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Conjunctival congestion in SUNCT syndrome

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Abstract The purpose of this observational-retrospective study was to assess which type of conjunctival injection and related phenomena are present during SUNCT attacks. We studied the videorecords of 23 SUNCT attacks in three patients. Conjunctival injection was found to mostly involve vessels of the palpebral territory stemming from both superior and inferior palpebral vessels that supply the tarsal conjunctiva and most of the ocular (bulbar) conjunctiva. Episcleral injection was also observed. During attacks, the appearance of the conjunctiva suggested conjunctival edema or chemosis. Furthermore, the presence of edema below the capsule of Tenon was strongly suspected. During SUNCT attacks, there was a dramatic conjunctival congestion of the eye on the symptomatic side that tended to extend to the rest of the external tunica of the eye.

Key words SUNCT syndrome • Cluster headache • Trigeminal neuralgia • Unilateral headaches

Introduction

Conjunctival injection, lacrimation, and nasal stuffiness or rhinorrhea on the symptomatic side are integral parts of the clinical picture of cluster headache, chronic paroxysmal hemicrania (CPH), and SUNCT (short-lasting unilateral neuralgiform headache with conjunctival injection and tearing) syndrome. In fact, International Headache Society (IHS) diagnostic criteria for cluster headache and CPH [1], and proposed criteria for SUNCT [2, 3] include these autonomic accompaniments as diagnostically crucial. In cluster headache, there are also distinctive signs such as those related to a possible latent sympathetic hypofunction (Horner-like syndrome) in the symptomatic side [4].

Duration of both pain and accompaniments is by far shorter in SUNCT as compared to cluster headache and CPH. In SUNCT, conjunctival injection occurs in a rather "explosive" fashion considering that vasodilation reaches full, dramatic development within a few seconds after the onset of attacks [5–7] and fades away as quickly as it started. In SUNCT, both abruptness and brevity of attacks may bring about subtle differences regarding the vessels involved in the development of conjunctival injection. Possibly, other signs can be recognized in the conjunctiva and in other tissues of the affected eye and orbital-periorbital area.

We endeavored to analyze which types of conjunctival injection and related phenomena are present during paroxysms and whether more signs of ocular congestion are present during SUNCT attacks. Eventually, this may help in differentiating SUNCT from other, similar headaches.

Table 1 Scales used to grade ocular signs

Lacrimation
Mild (+), Increased conjunctival brightness to increased conjunctival meniscus
Moderate (++), Tears clearly seen but hardly overflowing the palpebral edge
Severe (+++), Tears running down the face

Conjunctival injection
Mild (+), Increased number of vessels seen but not clearly dilated
Moderate (++), Clearly dilated vessels
Severe (+++), Engorged, tortuous and intensely red/violaceous vessels

Conjunctival edema or chemosis
Mild (+), Conjunctiva looks pale and loses transparency
Moderate (++), Bulging of the conjunctiva
Severe (+++), Chemosis prevents appropriate palpebral closure

Material and methods

High-quality videotapes of the attacks of 3 SUNCT patients (2 females and 1 male) were reviewed and analyzed independently by two authors (JAP, JY). One investigator (JY) neither knew the patients nor had access to the clinical records. He saw the patients' faces and the precipitation of attacks, but was not aware of the diagnosis. He was just expected to analyze the ophthalmological signs. He was not present during the recording of the movies.

To obtain an accurate judgement of the observed signs, only close-up scenes (n = 23 attacks) were considered. Attack duration ranged from 80 s to 140 s. The pattern of conjunctival injection and any other "macroscopic" signs in the symptomatic eye during attacks were analyzed.

The extent of lacrimation, conjunctival injection and conjunctival edema was graded according to the scales presented in Table 1. Color photographs of the videorecorded attacks were taken from the monitor by "freezing" selected scenes.

Results

The following observations were made when reviewing the videotaped recordings of 23 SUNCT attacks in three patients:

1. *Increased frequency of blinking*. Increased blinking heralded many attacks (and persisted during attacks). Since all observed attacks were precipitated, expectation and fear of the immediately ensuing attack may have contributed to the observation of this sign.
2. *Blepharospasm* was observed predominantly on the symptomatic side during most attacks in all three patients.
3. *Prominent vasodilation of conjunctival and episcleral vessels*. A few seconds after the onset of painful attacks, there was the rapid development of a dramatic, impressive bulbar and tarsal conjunctival injection (Table 2, Fig. 1) The dilated vessels belonged to the palpebral territory that supplies the tarsal and bulbar conjunctivae. The vascular injection extended toward the episcleral vessels. Injected episcleral vessels were clearly recognized in a deeper layer than conjunctival vessels (Fig. 1).
4. *Conjunctival edema and chemosis*. In the union of the tarsal and bulbar conjunctivae, there were modifications in the aspect of the conjunctivae that were attributed to probable conjunctival edema. These alterations were more intense near the external canthus and in the inferior union of the tarsal and ocular conjunctivae where they

Table 2 Assessment of ocular signs during 23 SUNCT attacks in 3 patients

Attack	Lacrimation	Conjunctival injection	Conjunctival edema
Patient 1			
1	+++	+++	+
2	+++	+++	+
3	+++	+++	++
4	+++	+++	++
5	+++	++	++
6	++	++	+
7	+++	++	+
8	++	++	++
9	++	++/+++	+
10	++	+++	++
11	++	+++	++
12	+++	+++	++
13	+++	+++	++
Patient 2			
1	+++	++	ND
2	+++	+++	+
3	+++	++	+
4	+++	++	+
5	+++	+++	ND
Patient 3			
1	+++	+++	ND
2	+++	+++	ND
3	+++	+++	+
4	+++	+++	+

ND, not determined

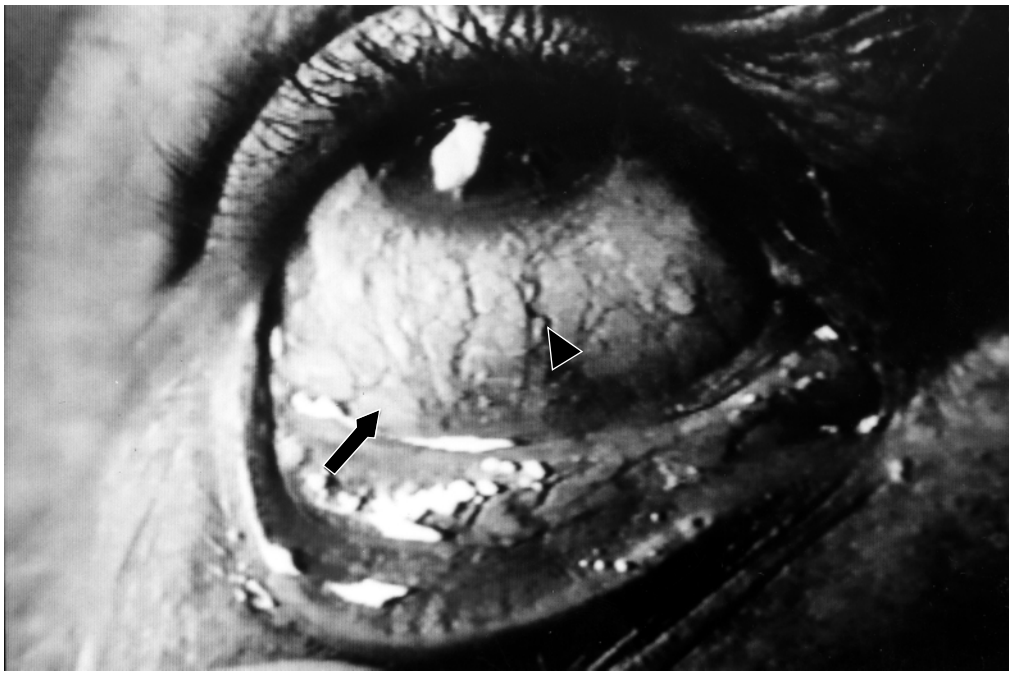

Fig. 1 SUNCT attack no. 3 in patient 1. An injection of the vessels of the palpebral and tarsal conjunctivae is observed. One particularly engorged conjunctival vessel is indicated (*arrowhead*). Episcleral injected vessels can be recognized as lying deeper than conjunctival vessels. Moderate conjunctival edema can be seen in the inferior union of the tarsal and ocular conjunctivae (*arrow*). That particular area increased in the course of the attack and quickly disappeared afterwards

were considered to be almost chemosis (Fig. 1). Edema below the capsule of Tenon was suspected but could not be confirmed. Assessment of conjunctival edema in the 3 patients is detailed in Table 2.

5. *Swelling of eyelids* and decreased palpebral width were observed, without associated miosis.
6. *Lacrimation* was prominent in virtually all observed attacks (Table 2).

The ocular signs observed during SUNCT attacks are summarized in Table 3.

Table 3 Ocular signs during SUNCT attacks, observed in the present study and in [5–7, 9]

Ocular sign	Location
Increased frequency of blinking	Bilateral
Blepharospasm	Predominantly on symptomatic side
Conjunctival edema and chemosis	Symptomatic side
Swelling of eyelids	Symptomatic side
Conjunctival injection	Symptomatic side
Episcleral injection	Symptomatic side
Prominent lacrimation	Symptomatic side
Increased IOP and CIP	Symptomatic side
Increased corneal temperature	Predominantly on symptomatic side

IOP, intraocular pressure; *CIP,* corneal indentation pulse amplitudes

Discussion

Conjunctiva is supplied by two vascular beds stemming from the ophthalmic artery (internal carotid system):

1. A great *palpebral territory* supplied by both superior and inferior palpebral vessels that feed the tarsal conjunctiva and most of the ocular (bulbar) conjunctiva.
2. A small *ciliary territory* supplied by the anterior ciliary vessels feeding the ocular conjunctiva surrounding the cornea. The ciliary territory is in close relationship with vessels of the ciliary muscle and iris.

Between both territories, anastomoses are scarce and limited to the boundaries. Thus, there is a certain independence of the respective vasoreactivities when involved in pathological processes [8].

This particular vascular bed produces two different types of conjunctival injection that are clearly related to the origin of the congestion. When a primary process affects the conjunctiva the palpebral territory is injected, whereas in processes involving ciliary muscle and iris an injection surrounding the cornea is prominent. In other words, the underlying mechanisms are different whether conjunctival injection or ciliary injection are present – or predominant.

Macroscopically, ciliary-type injection can be seen as a thin, delicate, reddish, vascular halo surrounding the cornea. In additon, there are associated signs involving pupil size, as in glaucoma associated with mydriasis, or iritis and cheratitis most frequently associated with miosis. In

conjunctival-type injection, the dilated vessels appear rather tortuous and engorged – typically in the periphery. Changes in pupil size are not typical of conjunctival-type injection.

Ciliary-type injection is related to intrinsic – corneal and anterior segment – ocular disorders. Conversely, conjunctival-type injection is frequently produced by local "irritation" of the conjunctiva. Contact with irritants and allergens may produce short-lived conjunctival edema and chemosis that may develop within seconds.

According to our observations, conjunctival injection during SUNCT attacks seems to be of the conjunctival-type. Congestive signs of the conjunctiva may include engorgement of both tarsal and bulbar conjunctival vessels, conjunctival edema, and eyelid swelling (both local edema and vessel engorgement may be the cause of eyelid swelling). These signs have been observed in our patients. Indeed, extension of the congestive process towards the episcleral vessels has also been observed. From the ophthalmological point of view, episcleral injection is frequently accompanied by conjunctival edema. Therefore, it is not surprising to observe both signs appearing in tandem during SUNCT attacks.

It was also suspected that the edema might have reached the space below the capsule of Tenon. Interestingly, distension of this virtual space is very painful. However, without a direct examination by the slit lamp it is difficult to ascertain this latter point.

Generally speaking, the presence of such an array of congestive signs points toward a sudden flow of blood into the orbita or a local release of vasoactive substances and mediators of the inflammation. The possibility of impaired venous blood drainage from the eye to the cavernous sinus seems to be unlikely, taking into account that during SUNCT attacks episcleral venous pressure does not change [9]. Dramatic onset and unilateral expression of the pain and accompaniments both suggest a neurogenic triggering of the process, supposedly through the first division of the trigeminal nerve. Moreover, the triggering of attacks by precipitating mechanisms mostly acting on structures supplied by the trigeminal nerve strengthens the concept of a neurogenic origin. Secondary activation of the parasympathetic fibers of the facial nerve may contribute to some of the autonomic features. However, it hardly explains the changes observed in the conjunctiva that were considered edema-chemosis and that are reminiscent of the processes that produce local "irritation" in such a mucous membrane. In the future, analysis of the lacrimal and nasal secretions – during SUNCT attacks – should be undertaken in order to disentangle whether vasoactive substances, neuropeptides, and mediators of inflammation are locally released into the orbit and, particularly, within the external tunica of the eye.

During attacks, there is an increase in intraocular pressure (IOP), corneal indentation pulse amplitudes (CIP), and corneal temperature (Table 3) [9]. This marked change in ocular variables may be explained by a sudden increase in local blood supply [9]. Nevertheless, impaired drainage of the aqueous humor from the anterior camera of the eye owing to both increased pressure in the episcleral arteries and local congestion might also contribute to the observed increase in IOP.

One ocular feature not observed in the present series, but worth mentioning, is conjunctival hemorrhage. Conjunctival bleeding may be a rare feature of conjunctival congestion maxima. Of course, vascular fragility diathesis should also be considered whenever this sign is observed. Interestingly, conjunctival hemorrhage has been only rarely observed in other ocular or periocular painful syndromes such as CPH and cluster headache, and in idiopathic stabbing headache (jabs and jolts syndrome) (personal observations and [10, 11]).

In conclusion, conjunctival injection during SUNCT attacks seems to be of the conjunctival type. Possibly, other signs of conjunctival involvement may be seen during SUNCT attacks, including conjunctival edema with small areas of chemosis, and extension of vasodilation to the episcleral vessels. Therefore, conjunctival congestion may be a more comprehensive term for the whole array of features observed. Whether these findings hold true for other paroxysmal headaches with local autonomic accompaniments must await further observations.

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