Case report

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Unilateral corneal arcus and conjunctival vessel alterations in cranial autonomic dysregulation: A case report

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Abstract

Background: Cranial autonomic dysregulation is a common symptom of patients suffering from cluster headache or migraine. The peripheral vascular dysfunction may increase the risk for ischemic or hemorrhagic strokes, myocardial infarction, retinal vasculopathy, cardiovascular mortality, and peripheral artery diseases. Furthermore, it may also manifest with ocular symptoms, e.g., increased lacrimation, conjunctival injection, and facial swelling.

Case presentation: We here report a case of a patient with migraine and ocular signs of a vascular dysregulation that have led to persisting changes of conjunctival vessels and to a corneal arcus.

Conclusions: Autonomic vascular dysregulation may not only cause headaches but also persisting changes of ocular tissues, e.g., conjunctival vessel alterations and a corneal arcus.

Keywords

Conjunctival hyperemia, conjunctival vessels, corneal arcus, cranial autonomic dysregulation, migraine

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Background

Cranial autonomic dysregulation is a common symptom of patients suffering from cluster headache or migraine.[1] The peripheral vascular dysfunction may increase the risk for ischemic or hemorrhagic strokes, myocardial infarction, retinal vasculopathy, cardiovascular mortality, and peripheral artery diseases.[2] Furthermore, it may also manifest with ocular symptoms, e.g., increased lacrimation, conjunctival injection, and facial swelling.[3]

We here report a case of a patient with migraine and ocular signs of such a vascular dysregulation that have led to persisting changes of conjunctival vessels and to a corneal arcus.

Case presentation

A 54-year-old woman was referred to our clinic due to recurrent unilateral eyelid and conjunctival swelling of her right eye(lid) for two decades and symptoms lasting each time for about 1–2 days. It used to be accompanied by headache but since few years, eye symptoms are also present without headache. Medical history revealed a known migraine with aura (mostly right-sided headache with photophobia, phonophobia and nausea, with symptoms lasting for half a day up to one or two days since youth) and an unremarkable brain MRI 15 years ago. Best-corrected visual acuity was 20/16 for both eyes, and intraocular pressure was normal and symmetric. In the ophthalmological examination during an attack, we found a

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swollen and ptotic upper eyelid on the right with conjunctival chemosis and hyperemia (Figure 1a). A unilateral corneal arcus on this eye was found (Figure 1b, arrow), whereas the cornea was clear on the left eye (Figure 1c). Even in the attack-free intervals, the slit lamp exam revealed a conjunctival hyperemia (Figure 1d-f) with vessel abnormalities (caliber changes) of the right eye (Figure 1g). Pupils were equal, round, and reactive to light, and fundoscopy was unremarkable. An MRI of the orbit with MR-angiography of the carotid artery showed periorbital swelling and enhancement on the right side. There were no signs of intraorbital vascular malformation,

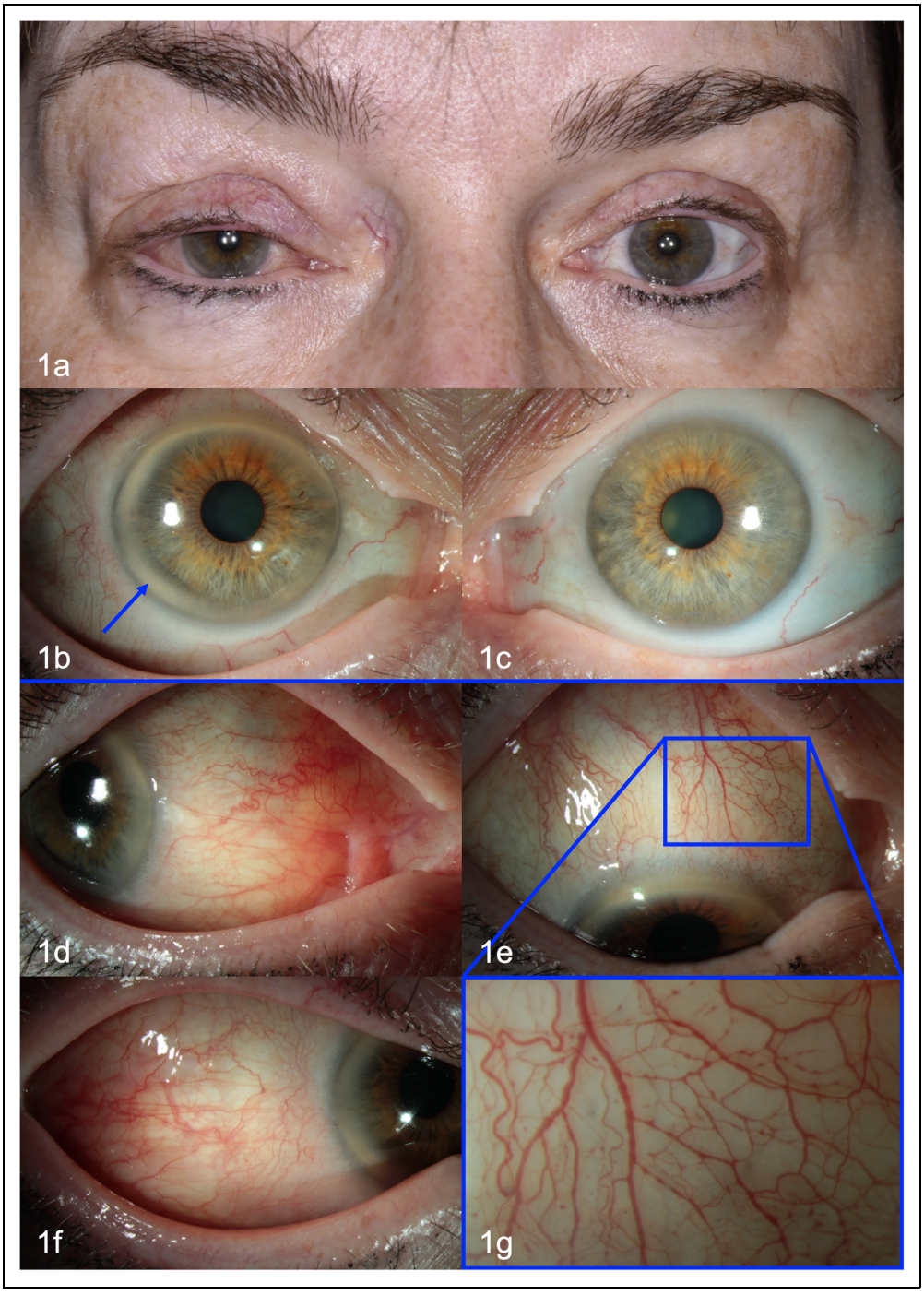

Figure 1. Patient with right sided ptosis, eyelid swelling, and increased conjunctival injection during a migraine episode (1a). Unilateral corneal arcus in the right eye (1b, arrow), whereas the left eye shows no arcus (1c). Persisting conjunctival changes with dilated conjunctival vessels and caliber changes in the affected right eye (1d-f) in an attack-free episode. Teleangiectatic vessels (1g).

carotid cavernous fistula, abscess, or atherosclerotic changes of the vessels. Laboratory investigations were unremarkable, particularly screening for thyroid disease, rheumatologic disease, and sarcoid disease were within normal limits. HIV testing was also negative.

Discussion and conclusion

This report describes a woman with migraine in medical history who presents with a unilateral corneal arcus and persisting conjunctival vascular abnormalities. Migraine is a primary recurrent headache disorder that usually occurs in attacks. The attacks typically last at least from 4 up to 72 h. Migraine has two major types: (A) Migraine without aura is a clinical syndrome characterized by headache with specific features and associated symptoms. (B) Migraine with aura is primarily characterized by transient focal neurological symptoms that usually precede or sometimes accompany the headache. Eye symptoms in context with migraine range from short lasting bilateral visual disorders (flickering/scintillation, partial visual field defects, flashes, and/or sensations of colors) over retinal vasculopathy to cranial autonomic parasympathetic symptoms. Parasympathetic activity in migraine induces meningeal vasodilatation due to release of vasoactive peptides including CGRP (calcitonin gene-related peptide), which also affects the eye and seems to appear in around half of migraine patients.[3] In particular, lacrimation and conjunctival injection, but also facial swelling are widely experienced by migraineurs.[3] In our patient, as a further differential diagnosis to migraine, trigeminal autonomic cephalgia (TAC), especially hemicrania continua from remitting subtype, has to be considered. Hemicrania continua from remitting subtype TAC is characterized by a persistent strictly one-sided pain, which is interrupted by remission periods of at least 24 h and is accompanied by ipsilateral cranial autonomic symptoms, such as lacrimation and conjunctival injection.[4]

In our patient we found strictly right sided symptoms and pure trigeminal autonomic presentation, which is rather uncommon for migraine, however, about 20% of the migraine patients have side locked headaches. On the other hand, atypical for TACs would be the recurrent symptoms appearing even without pain and the positive medical history for migraine. As a potential approach to confirm cluster headaches and differentiate them from migraine, a treatment with indomethacin was suggested, but it was not tried, as due to lifestyle changes, the patient had no further severe attacks. However, the dilated conjunctival vessels are persisting.

Interestingly, the autonomic vascular dysregulation in our patient has presumably led to persisting changes of the ocular tissue, i.e., conjunctival vessel alterations. Indeed, similar conjunctival vascular abnormalities, e.g., conjunctival aneurysms, have been reported before in patients with glaucoma and vasospastic syndrome (i.e., patients suffering from conditions where small blood vessels have spasms that limit blood flow). Most probably as a long-time result of this vascular dysregulation with increased vascular permeability, a unilateral corneal arcus has developed in our patient. To the best of our knowledge, this is the first report of such a corneal arcus in a patient with migraine. Unilateral corneal arcus is rare and has been reported in the contralateral eye of patients with occlusive carotid artery disease. Furthermore, it was described after trauma, in patients with Sturge-Weber syndrome, and after periocular erysipelas. In our patients, such etiologies were excluded by MR angiography, clinical examination, optical coherence tomography (OCT), visual field testing, and medical history. Abnormal vascular permeability is assumed to account for the unilateral arcus.

Although the basic pathophysiology of migraine remains unclear, recent evidence has emphasized a neurovascular origin, especially a vasospastic stimulation, and endothelial dysfunction. Hereby, endothelial dysfunction might be both, cause and consequence, of migraine.[5] In addition, the neuropeptide CGRP is thought to have a crucial role in the trigeminovascular system and has been found to be elevated in patients with migraine.[6] CGRP is abundant in trigeminal ganglion neurons and is released from the peripheral and central nerve terminals and secreted within the trigeminal ganglion.

To the best of our knowledge, this is the first description of a unilateral corneal arcus and persisting conjunctival vascular changes as a result of a cranial autonomic dysregulation.

Abbreviations

CGRP	calcitonin gene-related peptide
OCT	optical coherence tomography
TAC	trigeminal autonomic cephalgia.

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None.

Authors' contributions

CT, MD and MA collected the ophthalmological data. HK and RK interpreted neurological pathomechanisms. All authors analyzed and interpreted the patient data. MD and CT drafted the manuscript. All authors read and approved the final manuscript.

Availability of data and materials

All relevant data generated or analyzed are included in this published article.

Consent for publication

Written informed consent was obtained by the patient for the publication of the case report and any accompanying images.

Declaration of conflicting interests

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Ethical approval

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