Red Ear Syndrome: Case Series and Review of a Less Recognized Headache Disorder

Sir,

Red ear syndrome (RES), originally described by Lance (1994), is characterized by pain and redness of one or both external ears with a burning sensation.[1] Pain may radiate to the mandible, cheek, and occiput from the pinna.[1,2] The attacks can last for seconds to hours but commonly occur for 30 min to an hour.[2-4] Its etiology, pathophysiology, and treatment are unclear. It is not yet included in the international classification

of headache disorders version II or IIIβ [ICHD II and ICHD-IIIβ].[1]

Herein, we report three patients with RES and review the published literature on this rare syndrome.

Case 1: A 23-year-old female presented with episodic severe left ear pain with reddening of the pinna, 5–6 times per month, for 6 months, each episode lasting for 4–5 h [Figure 1]. Episodes were triggered by sunlight, wearing hairband, combing the hair prior to travel in the afternoon, lack of sleep, or travel post-lunch. She did not have a migraine or another headache. She felt relief with ice pack application, rest, or diclofenac 50 mg tablet. Her examination, MRI head and cervical spine were normal. Her symptoms remitted with 10 mg/day amitriptyline for 6 months and are currently on medicines.

Case 2: A 21-year-old male presented with daily episodes of severe left ear pain and reddening [Figure 2] with a throbbing hemicranial headache with nausea, photo-phonophobia, precipitated by missing breakfast, playing in the sun, or travelling for 3 months. While the reddening of ear and pain would last for 3–4 h, the headache would last for 6–8 h and improve by paracetamol or sleep. His examination and brain imaging were normal. He improved with 5 mg/day of flunarizine for 6 months and was symptom-free now without medicines for 3 months.

Case 3: A 26-year-old female presented with episodic bilateral earache with reddening of both ears, lasting 2–4 h with a burning sensation, precipitated by eating sour food, strong perfume, getting emotional, or speaking a lie, without any headache, nausea, and vomiting [Figure 3]. The episodes were infrequent, about 1–2/month but causing social embarrassment. The episodes would improve with rest or paracetamol. Her examination including dental examination and MRI brain and cervical spine were normal. Since she had only occasional nondisabling episodes she did not want prophylactic treatment.

These three patients we described had characteristics of RES with episodic paroxysmal attacks of unilateral/bilateral ear erythema along with burning ear pain.[1] In RES, the extent of pain and redness can be variable: pain can be dull, wounding, sharp and poking; the extent of pain-redness could be at the pinna, with the inclusion of cheek, temple, upper neck, and whole face ipsilateral to the influenced ear.[1-3,5,6] Heat, rubbing, touching the ear, neck movements, brushing of the hair, biting, tooth pounding, stress, or specific dietary elements work as triggers.[1,7]

Central, as well as peripheral dysfunction in variable ways, have been suggested by different authors as the pathophysiologic abnormality in RES. Central brainstem trigemino-facial neurovascular-axis dysfunction in lines with migraine and various trigeminal autonomic cephalalgias has been proposed by some scientists.[1,4,8] Others have suggested a peripheral dysfunction in the trigeminal nerve or C3 nerve root irritation as the proposed hypothesis for RES.[1,3] The link of RES to the skin disease erythromelalgia has also been suggested.[1,9,10] We suggest a unified hypothesis considering a combination of both central dysfunctions as well as peripheral dysregulation as the pathophysiologic dysfunction in RES [Table 1].

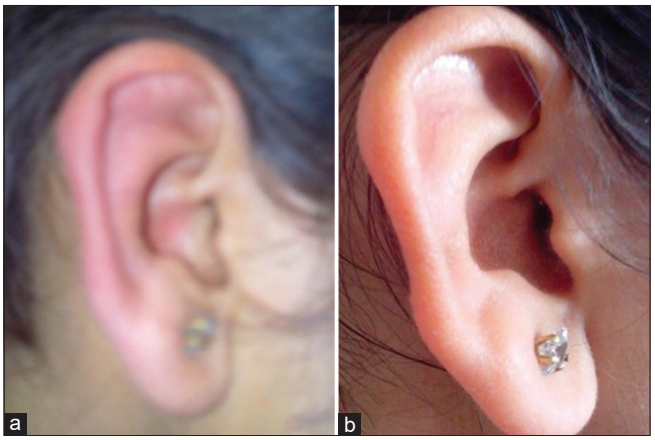

Figure 1: Red ear in case 1: (a) During the paroxysm of pain. (b) Normal ear when pain paroxysm is over

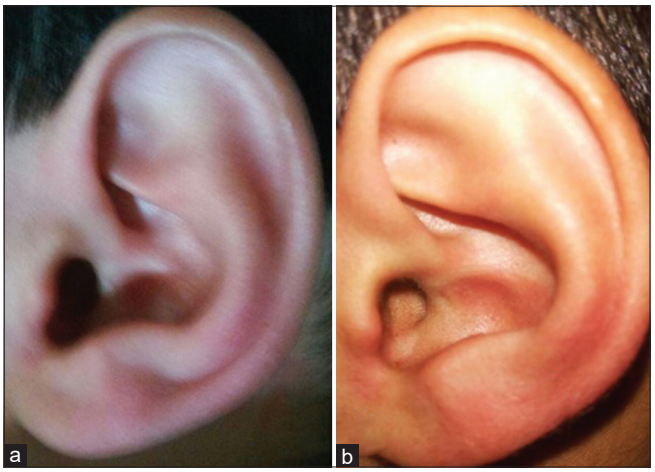

Figure 2: Red ear in case 2: (a) During the paroxysm of pain. (b) Normal ear when pain paroxysm is over

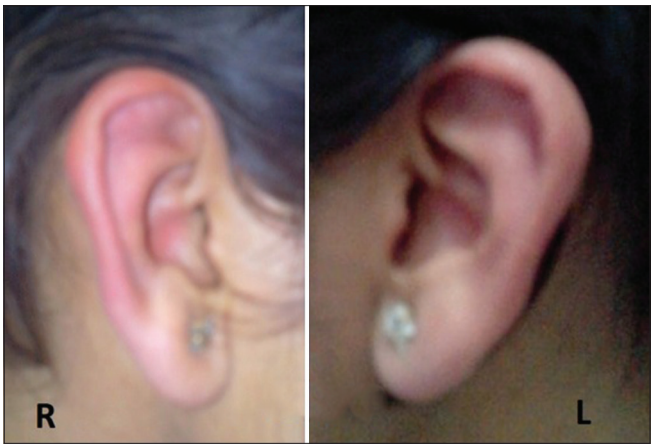

Figure 3: Case 3: Bilateral red ears during paroxysms of pain in R] Right ear L] Left ear

RES has been reported in association with migraine and temporomandibular joint dysfunction.[2] Raieli *et al*. suggested that RES associated with migraine can occur during its prodrome (as an aura of migraine) or during the period of headache and this association could be explained by trigeminovascular activation.[4,6] Synchronous ear vasodilation with facial vasoconstriction could be the reason for RES seen during a migraine attack.[11,12]

RES also has overlap with trigeminal autonomic cephalalgias (TAC) but the main differentiating feature of TAC is extra-auricular symptoms such as lacrimation, conjunctival redness, and nasal blockage which are not seen in RES.[13,14]

Lambru, Miller, and Matharu have proposed the diagnostic criteria for RES [Table 2]. However, there are fallacies in these diagnostic criteria still and some of them can be debated about. For example, the number of attacks required to be present in criteria A has been empirically numbered to be at least 20 attacks and this may not be required. The frequency of attacks may be important while considering whether prophylactic treatment is required or not, however, documented fewer attacks which satisfy other criteria would not negate a diagnosis of RES. The criteria do concede that patients may have fewer attacks and daily attacks may not be required but then these facts about frequency (20 attacks at least) should not be considered in the diagnostic criteria or the criteria should have a lesser number as the minimum required to attack.

A criterion of having the pain unilaterally also may not be required as patients may have bilateral red ears during an attack (as in one of our cases). If unilaterality is not must, then, having the feature as one of the criteria makes the criteria unnecessarily complicated. So laterality may not be mentioned at all in the diagnostic criteria and only 2 of 3 criteria (burning quality/mild-to-moderate intensity and triggered by cutaneous stimuli) may be considered. The same fallacy of describing the pain to be hemicranial in migraine has also been there for many years but the fact that headache can be bilateral in migraine (especially in children) is well-known now. [ICHD III].[15] Similarly, hemifacial spasm or trigeminal neuralgia have been known to have bilateral symptoms in rare cases and so the criteria to have unilateral pain also may not be clinically relevant.

RES can be classified into two types: 1) Primary RES and 2) Secondary RES. Primary type RES occurs more in younger individual and is short-lasting with variable recurrence and most of them have a history of migraine. In contrast, secondary RES occurs in older with female predominance and without a history of migraine or associated with the trigger.[2,7] Secondary RES has been attributed to a number of disorders such as cervical arachnoiditis, traction injury of upper cervical roots, atypical neuralgias, temporomandibular joint dysfunction, or thalamic syndrome.[2]

Table 1: Tabulated summary of different proposed hypotheses for the pathophysiology of red ear syndrome

	Different Hypothesis (Author) [References]	Description of hypothesis	Favoring points of the hypothesis	Pitfalls of the hypothesis
Central Theory	Brainstem trigemino- autonomic circuit dysregulation theory (Goadsby and Lipton)[1,8]	Dysfunction in the regulation of the brainstem connection between facial parasympathetic outflow in response to stimuli exciting the trigeminal nerve	This hypothesis in line with the central hypothesis of paroxysmal hemicranias, cluster headache and SUNA	What leads to the dysfunction in the link between trigeminal and facial nerve is unclear
	Trigemino-Vascular activation theory (Raielli)[1,4]	Trigeminal nerve activation by stimuli lead to the release of vasodilator substances such as substance P, CGRP, and nitric oxide causing pain and redness	Associated of RES with migraine can be explained by this theory	RES occurs due to sympathetic dysregulation and not by parasympathetic activation
Peripheral Theory	Antidromic discharges from C3 Root irritative lesions (Lance)[1,3]	Vasodilatation in the external ear region leads to reddening and pain. This occurs due to C3 root irritation	Relief by local anaesthesia, RES Triggered by temporomandibular joint pathologies	How C fibres get stimulated in primary RES is unclear
	Auricular Erythromelalgia Theory (Kalgaard and Orstavik)[1,9,10]	Combination of vascular misdistribution and sensory and sympathetic nerve dysfunction in the auricular region leads to skin hypoxia on exposure to specific triggers. Such hypoxia leads to neuropathic pain and redness in the auricular region	Clinical similarities to Erythromelalgia	Headache occurring in association with RES is not explained
Unified Theory	Combination of central as well as peripheral dysfunction in a variable manner may be responsible for RES. (We propose this)	Secondary RES (associated with cervical cord or TMJ lesions) may be due to peripheral nerve dysfunction leading to an exaggerated vasodilatory response to nonpainful cutaneous stimuli/triggers. Primary RES, as well as secondary RES in association with thalamic or brainstem pathology, would be a result of central brainstem dysfunction plus abnormal peripheral focal neurovascular bundle supplying the external ear region	This unified theory combines both the central and peripheral theories and can also differentiate the RES into primary and secondary subtypes.	What leads to the brainstem trigemino-facial circuit dysfunction is still unclear

Table 2: Diagnostic criteria for primary RES by Lambru, Miller, and Matharu[1]

A	At least 20 attacks fulfilling criteria B-E
B	Episodes of external ear pain lasting up to 4 h
C	The ear pain has at least two of the following characteristics: Burning quality Unilateral location Mild-to-moderate severity Triggered by cutaneous or thermal stimulation of the ear
D	The ear pain is accompanied by ipsilateral redness of the external ear.
E	Attacks occur with a frequency of more than equal to one per day, although cases with lower frequency may occur.
F	Not attributed to another disorder

RES is a rare, incompletely understood, probably under-recognized headache disorder characterized by short-lasting paroxysmal attacks of reddening of pinna with local pain involving the external ear. More awareness about this disorder, refining its diagnostic criteria, and consideration for inclusion of it in the ICHD classification should be the further steps in understanding and recognizing this rare headache disorder.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Ishani Patel, Devangi Desai[1], Soaham Desai[2]

Intern Doctor, [1]Departments of Medicine, and [2]Neurology, Shree Krishna Hospital, Pramukhswami Medical College, Karamsad, Anand, Gujarat, India

Address for correspondence: Dr. Soaham Desai, Department of Neurology, Shree Krishna Hospital, Pramukhswami Medical College, Karamsad, Anand, Gujarat - 388 325, India.
E-mail: drsoahamdesai@yahoo.com

References

1. Lambru G, Miller S, Matharu MS. The red ear syndrome. J Headache Pain 2013;14:83.
2. Al-Din AS, Mir R, Davey R, Lily O, Ghaus N. Trigeminal cephalgias and facial pain syndromes associated with autonomic dysfunction. Cephalalgia 200;25:605-11.
3. Lance JW. The red ear syndrome. Neurology 1996;47:617-20.
4. Raieli V, Monastero R, Santangelo G, Eliseo GL, Eliseo M, Camarda R. Red ear syndrome and migraine: Report of eight cases. Headache 2002;42:147-51.
5. Boulton P, Purdy RA, Bosch EP, Dodick DW. Primary and secondary red ear syndrome: Implications for treatment. Cephalalgia 2007;27:107-10.
6. Donnet A, Valade D. The red ear syndrome. J Neurol Neurosurg Psychiatry 2004;75:1077.
7. Chan CC, Ghosh S. Red ear syndrome precipitated by a dietary trigger: A case report. J Med Case Rep 2014;8:338.
8. Goadsby PJ, Lipton RB. A review of paroxysmal hemicranias, SUNCT syndrome and other short-lasting headaches with autonomic feature, including new cases. Brain 1997;120:193-209.
9. Kalgaard OM, Clausen OP, Mellbye OJ, Hovig T, Kvernebo K. Nonspecific capillary proliferation and vasculopathy indicate skin hypoxia in erythromelalgia. Arch Dermatol 2011;147:309-14.
10. Ørstavik K, Mørk C, Kvernebo K, Jørum E. Pain in primary erythromelalgia—A neuropathic component? Pain 2004;110:531-8.
11. Raieli V, Pandolfi E, La Vecchia M, Puma D, Calò A, Celauro A, *et al*. The prevalence of allodynia, osmophobia and red ear syndrome in the juvenile headache: Preliminary data. J Headache Pain 2005;6:271-3.
12. Picco PP, D'Alessandro M, Leoni M, Doglio M, Martini A. Primary red ear syndrome associated with cochleo-vestibular symptomatology: A paediatric case report. Cephalalgia 2013;33:1277-80.
13. Ryan S, Wakerley BR, Davies P. Red ear syndrome: A review of all published cases (1996–2010). Cephalalgia 2013;33:190-201.
14. Young WB, Peres MF, Rozen TD. Modular headache theory. Cephalalgia 2001;21:842-9.
15. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia 2018;38:1-211.

Submitted: 04-Jan-2020 **Revised:** 21-Jan-2020 **Accepted:** 08-Feb-2020
Published: 07-Apr-2020

DOI: 10.4103/aian.AIAN_1_20