case 1 (patient MM):

A) Patient MM presented a central retinal artery (CRA) occlusion of the left eye,

with the following key features: (2)

1. abrupt, painless, severe loss of vision of the left eye (0.1).
2. anterior segment examination was normal in both eyes.
3. the fundus of the left eye presented: (Figure 8)

- ischemic whitening of the retina.
- cherry-red spot in the center of the retina.
- the site of obstruction of CRA was not visible on ophthalmoscopy (no embolus was found).

B) Patient MM presented at the Spectral Doppler analysis of the retrobulbar vessels the following aspects: (10)

1. an increased resistance index (RI) in the central retinal arteries-CRA (the RI was higher

on the left affected side, than it was on the right unaffected side); with severe diminished blood flow velocities (especially end-diastolic velocities) in the CRA. (Figure 9. a, b)
2. less abnormalities were observed in the posterior ciliary arteries (PCA), (Figure 10. a, b) and in the ophthalmic arteries. (Figure 11. a, b)

Case 2 (patient TL):

A) Patient TL presented an anterior ischemic optic neuropaty of the left eye, with the following key features: (1)

1. visual acuity was 0.02 for the left eye, and 0.4 for the right eye.
2. anterior segment examination of both eyes was normal.
3. the ophthalmoscopy of the left eye revealed diffuse optic disc edema. (Figure 12)

B) Patient TL presented at the Spectral Doppler analysis of the retrobulbar vessels the following aspects: (10)

1. an increased resistance index (RI) in the posterior ciliary arteries (PCA) (the RI was higher on the left affected side, than it was on the right unaffected side); with severe diminished blood flow velocities (especially end-diastolic velocities) in the PCA. (Figure 13. a, b, c, d)
2. less abnormalities were observed in the central retinal arteries (CRA), (Figure 14. a, b) and in the ophthalmic arteries (OA). (Figure 15. a, b)

4. DISCUSSIONS

Giant cell arteritis is an autoimmune vasculitis of unknown origine. The disease mainly affects patients older than 50 years of age. Horton's disease involves predominantly extracranial vessels, especially superior temporal arteries, the ophthalmic arteries, the posterior cilliary arteries, and other peripheral arteries. The typical predominantly extracranial vascular involvement is explained by the affinity of inflammation to the elastic fibers. As intracranial arteries have less elastic fibers in the media, they are seldom involved. A definitive diagnosis is made following the criteria of the American College of Rheumatology which includes: age, temporal headache, swollen temporal arteries, jaw claudication, eye involvement, the temporal artery biopsy, and its histologic evaluation. Because of segmental (discontinous) involvement of temporal arteries, a biopsy result may be negative in 9-44% of patients with clinical positive signs of temporal arteritis. (3-6) Our patients accomplished all criterias of the American College of Rheumatology.

Both patients presented large vessel giant cell arteritis, which is a subgroup of temporal arteritis in at least 17% of cases. In these patients inflammation occurs also at the level of the subclavian, the axillary arteries, etc.(3-6) Interestingly, in both our cases, the common carotid and the internal carotid arteries were, also, involved.

Our first patient presented a central retinal artery obstruction (CRAO), which is the result of an abrupt diminuation of blood flow in CRA, severe enough to cause ischemia of the inner retina.. Because of the fact that there are no functional anastomoses between choroidal (PCA) and retinal circulation (CRA), CRAO determines severe and permanent loss of vision. Therefore, it is very important to identify the cause of CRAO, in order to protect the other eye. The majority of CRAO are thrombotic or embolic. The site of the blockage is within the optic nerve substance and for this reason, it is generally not visible on ophthalmoscopy. All patients with CRAO should undergo a systemic evaluation, because of strong association with systemic diseases: carotid artery diseases, especially atherosclerosis, aortic or mitral valvular lesions, haematological disorders, and last but not least systemic vasculitis (including Horton's disease, like in our two cases). (3-6)

Our second patient suffered from an arteritic anterior ischemic optic neuropathy (AAION). AAION results from short posterior ciliary (SPCA) vasculitis and the consecutive optic nerve head infarction. Human autopsy studies of acute AAION demonstrate optic disc edema with ischemic necrosis of the prelaminar, laminar, and retrolaminar portions of the optic nerve and infiltration of the SPCA's by chronic inflammatory cells. In some cases segments of these vessels have been occluded by inflammatory thickening and thrombus. Typically, like in our second case, AAION is exhibited in elderly patients, with a mean age of 70 years, with severe visual loss, temporal headache, tender of temporal arteries, etc. (3-6)

5. CONCLUSIONS

1. The eye involvement in Horton's disease consists in anterior ischemic optic neuropaty or central retinal artery occlusion with abrupt, painless, severe loss of vision of the involved eye.
2. Color Doppler Imaging of the retrobulbar vessels may be helpful for detecting the blood

flow in the orbital vessels, especially in cases of opacity of the medium, or when the clinical appearance of ophthalmologic complications in temporal arteritis is athypical.

3. The Spectral Doppler Analysis of the orbital vessels in giant cell arteritis reveales low blood velocities, especially end-diastolic velocities, and high resistance index in all retrobulbar vessels, in both orbits, for both patients (especially on the affected side).
4. Ultrasound technique is a valuable diagnostic tool to investigate Horton's disease.

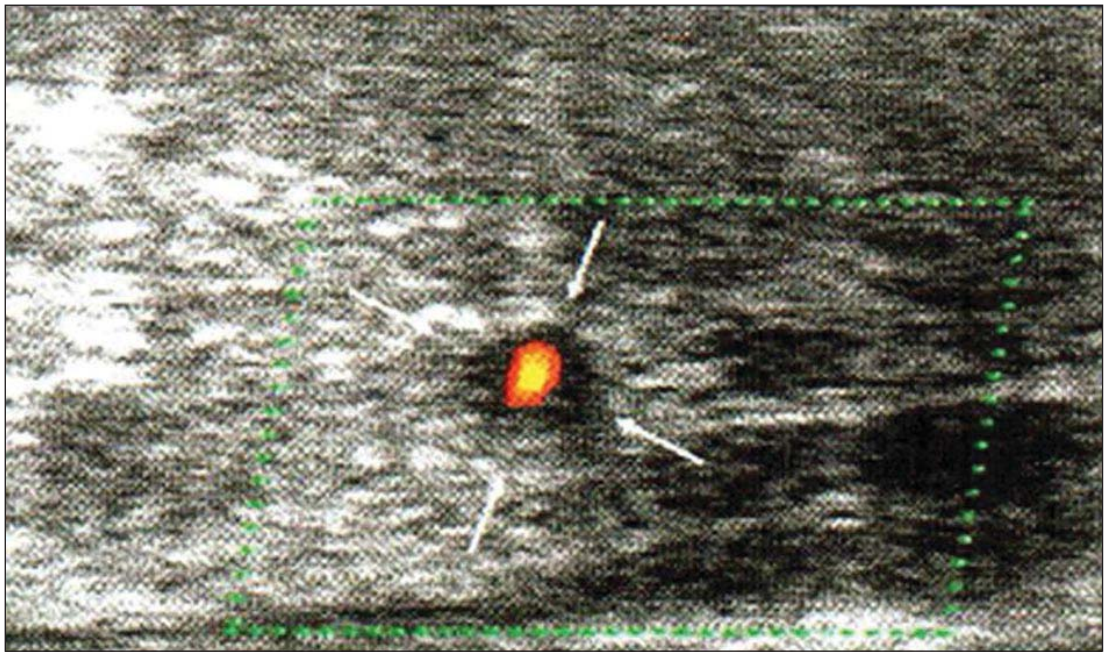


Figure 1. *Patient MM-Extracranial Duplex (linear transducer 11 Mhz). The right superior temporal artery (STeA) shows reduced color filling and a vessel wall thickening in form of a dark halo sign (arrows).*

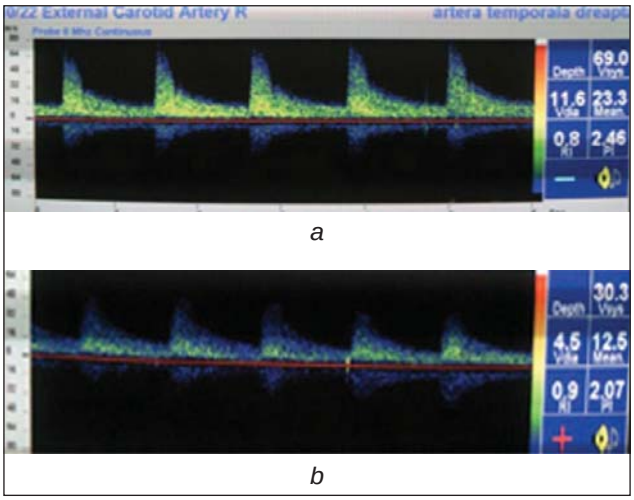


Figure 2. *Patient TL-Spectral analysis cw-Extracranial Doppler ultrasound of the left temporal artery. Peak flow velocity is 69 cm/s in the stenosis (a), and 30.3 cm/s behind the stenosis (b).*

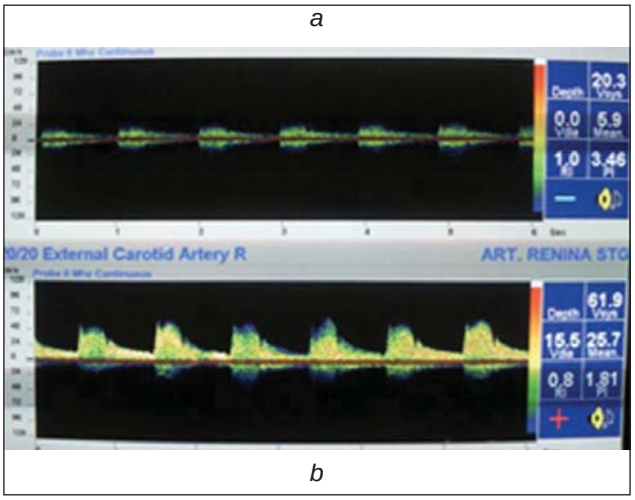


Figure 3. *Patient TL-Spectral analysis cw-Extracranial Doppler ultrasound of the left lingual artery. Peak flow velocity is 61.9 cm/s in the stenosis (A), and 20.3 cm/s behind the stenosis (B).*

Figure 4. *Patient MM-B mode insonation in large vessel giant cell arteritis Transverse view of the left CCA. Hypoechoic wall swelling*

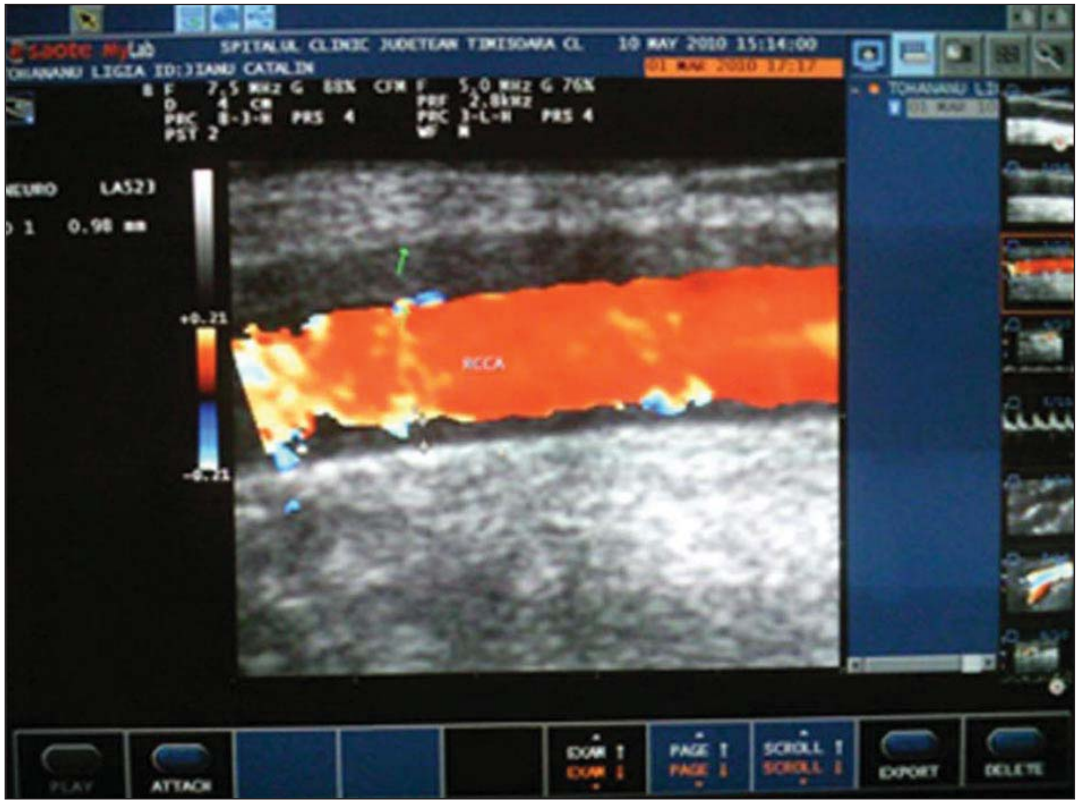


Figure 5. *Patient TL-Color Doppler ultrasound in large vessel giant cell arteritis Longitudinal view of the right CCA with hypoechoic wall swelling.*

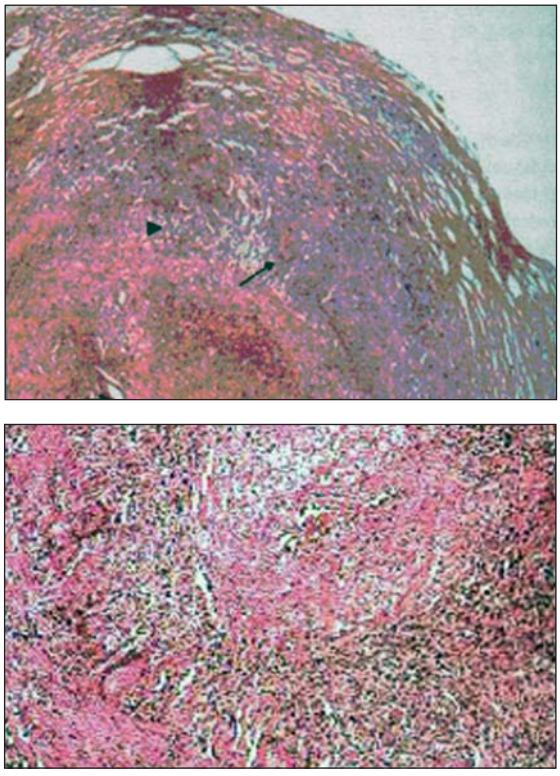


Figure 6. *Patient MM-Typical temporal arteritis. A-Histological section shows a vasculitis involving all coats of the temporal artery. B-Increased magnification shows the typical giant cell granulomatous inflammation.*

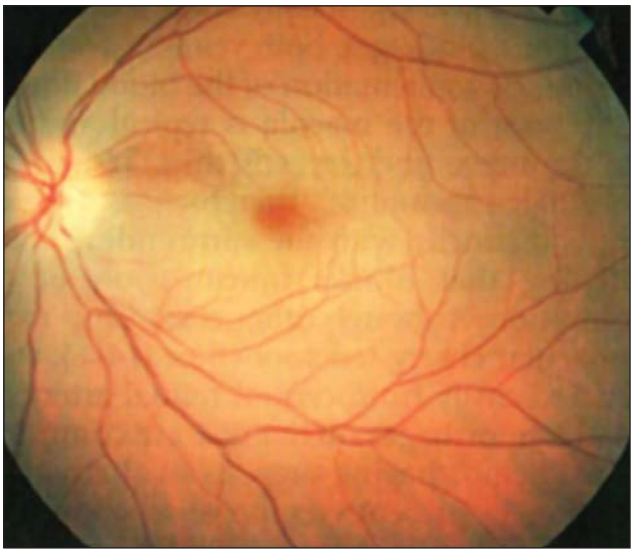


Figure 8. *Patient MM-Fundus view of the left eye. Central retinal artery obstruction. A prominent cherry-red spot with cilio-retinal artery sparing in the papillo-macular bundle.*

Figure 7. *Patient MM-CT-A. Occlusion of the left CCA, ECA, and ICA.*

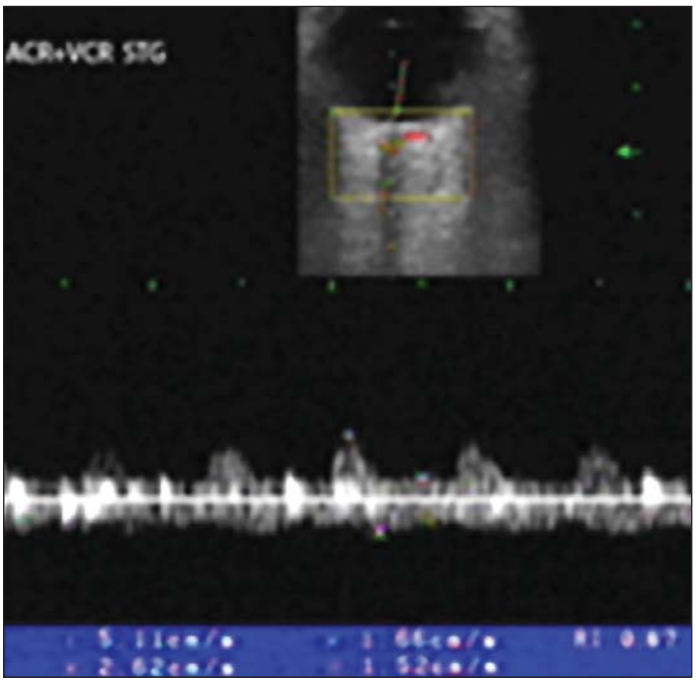


Figure 9. a. *Pacient MM-Spectral Doppler analysis of central retinal arteries (CRA).*

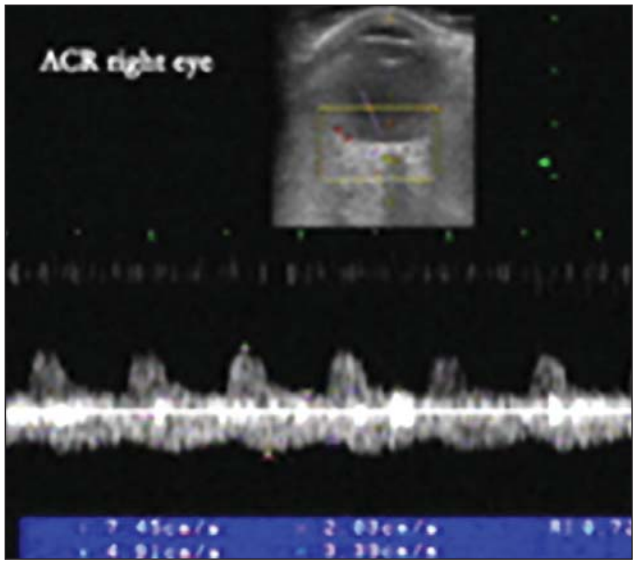
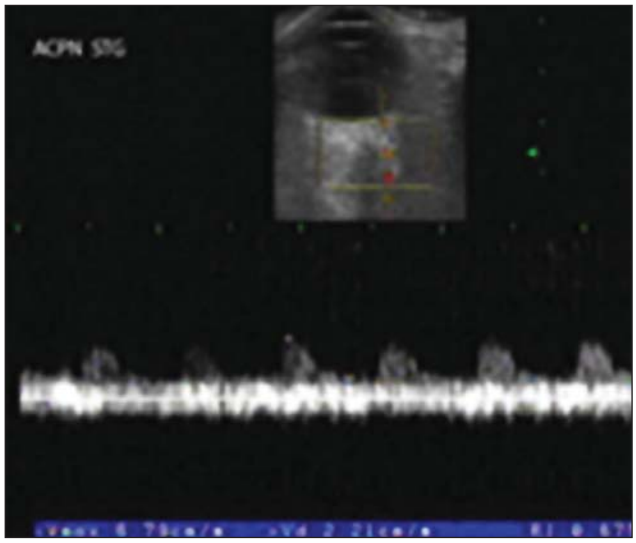


Figure 9. b. *Pacient MM-Spectral Doppler analysis of central retinal arteries (CRA).*



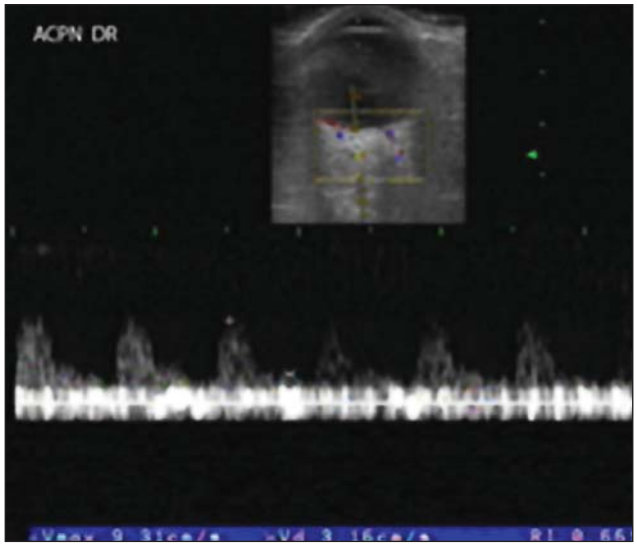
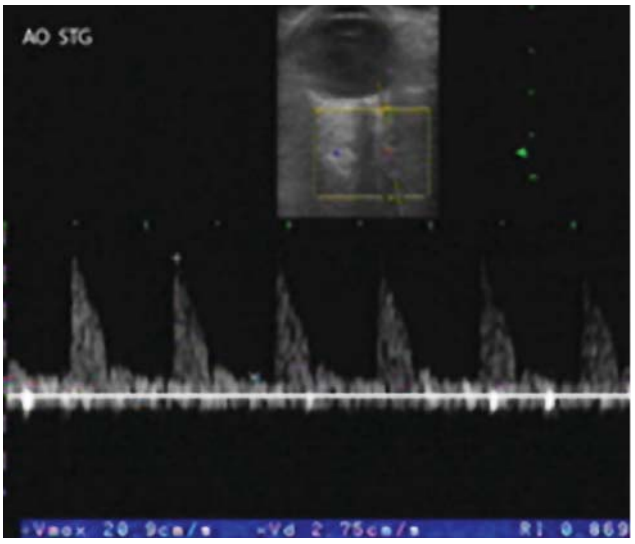


Figure 10. a, b. *Pacient MM-Spectral Doppler analysis of posterior ciliary arteries (PCA).*



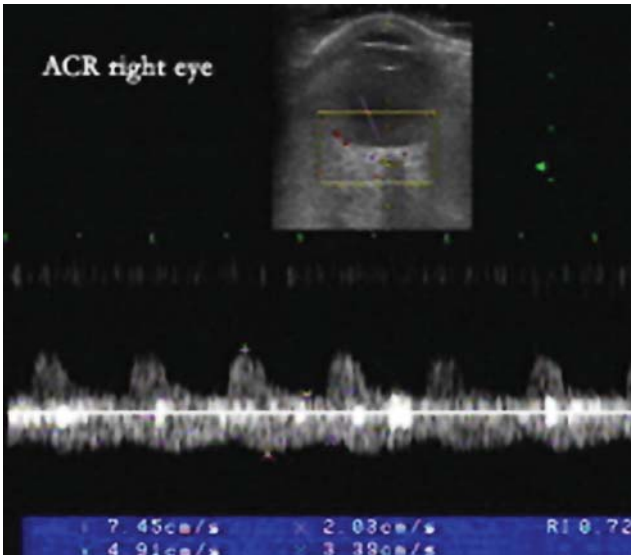
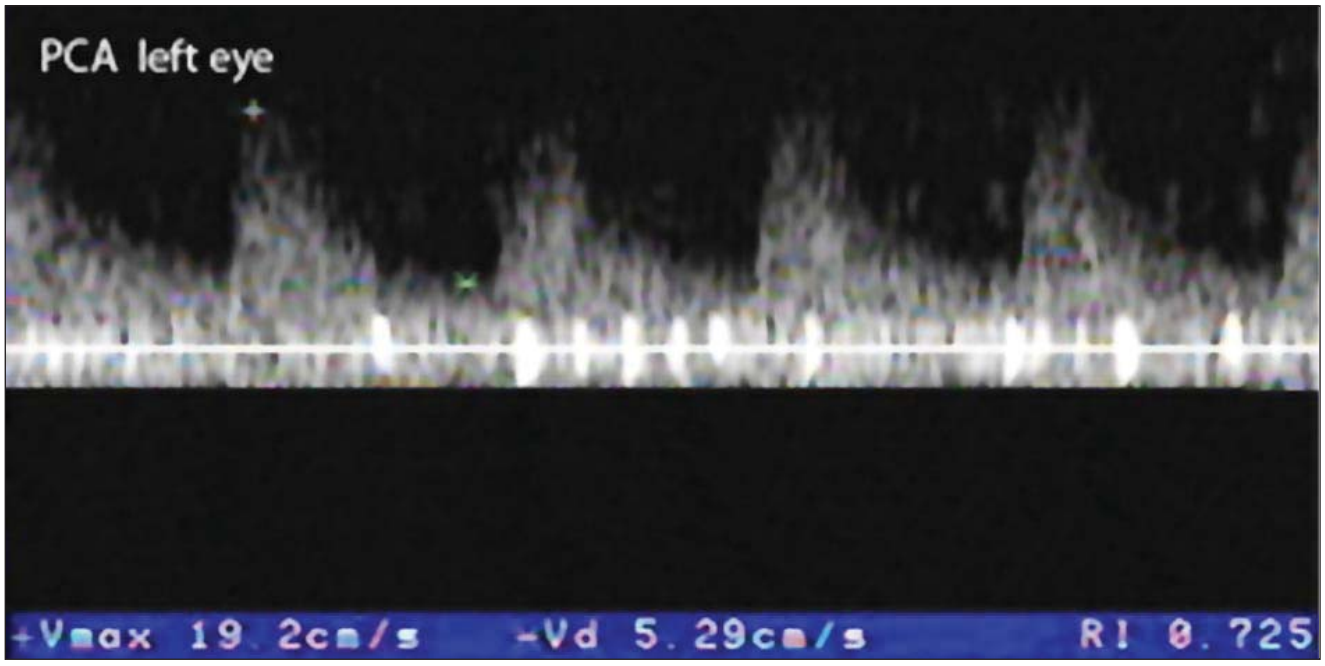


Figure 11. a, b. *Pacient MM-Spectral Doppler analysis of ophthalmic arteries (OA).*

Figure 12. *Pacient TL-Fundus view of the left eye. Arteritic anterior ischemic optic neuropaty (AAION). The optic disc demonstrates pale with diffuse edema.*



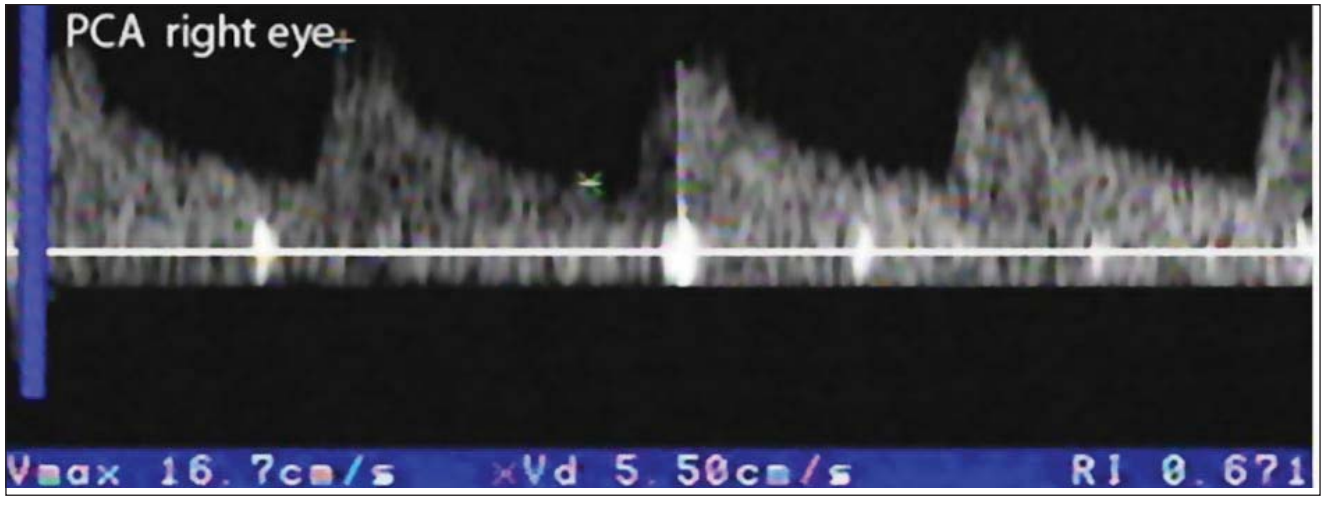


Figure 13. a, b. *Pacient TL-Spectral Doppler analysis of posterior ciliary arteries (PCA).*

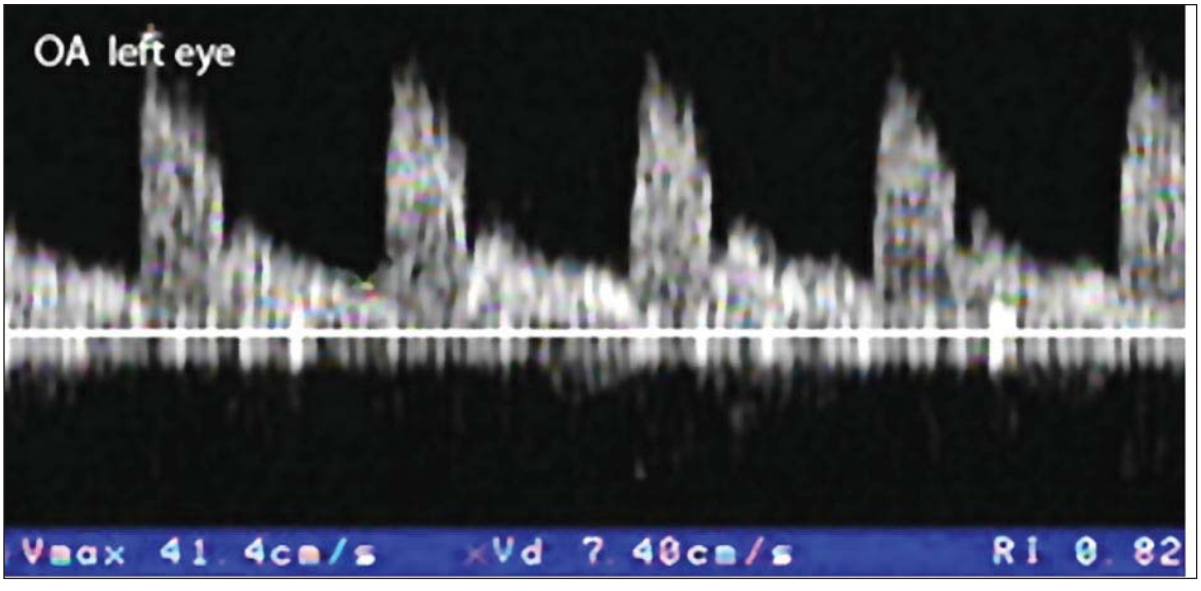


Figure 15. a, *Pacient TL-Spectral Doppler analysis of ophthalmic arteries (OA).*

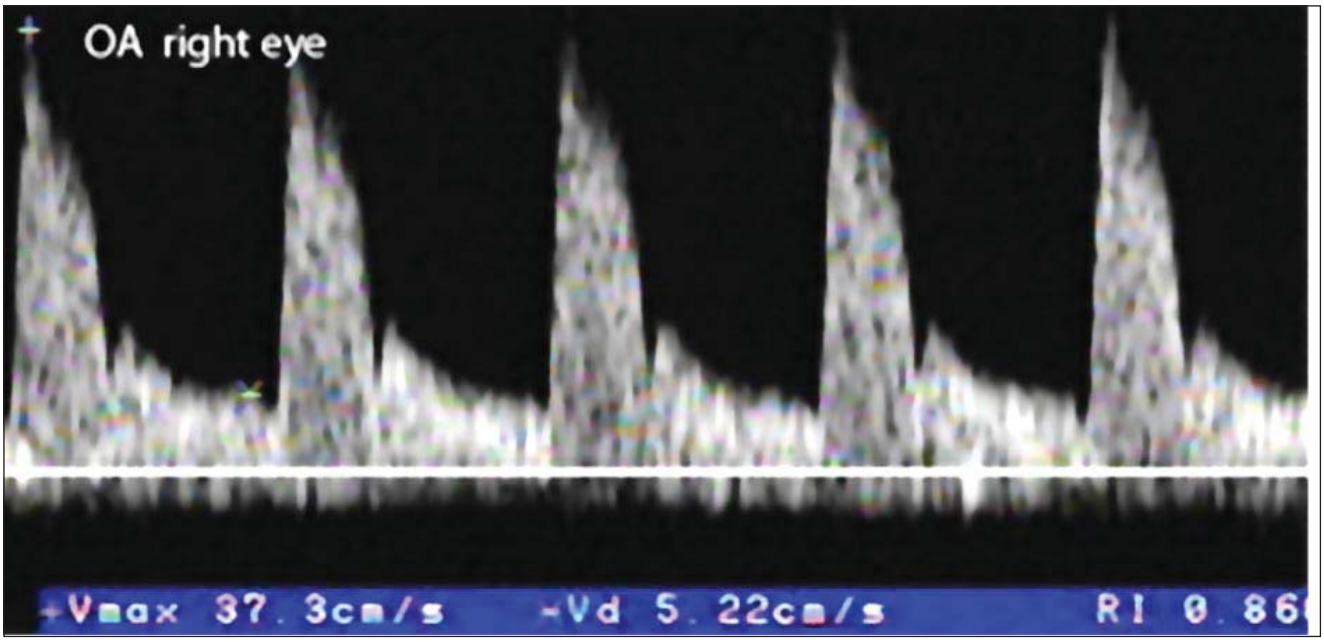


Figure 15. b *Pacient TL-Spectral Doppler analysis of ophthalmic arteries (OA).*

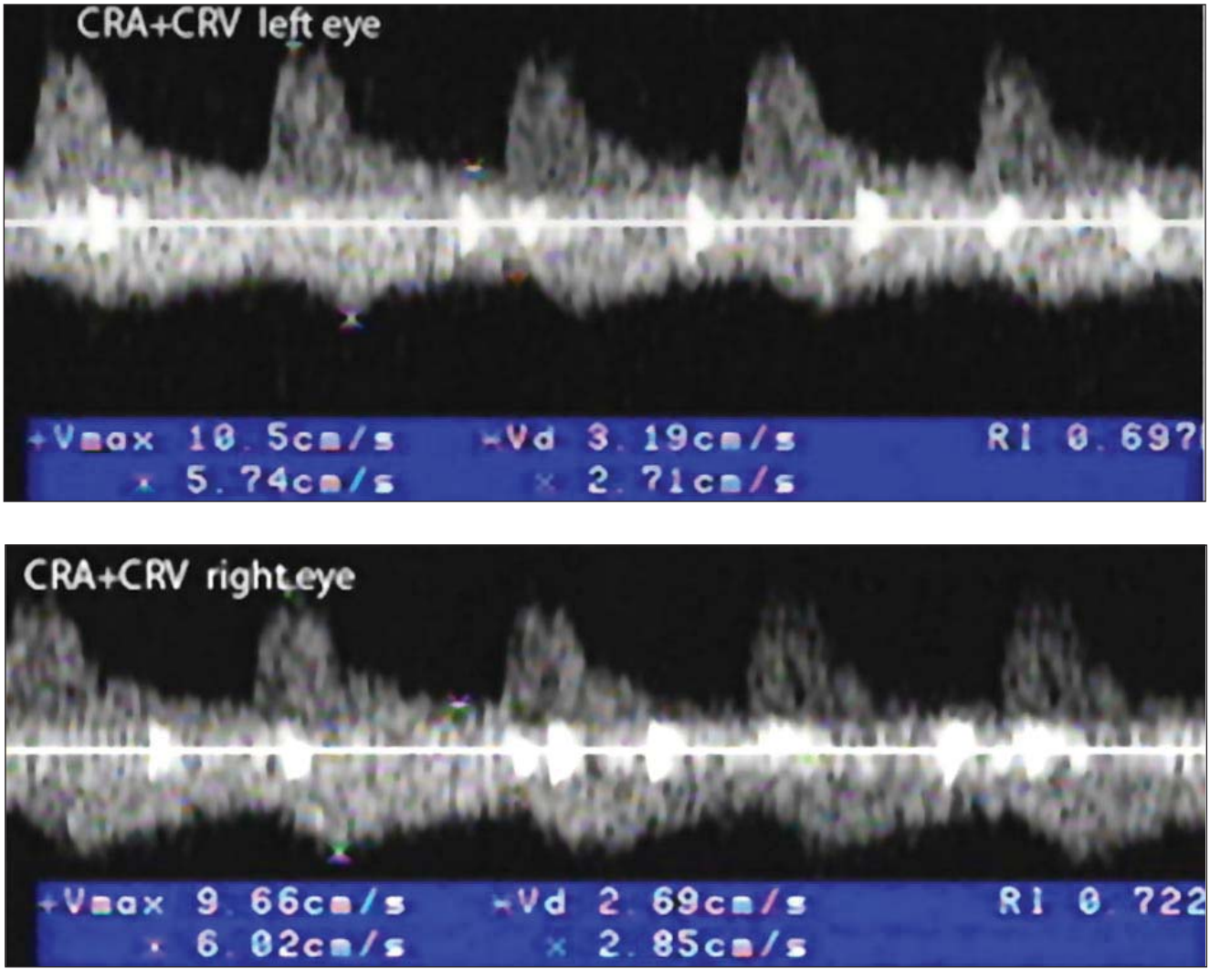


Figure 14. a, b. *Pacient TL-Spectral Doppler analysis of central retinal arteries (CRA).*

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