Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome is a rare form of primary headache disorder, although secondary causes, particularly posterior fossa abnormalities, are well known. We report a case of SUNCT syndrome secondary to a prolactinoma. Administration of dopamine agonists led to complete resolution of the SUNCT attacks. This case, together with other similar case reports in the literature, highlight the importance of excluding a diagnosis of pituitary adenoma in all suspected cases of SUNCT syndrome, especially as the headache can precede more classical pituitary symptoms by a considerable period of time. Clinicians managing patients with suspected SUNCT syndrome should elicit a history of symptoms associated with pituitary neoplasms, perform a magnetic resonance imaging scan of the brain and pituitary, and screen for serum hormonal abnormalities.

S

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome is a rare form of strictly unilateral headache that occurs in association with cranial autonomic features. The pain is abrupt in onset, of short duration, stabbing or burning occurs in association with cranial autonomic features. The usual duration of the pain was 20–30 s, although the range was 10–60 s. The pain came on rapidly, was maintained at a plateau phase, and then resolved rapidly. He denied any superimposed spikes or variations in the severity of the pain. He had 1–6 attacks daily at presentation. The attacks were associated with prominent ipsilateral conjunctival injection and lacrimation, and bilateral facial flushing. He denied all other cranial autonomic features, or any nausea, vomiting, photophobia, phonophobia or osmophobia. He felt restless during the pain and would often pace up and down. He denied any aura symptoms. Occasionally he had a dull interictal pain that lasted 10–60 min. The headaches could be triggered consistently by exercise, particularly cycling, and inconsistently by stressful situations. He denied having any other triggers for the headaches. He was unsure whether there was a refractory period after an attack; the shortest interval between two spontaneous attacks was about 3 min. He could abbreviate the attack by coughing. There was no past history of headaches.

The patient had previously tried ibuprofen, aspirin, distalgesic and indomethacin 50 mg three times daily for the headaches without any benefit. At presentation, he was not taking any drugs. In the past medical history, he had had a tonsillectomy in childhood. His sister was diagnosed as having cancer of the thyroid gland at the age of 21 years. There is no family history of headaches. He is a non-smoker and drinks 25–30 units of alcohol per week.

General and a detailed neurological examination were entirely normal. An MRI scan of the brain revealed a pituitary adenoma on the right extending towards the right cavernous sinus, causing displacement of the right internal carotid artery; the pituitary adenoma also extended into the chiasmatic cistern but did not impinge on the optic chiasm (fig 1). Formal visual field testing was normal. Blood tests showed that the serum prolactin level was markedly elevated at 10 000 mU/L (normal range 0–635) and testosterone was at the lower end of the normal range (10.1 nmol/L; normal range 9–33) with no elevation of luteinizing hormone (LH) or follicle stimulating hormone (FSH), while free thyroxine, thyroid stimulating hormone (TSH), basal cortisol, adrenocorticotropic hormone (ACTH), oestrogen, and growth hormone (GH) levels were normal. A diagnosis of macroprolactinoma was made.

At that point, the patient was re-interviewed to elicit features of pituitary disease. He reported that, in retrospect, there might have been a slight reduction in libido over the

**Abbreviations:** ACTH, adrenocorticotropic hormone; fMRI, functional magnetic resonance imaging; FSH, follicle stimulating hormone; GH, growth hormone; LH, luteinising hormone; SUNCT, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing; TSH, thyroid stimulating hormone
previous few years, although sexual function had otherwise been normal. He had always had scanty beard growth, but otherwise secondary sexual development and characteristics were normal. There was no galactorrhoea and vision was normal.

The patient was started on bromocriptine 2.5 mg twice daily, and the prolactin level normalised within 3 months. The bromocriptine dose was thereafter reduced to 1 mg twice daily. The headaches started improving shortly after starting bromocriptine and resolved completely within 3 months. A repeat MRI scan of the brain in February 2000 showed that the pituitary adenoma had virtually disappeared. The patient continued taking bromocriptine for 18 months but began to develop nausea and nasal blockage; bromocriptine was therefore substituted with cabergoline 1.5 mg/week. He has been on cabergoline over the last 18 months, on which the prolactin level remains normal and there are no side-effects. He denies having had any further headaches and his libido and energy levels have significantly improved.

**DISCUSSION**

The phenotype of this headache satisfies diagnostic criteria for SUNCT.\(^4\)\(^5\) Medical treatment of the prolactinoma led to a resolution in headache symptoms, which strongly indicates that the prolactinoma was pivotal in triggering the headache, rather than a coincidental pathology.

Most cases of SUNCT syndrome described in the medical literature are primary but several cases of secondary SUNCT have been reported. The secondary SUNCT cases can be divided into two groups: posterior fossa and pituitary abnormalities. There are seven case reports of SUNCT syndrome secondary to a posterior fossa abnormality including: homolateral cerebellopontine angle arteriovenous malformations in two patients;\(^6\)\(^7\) a brainstem cavernous hemangioma;\(^8\) a posterior fossa lesion in an HIV/AIDS patient;\(^9\) severe basilar impression causing pontomedullary compression in a patient with osteogenesis imperfecta;\(^10\) craniosynostosis resulting in a foreshortened posterior fossa;\(^11\) and ischaemic brainstem infarction.\(^12\)

There are four case reports of SUNCT syndrome secondary to pituitary adenomas in the medical literature. Before the acronym “SUNCT syndrome” had been proposed, a patient with a non-functioning pituitary macroadenoma and trigeminal neuralgia attacks, compatible with a diagnosis of SUNCT syndrome, had been described.\(^12\) The differential diagnosis between trigeminal neuralgia and SUNCT can be difficult.\(^13\)\(^14\)\(^15\)\(^16\)\(^17\)

Two patients with SUNCT syndrome and prolactinomas have been described; one of the patients had a microprolactinoma while the other had a macrolactinoma with cavernous sinus invasion.\(^18\) Recently, we have described a patient with SUNCT syndrome and microlactinoma.\(^19\)

The pathophysiology of pituitary-associated headache is poorly understood, although dural stretch, cavernous sinus invasion, and local pressure effects have been proposed as possible mechanisms.\(^10\)\(^11\)\(^12\) In all the case reports of SUNCT syndrome secondary to pituitary tumour, including the case described here, the SUNCT attacks occurred on the side ipsilateral to the side of the tumour, suggesting a role for a mechanical mode of action. However, though a mechanical effect may be a conceivable mechanism for the headache in cases of pituitary macroadenomas and cavernous sinus invasion, it is not a satisfactory explanation for macroadenomas. Furthermore, a recent study in patients with pituitary tumours has demonstrated that there is a poor correlation between headache score and tumour size (hence dural stretch), cavernous sinus invasion and degree of carotid artery invasion.\(^20\) A plausible explanation is that the headaches are predominantly neurohumorally mediated rather than mediated by the size or invasiveness of the tumour. In this respect, it is interesting to note that in all case reports of SUNCT syndrome secondary to pituitary tumour, except the case reported here, dopamine agonists induced SUNCT attacks. This indicates a role for the dopamine axis in the aetiology of pituitary-associated headache and indicates that understanding the role of the hypothalamopituitary axis in the pathophysiology of these headaches may be very instructive.

This case report highlights the importance of appreciating that SUNCT syndrome can occur secondary to a pituitary tumour. The significance of this is further underlined by the findings that headache was the initial presentation in all cases reported hitherto, and that there can be a considerable time lag before the onset of pituitary-related symptoms. In the case reported by Ferrari et al, there were no pituitary symptoms despite a 10-year history of headaches.\(^21\) In the two cases reported by Massiou et al, the headaches preceded the pituitary symptoms by 3 and 4 years respectively.\(^22\) All patients presenting with suspected SUNCT syndrome should be questioned for symptoms associated with pituitary neoplasms, either due to hormonal imbalance or mass effect. Both the pituitary and posterior fossa abnormalities emphasise the absolute need for a cranial MRI, including an adequate view of the pituitary. In addition, these patients should have a screen for basal hormone measurements, including prolactin, TSH, free thyroxine, cortisol, ACTH, LH, FSH, oestradiol, testosterone and GH.

**ACKNOWLEDGEMENT**

PJG is a Wellcome Trust Senior Research Fellow.

**Authors’ affiliations**

M S Matharu, M J Levy, P J Goadsby, Headache Group, Institute of Neurology

R T Merry, The National Hospital for Neurology and Neurosurgery, Queen Square, London, UK

Competing interest: None declared

Correspondence to: Professor P J Goadsby, Institute of Neurology, Queen Square, London WC1N 3BG UK; peterg@ion.ucl.ac.uk
REFERENCES


Want to know more?

Data supplements

Limited space in printed journals means that interesting data and other material are often edited out of articles; however, limitless cyberspace means that we can include this information online.

Look out for additional tables, references, illustrations.

www.jnnp.com
SUNCT syndrome secondary to prolactinoma

M S Matharu, M J Levy, R T Merry and P J Goadsby

*J Neurol Neurosurg Psychiatry* 2003 74: 1590-1592
doi: 10.1136/jnnp.74.11.1590

Updated information and services can be found at:
http://jnnp.bmj.com/content/74/11/1590

These include:

**References**
This article cites 20 articles, 3 of which you can access for free at:
http://jnnp.bmj.com/content/74/11/1590#BIBL

**Email alerting service**
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Topic Collections**
Articles on similar topics can be found in the following collections

- Headache (including migraine) (459)
- Neurooncology (237)
- Pain (neurology) (763)
- Drugs: CNS (not psychiatric) (1945)
- Radiology (1747)
- Radiology (diagnostics) (1309)

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/