

SUNCT responding to botulinum toxin

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BACKGROUND

- Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is a disabling primary headache disorder: a form of trigeminal autonomic cephalalgias (TACs)^[1-4].
- SUNCT is characterized by repetitive short-lasting attacks of moderate to severe, strictly unilateral pain, occurring in the distribution of the ophthalmic and maxillary divisions of the trigeminal nerve, associated with ipsilateral marked conjunctival injection and lacrimation.
- The duration of headaches typically range from 5–240 seconds and the frequency varies from three attacks daily to more than 60 attacks per hour.
- Other autonomic accompaniments include rhinorrhea, nasal stuffiness, forehead and facial sweating, hyperventilation, increased intraocular pressure on the symptomatic side, vascular engorgement and swelling of the eyelids with decreased palpebral width (pseudoptosis).
- SUNCT is more common in males and the mean age at onset is around 50 years, with a range of 10 to 77 years.
- Treatment can be challenging, but lamotrigine may be helpful as a preventive treatment of SUNCT based on case reports and small open-label series^[5].
- We describe a case of SUNCT that responded well to botulinum toxin injection.

CASE REPORT

- A 46 year old male developed an episodes of viral meningitis in 1994 which was complicated by non-communicating hydrocephalous and was treated with ventriculo-peritoneal shunt. He had a shunt revision in 2001 which was complicated by shunt infection which was then removed after two weeks. Following this he started to develop episodic excruciating headaches.
- A typical headache is characterized by severe pain which starts in the right suprascapular area and radiates to the right side of face and right retro-orbital region associated with right eye injection and tearing, right eye lid swelling and small palpebral fissure. There was no papillary change.
- Each episode lasts 30–90 seconds and recurs up to 5–6 times per hour. Over time he developed mild right side cervical dystonia with elevation of the right shoulder and tilting of the head to the right side which may become prominent during an episode of headache.
- Sumatriptan, valproic acid, gabapentin and indomethacin failed to provide benefit. He underwent onabotulinumtoxinA (Botox) injection as follows: right brow 30 units, right masseter 75 units, right temporalis muscle 50 units, right trapezius 100 units, and right splenius 100 units.



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RESULTS

- There was dramatic improvement in the intensity and frequency of headache (from 20–30/day to only 2–3/week) following onabotulinumtoxinA injection. The benefit was sustained for 3–4 months after which time the frequency and intensity of the headaches gradually return but never to the baseline frequency.
- The patient continued to benefit from repeat injections 3–4 times per year for the past three years without any adverse effects. MRI brain did not show any pituitary abnormality.



Figure 1: This picture was taken immediately following an episode of SUNCT showing right eye conjunctival injection tearing and pseudoptosis. These resolved within the next 5 to 10 minutes.

Table 1: International Headache Society diagnostic criteria for SUNCT

- At least 20 attacks fulfilling criteria B-D
- Attacks of unilateral orbital, supraorbital, or temporal stabbing or pulsating pain lasting 5–240 seconds
- Pain is accompanied by ipsilateral conjunctival injection and lacrimation
- Attacks occur with a frequency from 3 to 200 per day
- Not attributed to another disorder

Table 2: Differential diagnosis of very brief headaches

- SUNCT (primary and secondary forms)
- SUNA (short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms)
- Trigeminal neuralgia
- Primary stabbing headache
- Paroxysmal hemicrania

Table 3: Secondary causes of SUNCT

- **Posterior fossa abnormality**
 - Ipsilateral cerebellopontine angle arteriovenous
 - A brainstem cavernous hemangioma
 - A posterior fossa lesion in a patient with HIV/AIDS
 - Severe basilar impression causing pontomedullary compression in a patient with osteogenesis Imperfecta
 - Craniosynostosis resulting in a foreshortened posterior fossa
 - Ischemic brainstem infarction.
- **Pituitary abnormality**
 - Pituitary micro or macro adenoma

DISCUSSION

- Our patient satisfied the International Headache Society diagnostic criteria for SUNCT (Table 1). The differential diagnosis of very brief headaches are listed in Table 2.
- Majority of SUNCT cases are idiopathic, but a minority of them can be secondary to intracranial abnormalities (Table 3). Our patient likely falls in to secondary category given the underlying non-communicating hydrocephalus but what actually triggered the onset of headaches is unclear.
- The activation of the posterior hypothalamus during attacks, demonstrated in functional neuroimaging studies, has led to the hypothesis that the posterior hypothalamus plays a crucial role in the pathophysiology of SUNCT syndrome.
- The effectiveness of botulinum toxin (BoNT) in chronic migraine has been shown in multiple double blind placebo controlled studies.
- BoNT in SUNCT has not been formally evaluated. There is one case report showing sustained benefit of BoNT in a patient with SUNCT refractory to oral treatments. In this patient onabotulinumtoxinA was infiltrated at four points around the orbit, injecting 10 U at each site and was injected every three months and the benefit was sustained after 2.5 years of follow-up^[6].
- The mechanisms by which BoNT helps in migraine prophylaxis and other headache disorders is still incompletely understood. Studies have shown that BoNT can affect the release of various neurotransmitters, particularly those involved in pain (e.g., substance P, glutamate, and calcitonin-gene-related peptide). Central mechanisms may also play a role^[7].
- There was return of symptoms, although not as severe, when the effect of BoNT was wearing off suggesting a more direct analgesic effect. BoNT effectively controlled both the pain as well as the autonomic activation in our patient.

CONCLUSIONS

- OnabotulinumtoxinA injection is an option for refractory cases of SUNCT. The benefit was sustained for the follow up period of three years.
- Further studies are needed to confirm this observation.

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