

## Red ear syndrome: Three new pediatric cases

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### Abstract

Red ear syndrome (RES) is characterized by paroxysmal burning sensation, pain, edema and reddening of unilateral or bilateral ear lobe, usually provoked by different triggers. The exact prevalence in the pediatric age group is unknown but pediatric cases reported in the literature are very small in number. Etiology and pathomechanism of RES still remain controversial. Several therapeutic approaches have been tried with heterogeneous clinical response. We report three new pediatric cases, one of which is the youngest case ever reported in the literature.

**Keywords:** Auricula, erythromelalgia, red ear syndrome, migraine, pediatric, case report

### INTRODUCTION

Red ear syndrome (RES) is a rare neurological syndrome that was first described in 1994 as flushing and a sensation of tightness or pain in one ear lobe in three patients.<sup>1</sup> With the increase in case reports in the literature, bilateral involvement and triggering by touch or changes in ambient temperature have also been described. To date, only 19 pediatric cases have been reported. RES is classified into two groups as primary (idiopathic) RES associated with primary headache syndromes and secondary RES due to upper cervical spine pathologies and temporomandibular joint dysfunction (TMJD).<sup>2</sup> For the time being, primary RES is an diagnosis of exclusion with proposed but not universally

accepted diagnostic criteria (Table 1).<sup>3</sup> Differential diagnosis includes allergic reactions, contact dermatitis, chilblains (associated with cold exposure), relapsing polychondritis (autoimmune condition characterized by recurrent inflammation of the cartilage, but sparing the earlobe unlike RES), and infectious perichondritis caused by *Pseudomonas aeruginosa*, often following a local trauma. In all these, the anamnesis, physical examination, and sometimes basic laboratory tests are enough to rule out the differential diagnosis.<sup>4,5</sup> The disease pathomechanism is not fully understood, resulting in no clear consensus on the treatment.<sup>2</sup> Here, three new pediatric cases with RES are reported.

**Table 1: Proposed diagnostic criteria for primary red ear syndrome by Lambru *et al.*<sup>3</sup>**

A	At least 20 attacks fulfilling criteria B-E
B	Episodes of external ear pain lasting up to 4 hours.
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 $\geq 1$ per day, although cases with lower frequency may occur.
F	Not attributed to another disorder.

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## CASE REPORTS

### Patient 1

An eight-year-old male patient presented with a pain, burning sensation, and redness in both ears that lasted between 15 minutes and 1 hour and started two years ago (Figure 1). His attacks were about 10 times a day, more often in the evenings. The patient had a history of hematemesis at the age of 11 months, leading to diagnosis of portal hypertension and a Rex shunt was performed six years ago. The patient was currently not using any medication other than pantoprazole and he felt relief by washing his ears with cold water. The patient did not complain of headache, but his older brother suffered from migraine.



Figure 1. Red and slightly swollen right ear of Patient 1

### Patient 2

A corrected 15-month-old (birth at 31st gestational week) male patient presented with routine post-delivery follow-up. The patient, whose neurodevelopment was normal for the corrected age, complained of redness, swelling and pain in both auricles every 15 days for four months (Figure 2). These attacks lasted approximately two days. His mother was under follow-up for migraine.

### Patient 3

A nine-year-old girl presented with complaints of pain, warmth, swelling and redness in both ears recurring every 10-20 days for one year. The attacks lasted 1-2 days; no triggering factor or responding to local steroid and ice application. There was also no history of headache, family history of migraine or any other headache syndromes.

All three patients shared common features such as sparing the hands and feet, no history of triggering factors, normal neurologic / otorhinolaryngologic examinations including audiologic tests, and normal routine blood tests, brain and cervical MRIs. Allergic reactions, contact dermatitis, chilblains and infectious perichondritis were excluded with the absence of a history of drug use, cold exposure, or trauma. Clinical features like onset at pediatric age, sparing the ear lobe and absence of nasal chondritis, auricular, laryngotracheal, cardiopulmonary and dermatological involvements exclude the diagnosis of relapsing polychondritis. Propranolol



Figure 2. Red and swollen right and left ears of Patient 2

was recommended to Patient 1 whose attacks did not restrict his daily activities but occurred frequently. The family did not consent to the treatment. The parents of Patient 2 and 3, whose attack frequencies were relatively low, were informed about the treatment options, if the attack frequency and/or severity increase. The families decided not to take drug treatment. All the three patients were still being followed up. Demographic and clinical features of these patients are shown in Table 2.

## DISCUSSION

RES is a very rare disorder with only 19 pediatric cases reported up to the present.<sup>2</sup> In this article, we reported three new cases. While the mean age of pediatric cases has been reported as 8.7 (4-14 years)<sup>2</sup>, the ages of our cases were 15 months, eight and nine years. Patient 2, who was 15 months old, was the youngest patient ever reported. Of the pediatric cases reported, 89.4% were male.<sup>2</sup> Two of our three cases were also male. In the literature, there is no significant difference in pediatric RES cases in terms of unilateral (n: 10, 52.6%) or bilateral (n: 9, 47.4%) auricular involvement.<sup>2</sup> In our cases, bilateral involvement was seen. Episodes can occur spontaneously or be triggered, most frequently by rubbing or touching the ear, heat or cold, trauma, brushing of the hair, neck movements or exertion, chewing.<sup>3,6-8</sup> No trigger factor was identified in our three cases.

The pathomechanism theories of RES, which have been the subject of widespread debate and still remain unclear, can be divided into central, involving trigeminal autonomic reflex (TAR)- a brainstem connection between the trigeminal nerve and the facial parasympathetic outflow providing parasympathetic control to facial vascularization- dysfunction, and peripheral, including cervico-spinal-temporomandibular dysfunction.<sup>3</sup> Although TAR has been proposed as a possible explication of the link between migraine and RES<sup>9,10</sup>, vasodilation of only the ears, not the entire face has been explained by assimilation of RES to an “auriculo-autonomic cephalalgia” where link between cervical sensitive afferents and parasympathetic brainstem efferents result in a TAR equivalent.<sup>11</sup>

Primary RES is typically seen in children or young adults, while secondary RES is more common in adults.<sup>2</sup> Migraine and nonspecific headache were found in 57.9% and 5.2% of pediatric RES cases, respectively.<sup>4,6,12-15</sup> However, family history of migraine in primary RES

**Table 2: Demographic and clinical features of the patients**

	Gender	Age (months)	Age of symptom onset (months)	Burning sensation	Pain	Redness	Edema	Frequency	Duration (pain)	Duration (other symptoms)	Trigger	Headache	Family history of headache	Comorbidity	Medication
Patient 1	Male	96	72	(+)	(-)	(+)	(-)	10 times/day	2 hours	15 minutes-1 hour	None	(-)	(+)	Portal hypertension, horseshoe kidney	Pantoprazole
Patient 2	Male	15	11	(-)	(+)	(+)	(+)	Every 15 days	30 minutes-1 hour	6-48 hours	None	(-)	(+)	Prematurity	None
Patient 3	Female	108	96	(-)	(+)	(+)	(+)	Every 15-20 days	2 hours	24-48 hours	None	(-)	(-)	None	None

patients has not been mentioned in the literature but considering the genetic nature of migraine, it can be concluded that migraine may be common in families of primary RES patients. Neither migraine nor nonspecific headache was reported in our cases, but in two patients, a family history of migraine was present, consistent with this hypothesis.

Diverse secondary causes of RES have been reported in the literature, mostly in the adulthood. These etiologies are classified into two main groups, each occurring in 44% of cases: TMJD and upper cervical spine lesions including cervical spondylosis, congenital cervical abnormalities, and neural foramen narrowing. Other rare causes include thalamic syndrome and herpes zoster infection.<sup>3,6,16-18</sup> Neuropsychiatric systemic lupus erythematosus and TMJD have been reported in pediatric secondary RES cases.<sup>6,19</sup> Secondary causes predisposing to RES were not found in our cases. Considering the association of secondary RES with upper cervical spine pathology and TMJD, cervical MRI should be performed.<sup>3</sup> Brain and cervical MRIs of our cases were all normal. Evaluation of the indomethacin response in patients with attacks more than once a day is proposed to rule out chronic paroxysmal hemicrania, which may have RES-like presentation due to unilateral and short-duration headache, episodes of auricular redness/pain.<sup>3</sup> This trial was not performed since our cases did not have headaches, were in the pediatric age group, and had bilateral manifestations.

Diagnostic criteria for the primary RES have been proposed<sup>3</sup> but have not yet been included in the International Classification of Headache Disorders due to the controversies in the pathogenesis.<sup>20</sup> These criteria, which were created with inclusion of both pediatric and adult patients, were mostly met by our cases in terms of the number, frequency and characteristics of attacks. Although Case 2 could not verbally express the presence of pain due to the young age, we interpreted the 30-60 minutes of restlessness and crying in his attacks as pain. In aspects of etiology and pathomechanism, RES could not be fully elucidated, leading to the imprecision for treatment options. Preventive strategies such as avoiding extreme temperatures, trauma and/or other triggering factors are proffered. Ice application and cooling sprays were used for symptomatic relief<sup>2</sup>, but caution is necessary as it is also a possible trigger. Although some prophylactic migraine treatments have been reported to have mild to moderate benefits especially

in patients with migraine-related episodes, RES is generally regarded to be refractory to medical treatments. Nimodipine, carbamazepine, verapamil, flunarizine, propranolol, cetirizine and gabapentin were found to have varying frequency of beneficial effects in children.<sup>2</sup> Propranolol treatment was recommended immediately to Patient 1 with frequent attacks, and to Patient 2 and 3 when there was frequent attacks, but in none of whom, the parent consented to receive the medical treatment.

In conclusion, we reported three new primary RES cases in one of whom was the youngest case ever reported. Although the pathophysiology and treatment options of RES are controversial, the place of neuroimaging in the differential diagnosis process is well accepted.

## DISCLOSURE

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Conflicts of Interest: None.

## REFERENCES

1. Lance JW. The mystery of one red ear. *Clin Exp Neurol* 1994;31:13-8.
2. Cinats AK, Haber RM. Pediatric red ear syndrome: A case report of an erythromelalgia type and review of the literature. *Pediatr Dermatol* 2019;36(5):686-9. doi:10.1111/pde.13911.
3. Lambro G, Miller S, Matharu MS. The red ear syndrome. *J Headache Pain* 2013;14(1):83. doi:10.1186/1129-2377-14-83.
4. Hernández CR, Loza SM, López RL, Del Mar García Romero M. Red ear syndrome in a pediatric patient. *Pediatr Emerg Care* 2018;34(10):e199-e200. doi:10.1097/PEC.0000000000001633.
5. van der Hulst AE, Sukhai RN. A red ear. *J Pediatr* 2009;154(4):624. doi:10.1016/j.jpeds.2008.10.014.
6. Lance JW. The red ear syndrome. *Neurology* 1996;47(3):617-20. doi:10.1212/wnl.47.3.617.
7. Brill TJ, Funk B, Thaçi D, Kaufmann R. Red ear syndrome and auricular erythromelalgia: the same condition? *Clin Exp Dermatol* 2009;34(8):e626-8. doi:10.1111/j.1365-2230.2009.03342.x.
8. Selekler M, Kutlu A, Uçar S, Almaç A. Immediate response to greater auricular nerve blockade in red ear syndrome. *Cephalalgia*. 2009;29(4):478-9. doi:10.1111/j.1468-2982.2008.01756.x.
9. Raieli V, Compagno A, Brighina F, et al. Prevalence of red ear syndrome in juvenile primary headaches. *Cephalalgia*. 2011;31(5):597-602. doi:10.1177/0333102410388437.
10. Raieli V, Giordano G, Spitaleri C, et al. Migraine and cranial autonomic symptoms in children and adolescents: a clinical study. *J Child Neurol* 2015;30(2):182-6. doi:10.1177/0883073814535494.
11. Evans RW, Lance JW. The red ear syndrome: an auriculo-

- autonomic cephalgia. *Headache* 2004;44(8):835-6. doi:10.1111/j.1526-4610.2004.04157.x.
12. Raieli V, Monastero R, Santangelo G, Eliseo GL, Eliseo M, Camarda R. Red ear syndrome and migraine: report of eight cases. *Headache* 2002;42(2):147-51. doi:10.1046/j.1526-4610.2002.02033.x.
  13. 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(15):1277-80. doi:10.1177/0333102413493527.
  14. Coleman J, Ream MA. A boy with red ears. *Semin Pediatr Neurol* 2018;26:92-3. doi:10.1016/j.spen.2017.03.015.
  15. La Mantia I, Andaloro C. An unusual headache: Red ear syndrome. *Eurasian J Med* 2018;50(2):139. doi:10.5152/eurasianjmed.2018.17344.
  16. Boulton P, Purdy RA, Bosch EP, Dodick DW. Primary and secondary red ear syndrome: implications for treatment. *Cephalalgia* 2007;27(2):107-10. doi:10.1111/j.1468-2982.2007.01270.x.
  17. Arjona A, Serrano-Castro PJ, Fernández-Romero E, Rubí J. The red ear syndrome: five new cases. *Cephalalgia* 2005;25(6):479-80. doi:10.1111/j.1468-2982.2005.00902.x.
  18. Al-Din AS, Mir R, Davey R, Lily O, Ghaus N. Trigeminal cephalgias and facial pain syndromes associated with autonomic dysfunction. *Cephalalgia* 2005;25(8):605-11. doi:10.1111/j.1468-2982.2005.00935.x.
  19. Lee HS, Bae SC, Uhm WS, Jun JB, Lee IH, Kim SY. Red ear(s) syndrome associated with child neuropsychiatric systemic lupus erythematosus. *Lupus* 2000;9(4):301-3. doi:10.1191/096120300680198908.
  20. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. *Cephalalgia* 2018;38(1):1-211. doi:10.1177/0333102417738202.