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Case Report

SUNCT: The Rarest Primary Headache in Indian Perspective

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Abstract

Short lasting neuralgiform headache with conjunctival injection and tearing (SUNCT) is perhaps the rarest primary headache, which has lately been classified by the international headache society (IHS). The very term, SUNCT, enumerates its features; however, due to its rarity and poor awareness on the part of treating physicians, it is liable to be misdiagnosed and wrongly treated. Demographic data are lacking in India and worldwide, and most of the informations we have, are through case reports and case series. This is also a small effort, one step forward, in that line to share our observations and views of a series of nine patients at our centre in India.

Keywords: Short lasting unilateral neuralgiform headache with conjunctival injection and tearing; Primary headache; Trigeminal autonomic cephalgias; Paroxysmal hemicranias; Cluster headache; Trigeminal neuralgia

Abbreviations

SUNCT: Short lasting Unilateral Neuralgiform headache with Conjunctival injection and Tearing; TAC: Trigeminal Autonomic Cephalgias; PH: Paroxysmal Hemicranias; CH: Cluster Headache; TN: Trigeminal Neuralgia; MRI: Magnetic Resonance Imaging; DBS: Deep Brain Stimulation

Introduction

Short lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) is a very rare primary headache, characterized by brief paroxysm of agonizing unilateral orbital or periorbital pain, associated with marked autonomic symptoms [1]. It was first described in 1978 by Sjaastad [2], and was classified among the subclasses of Trigemino-autonomic-cephalgias (TAC) by the International Headache Society (IHS) in 2004 [3]. However, owing to its rarity and overlap with the symptoms of Trigeminal Neuralgia (TN), Paroxysmal Hemicranias (PH) and Cluster Headache (CH), it is liable to be misdiagnosed and wrongly treated. Here we are reporting a series of nine cases of SUNCT from northern India, discussing the demographic and clinical details experienced at our centre; with the aim to highlight its distinguishing features, thus facilitating prompt diagnosis and management.

Materials and Methods

This observational study was conducted at SMS Medical college Hospital, Jaipur; a large teaching hospital of northern India, over a period of 10 years. All patients presenting with brief paroxysm of unilateral craniofacial pain and autonomic symptoms were considered for further clinico-historical evaluation of attacks; in terms of frequency, duration, location, severity, trigger ability and response to any previous medication. A final diagnosis was made only after thorough neuro-ophthalmological examination, neuroimaging, and applying IHS diagnostic criteria for SUNCT (Table 1). A total of 89 patients were taken for evaluation, of which only 9 patients were finally diagnosed as SUNCT.

Results and Discussion

Various demographic and clinical characteristics have been elucidated in the Table 2. Of the nine patients, all but one were males; reaffirming its male preponderance. Exact gender predilection is not known, however, 2:1 to 17:2 Male: Female (M: F) ratio have been proposed in various studies [4,5]. Though a small series, our study also had 8:1 M:F ratio, quite in keeping with the other series. However, socio-cultural issues may also influence the gender predilection especially in rural population.

The mean age of presentation was 53 years, ranging from 27 years to 63 years; however most of them (7/9) were above 50-year, which is also in keeping with the previous series where 50-year was the mean age [6].

Total duration of illness at the time of presentation ranged from 1-4 years, which perhaps speaks about the poor awareness and access to the health facilities in this part of the country.

Most of them (5/9) had frequency in the range of 40-60 attacks per day, three of them had frequency in the range of 100 and above per day, whereas one of them had 20-30 attacks per day. Most of the attacks (6/9) subsided in less than a minute time, and all by three minutes; commensurating with the diagnostic time frame (5-240 sec). Frequency of the attacks may range from 3-200 per day, however at least 20 attacks of the specified character and duration are required to qualify the diagnostic definition of SUNCT [3].

Attacks in all the cases were very severe and periorbital in

Table 1: Diagnostic criteria for SUNCT syndrome.

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

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Table 2: Demographic and clinical details of the patients.

name	age/ Sex	duration of illness	frequency	severity	character	location	laterality	autonomic features	attack duration	trigger	neuro -logic deficit	MRI-brain	treatment	response	others
a.s.	60/m	2 y'rs	40-50/d	very severe	electric current like	periorbital	right	redness, lacrimation, ptosis, nasal stuffiness	1-2 min.	cold water, cold breeze	none	normal	lamotrigine	fair	none
b.k.	27/m	4 y'rs	>100/d	very severe	stabbing	periorbital	right	redness, lacrimation, nasal stuffiness	50-60 sec.	none	none	normal	lamotrigine	fair	none
n.m.	58/m	3 y'rs	50-60/d	very severe	burning, throbbing	orbital and periorbital, upper face	left	redness, lacrimation, ptosis, nasal stuffiness	2-3 min.	cold water, cold breeze, hot food	none	normal	lamotrigine and carbama-zepine	partial	none
m.d.	63/f	1 y'r	50-60/d	very severe	electric current like	periorbital	right	redness, lacrimation, nasal stuffiness	1-2 min.	none	none	normal	lamotrigine	fair	none
y.a.	55/m	2 y'rs	~100/d	very severe	electric current like	orbital and periorbital	right	redness, lacrimation, ptosis, nasal stuffiness	30-40 sec.	none	none	normal	lamotrigine, amitryptiline	fair	h/o TTH
g.r.	59/m	3 y'rs	40-50/d	very severe	stabbing	periorbital and temporal	left	redness, lacrimation, nasal stuffiness	30-40 sec.	none	none	normal	lamotrigine	fair	none
o.s.	60/m	1 y'r	20-30/d	very severe	electric current like	periorbital	right	redness, lacrimation, ptosis, nasal stuffiness	40-50 sec.	none	none	normal	lamotrigine	partial	none
k.m	53/m	2 y'rs	100-150/d	very severe	electric current like	orbital and periorbital	right	redness, lacrimation, nasal stuffiness	40-60 sec.	none	none	normal	lamotrigine	fair	none
d.k.	48/m	1 y'r	40-50/d	very severe	electric current like	periorbital	left	redness, lacrimation, nasal stuffiness	30-45 sec.	none	none	normal	lamotrigine	fair	none

location, with orbital (3/9), temporal (1/9) and upper face (1/9) being additional locations. Though these are the chief locations, yet it could be located anywhere in the head [3,7].

Most of them (6/9) described the character of pain to be electric shock like; two of them, stab like and one patient described it to be burning or throbbing. Attacks may be in the form of single stab, a group of stabs, or a longer attack comprising many stabs between which the pain does not resolve completely (saw-tooth phenomenon) [7].

Interestingly, majority (6/9) of the patients had it on right side and three of them had it on left side. Though only a few studies have considered laterality issue so far [7]; we propose other series should also look into the laterality aspect to substantiate it further.

All the patients (9/9) had prominent autonomic symptoms like redness of eye, due to conjunctival injection; lacrimation; and nasal stuffiness. Four of them additionally had ptosis of the same side. These are the most distinguishing features of this condition [3,8].

Most of the patients had the attacks spontaneously; however, history of triggers in the form of splash of cold water, cold breeze against the face and hot food were forthcoming in two patients on probing. Presence of cutaneous triggers is the point which helps in differentiating this condition from PH and CH [9].

One of the nine patients had additional history of Tension Type Headache (TTH), reaffirming the fact that more than one type of headache can coexist in the same patient warranting treatment for the two separately [10].

Detailed neuro-ophthalmological examination was normal in all the patients in between attacks. MRI- brain including pituitary and posterior fossa structures were normal in all the patients, so also the routine blood parameters including pituitary function tests, and CXR; thus ruling out the possibility of secondary SUNCT.

Seven of the nine patients showed fair response to the treatment with lamotrigine in the dose of 100-200 mg/d over 2-3 weeks time, but two of them showed only partial response. One of those who showed only partial response had to be given lamotrigine in the dose of 400mg/d with little additional benefit. Many drugs like lamotrigine, topiramate, gabapentine, valproate, verapamil, nifedipine have been used in the treatment of SUNCT but recent reports suggest lamotrigine to be more efficacious than others, hence was our obvious choice [11]. Hypothalamic Deep Brain Stimulation (DBS) is a potential tool and hope for the future therapeutic avenues. Surgical treatments like Janetta procedure, percutaneous trigeminal

Table 3: Differential diagnosis of short lasting headaches.

	CH	PH	TN	SUNCT
Gender	M>F	F≥M	F>M	M>F
Pain Character Severity Location	 Stabbing, boring Very severe Orbital, Temple	 Throbbing, boring, Stabbing Very severe Orbital, temple	 Stabbing, electric Shock like Very severe V2/V3>V1	 Stabbing, burning, electric shock like Severe to very severe Periorbital, orbital
Frequency of attack	1/alternate day-8/day	1-40/day (5/day for more than half the time)	Any	3-200/day
Duration of attack	15-180 min	2-30 min	<1 sec	5-240 sec
Autonomic features	+	+	-	+
Alcohol trigger	+	±	-	-
Cutaneous triggers	-	-	+	+
Response to Indomethacin	-	+	-	-
Refractory period			+	-

ganglion compression, etc.; are reserved for cases refractory to medical management. SUNCT, secondary to posterior fossa or pituitary lesions, also warrant surgical treatment. However, results are not very encouraging.

Two of the nine patients were treated earlier with indomethacin, considering it to be Paroxysmal Hemicrania (PH), but to no avail. PH is definitely a close differential owing to its unilaterality, orbital or temporal location, brevity of attack and presence of autonomic symptoms. However, relatively longer duration of attack (2-30 min), lesser frequency (1-40/d), absence of cutaneous triggers and marked response to indomethacin distinguish it from SUNCT (Table 3) [11,12].

Two of them were labeled trigeminal neuralgia and receiving carbamazepine with minimal improvement. Again, trigeminal neuralgia is an important differential; as the pain is brief, usually unilateral, electric shock like, very much akin to SUNCT. However, TN rarely involves orbital or periorbital regions (V1 distribution) plus presence of refractory period and lack of autonomic symptoms make its diagnosis obvious [10,13].

Being one of the TACs, cluster headache could form another differential; however, longer duration of attack (15-180 min), lesser frequency (1/alternate day-8/d), periodicity, presence of alcohol trigger and absence of cutaneous triggers are the hallmark of the disease [3].

Goadsby and Lipton suggested that the pathophysiology of the two chief components of TACs, viz. trigeminal distribution of pain and ipsilateral autonomic features, result from the central disinhibition of trigeminal-autonomic reflexes. Much is not known about natural history of the disease but it is considered to be a lifelong disorder, though cases undergoing remission for variable periods are on record. And it does not cause any long term neurological sequelae [4].

Conclusion

SUNCT is one of the rarest of primary headaches, classified under the group called TACs by IHS. It is a disease of past middle age with obvious male preponderance. No incidence or prevalence data are available, worldwide in general and India in particular. Under-recognition and misdiagnosis of this condition due to poor awareness on the part of physicians could be one of the reasons. So for all practical purposes, any paroxysmal, unilateral headache lasting for less than four minutes, with marked redness and lacrimation of ipsilateral eye should be further observed and evaluated to rule out SUNCT. Though it is often misdiagnosed as PH, CH or TN; yet carefully taken history and patient clinical observations would help make a confident diagnosis. It is generally thought to be poorly responsive to treatment but of late lamotrigine has been found to be particularly efficacious in many case series including ours.

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