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The retrobulbar spot sign in sudden blindness — Sufficient to rule out vasculitis?

Michael Ertl[a,*,1], **Mathias Altmann**[b,1], **Elisabeth Torka**[a], **Horst Helbig**[b], **Ulrich Bogdahn**[a], **Anreea Gamulescu**[b], **Felix Schlachetzki**[a]

[a] *Department of Neurology, University of Regensburg, Bezirksklinikum Regensburg, Universitätsstraße 84, 93042 Regensburg, Germany*
[b] *Department of Opthalmology, University of Regensburg, Franz-Josef-Strauss-Allee 11, 93053 Regensburg, Germany*

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Summary
Introduction: Sudden retinal blindness is a common complication of temporal arteritis (TA). Another common cause is embolic occlusion of the central retinal artery (CRA). The aim of this prospective study was to examine the diagnostic value of hyperechoic material in the CRA for exclusion of vasculitis as a cause. The authors used orbital color-coded sonography (OCCS) for the detection of hyperechoic material.
Materials and methods: Twenty-four patients with sudden visual loss were included in the study after opthalmoscopic exclusion of other causes (e.g. vitreous bleeding, retinal detachment). Parallel to routine diagnostic workup OCCS was performed in all patients.
Results: 7 patients with the diagnosis of TA presented with different degrees of hypoperfusion in the CRA without hyperechoic material (referred to as a ''spot sign'') detected by OCCS.

Diagnostic workup in the remaining 17 patients did not reveal any signs of TA. The hyperechoic spot sign was visible in 10 of 12 patients (83%) with embolic CRA occlusion. Altogether the frequency of the spot sign in this group was 59%.

Detection of embolic CRAO using the spot sign had a sensitivity of 83% and a specificity of 100%. The missing spot sign in patients with TA was a highly specific finding (p-value 0.01).
Conclusions: The ''spot sign'' is a highly specific finding, and its detection excludes the diagnosis of temporal arteritis in patients with sudden blindness. The finding of a spot sign helps prevent patients from receiving long-term steroid treatment, or an invasive temporal artery biopsy, with its immanent risks.

Abbreviations: ACR, American College of Rheumatology; AFIB, Atrial fibrillation; AION, Anterior ischemic optic neuropathy; CRA, Central retinal artery; CRAO, Central retinal artery occlusion; DM 2, Diabetes mellitus type 2; ECST, European Carotid Surgery Trialists; ESR, Erythrocyte sedimentation rate; FA, Fluorescence angiography; ICA, Internal carotid artery; ION, Ischemic optic neuropathy; MI, Mechanical index; OCCS, Orbital color-coded sonography; OCT, Optic coherence tomography; PCA, Posterior ciliary artery; PION, Posterior optic neuropathy; TA, Temporal arteritis.

* Corresponding author. Tel.: +49 941 941 0; fax: +49 941 941 3015.
E-mail address: Michael.Ertl@medbo.de (M. Ertl).
[1] Contributed equally.

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Introduction

Sudden retinal blindness is a common complication of temporal arteritis (TA) due to ischemic optic neuropathy (ION) caused by vasculitic occlusion of the central retinal artery (CRA), the posterior ciliary artery (PCA) and other orbital arteries [1]. Depending on the affected arteries central retinal artery occlusion (CRAO), anterior optic neuropathy (AION) or posterior optic neuropathy (PION) are the results. In the elderly other common causes for hypoperfusion of the retina are thromboembolic events [2,3]. As a tool for the detection of TA, high-resolution ultrasonography of the superficial temporal artery has had a significant impact, with a high positive predictive value for the diagnosis of TA (specificity of 91%). However, a missing ''halo'' sign, suggestive for vessel wall inflammation seen on ultrasonography, does not sufficiently rule out presence of the disease (sensitivity 68%) and, therefore, superficial temporal artery biopsy remains the gold standard in the diagnosis of TA [4]. The differentiation of embolic versus arteritic occlusion remains a diagnostic challenge in elderly patients with ischemic optic neuropathy, because symptoms of TA, such as headache and elevation of inflammatory parameters, often coexist with significant cerebrovascular risk profiles. Additionally, depending on the cause of occlusion, different acute management strategies need to be applied quickly to improve long-term outcomes in these patients.

It is evident that we still need additional criteria with high negative predictive values to exclude the presence of vasculitis.

In a previously published series of patients with criteria for TA and sudden blindness, we found a hyperechoic embolic occlusion of the CRA in the area of the optic nerve head, which could be used to exclude TA; we called this a retrobulbar ''spot sign'' [5]. Foroozan et al. published a series of 29 patients with acute vision loss irrespective of the criteria for TA and observed this phenomenon in 9 patients with central retinal artery occlusion (CRAO) detected by retinal fluorescence angiography [6].

High-resolution color-coded ultrasonography can also be applied to the orbit since vitreous gel does not lead to any significant absorption of the incidental ultrasound beam. Orbital color-coded sonography (OCCS) allows detection of retrobulbar arteries and veins in addition to an assessment of orbital structures [7]. An analysis of Doppler flow spectra further aids the assessment and, to some degree the quantification, of retinal hypoperfusion due to CRA stenosis or occlusion. Normal flow velocity values within the CRA have been established previously [8].

This is the first prospective study in which patients suffering from acute vision loss due to either thromboembolic events or vasculitic changes in vessel walls were examined to identify the frequency of the ''spot sign'' in these specific disease patterns. We demonstrate that OCCS can be used to significantly discriminate embolic CRAO from arteritic causes of sudden ocular blindness in the elderly.

Materials and methods

Population and study protocol

The study protocol was approved by the local ethics committee at the University of Regensburg in accordance with the Declaration of Helsinki. Patients were first seen and screened at the Department of Ophthalmology of the University Hospital Regensburg. After exclusion of other reasons for visual loss, such as vitreous bleeding or retinal detachment, patients were referred to the Department of Neurology for OCCS and a routine neurovascular workup that included assessment of the superficial temporal artery. The funduscopic results were not disclosed before OCCS was performed. Before enrollment in the study, patients were made aware of the noninvasive and safe nature of OCCS and provided their written informed consent. In accordance with the study protocol, patients underwent routine diagnostic workups in the Departments of Ophthalmology and Neurology at our hospital, including registration of cerebrovascular risk factors, laboratory tests to detect criteria associated with TA (including the erythrocyte sedimentation rate [ESR]) according to American College of Rheumatology (ACR) criteria, a visual acuity test, retinal fundoscopy and color-coded sonography of brain-supplying arteries. All tests were performed within 24 h after admission.

Ultrasound equipment and data acquisition

For the visualization of retrobulbar structures, a high-resolution linear-array transducer with frequencies ranging from 8 to 15 MHz was used in combination with a Siemens Acuson system (Siemens AG, Erlangen, Germany) and a Toshiba XarioXG device (Toshiba, Tokyo, Japan). The acoustic output of the ultrasound systems was adjusted to the requirements of orbital sonography according to the ALARA principle (''as low as reasonably achievable'') to avoid damage to the lens and retina [9]. The settings for orbital sonography were the following: for B-mode, transmit frequency 14 MHz, mechanical index (MI) = 0.1, single focal zone at 2.5 cm, and bandwidth 74 dB; for C-mode, transmit frequency 10 MHz, MI = 0.2, color scale optimized for low velocities, and no wall filter; and for PW-mode, transmit frequency 2 MHz and MI < 0.44.

For OCCS the patients were placed supine with their eyes closed and asked to gaze forward. From above and slightly lateral, the transducer was placed with minimal pressure on the patient's orbit using plenty of contact gel. By definition the nasal side is depicted on the left image side.

Patient groups and statistical analysis

Depending on the final diagnosis and specific findings, patients were sorted into two different groups: (1) patients with a final diagnosis of TA; and (2) patients with visual loss on the basis of other pathologies. Patients were then further sorted depending on their funduscopic findings.

The frequency of the retrobulbar ''spot sign'' in patients with TA (group 1) was compared with that in patients