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Review Article

Red ear syndrome: Literature review and a pediatric case report☆

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ABSTRACT

Red ear syndrome (RES) is characterized by recurrent unilateral or bilateral painful attacks of the external ear, accompanied by ear redness, burning, or warmth. Proposed etiologies of this rare condition include dysregulation of sympathetic outflow, upper cervical pathology, glossopharyngeal and trigeminal neuralgia, TMJ dysfunction, thalamic syndrome, and primary headache syndromes. Idiopathic cases also exist in the literature. Pediatric cases are particularly rare and more commonly associated with migraine. Given the various potential etiologies, no single treatment is effective in all cases. This paper summarizes the current understanding and management of RES, and describes a case of idiopathic pediatric RES.

Contents

1. Introduction

Red ear syndrome (RES) was initially described in the literature by Lance in 1994 [1,2]. The condition involves recurrent attacks of unilateral or bilateral erythema of the ear, accompanied by burning, pain, or increased skin temperature of the affected ear [3]. The attacks occurred with varying duration and frequency. Symptoms are often precipitated by touch or changes in temperature, although in some cases they are spontaneous [3]. The etiology of the syndrome is still not well established, although multiple publications have described migraine or other neurological disorders as a possible linkage [2–11]. The treatment protocol varies and is usually symptomatically driven. We report a rare case of pediatric RES and review the current literature on pediatric and adult RES.

2. Case report

A 5-year old boy was referred to the Department of Otolaryngology at McMaster University with a 3-year history of intermittent erythema of ears. Although no precipitating factors could be identified, the patient usually presented with unilateral or bilateral erythema of ears, accompanied by occasional ear swelling, discomfort and warmth of the skin (Figs. 1–3). The symptoms would last for 1 h and then resolve spontaneously. Initially, the episodes were reported to occur approximately each week. In follow up appointments, the reported frequency had decreased to 2–3 times each month. He had a history of exaggerated local reaction with erythema and urticaria with wasp and spider bites, however there was no apparent association with the presenting symptoms of RES. He denied otalgia and any other otologic symptoms and had no previous history of migraine or associated headaches. There is no family history.

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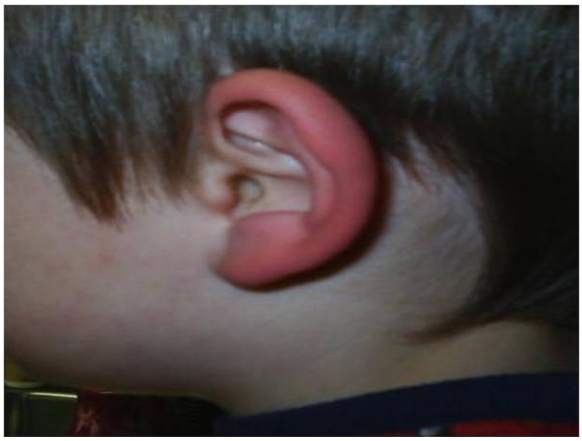

Fig. 1. Left-sided RES.

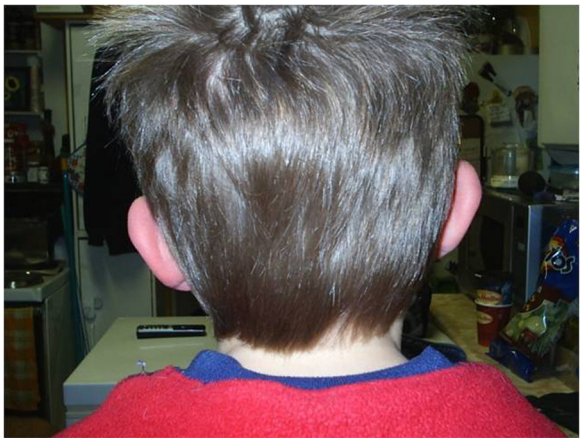

Fig. 3. Bilateral RES.

On examination, he was found to have normal ears bilaterally with normal tympanic membranes on microscopy. The remainder of the head and neck exam was unremarkable. Audiogram revealed normal hearing bilaterally. Allergic workup to identify a possible trigger was also negative. The patient showed minimal improvement with a treatment regimen of two weeks of Cetirizine. The patient continues to have these attacks intermittently.

3. Discussion

Red ear syndrome was initially described by Lance in twelve patients as a syndrome of recurrent attacks of unilateral ear pain or burning sensation with erythema of the ear [1,2]. The episodes vary in duration and frequency. RES can often affect both ears in isolation or simultaneously. The frequency of laterality is not known. Although multiple triggers have been described in the literature, some cases do occur spontaneously and therefore the exact etiology is still debated.

Based on an extensive search in scientific databases, fifty four adult cases of RES were identified. A summary of the adult cases of RES currently published in the literature is presented in Table 1 [2,4–7,10,11,13,15–17,23–27].

Twelve cases of RES have been described in detail in the pediatric population [2,7,9,21,22]. Raieli reports an additional sixteen cases in the pediatric population with minimal details [20]. A summary of the pediatric cases of RES currently published in the literature is presented in Table 2 [2,7,9,20–23].

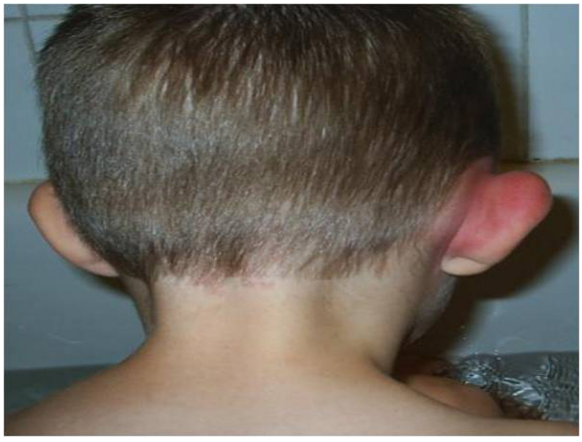

Fig. 2. Right-sided RES.

Current literature describes cases with various proposed etiologies. RES has been associated with upper cervical root pathology, glossopharyngeal and trigeminal neuralgia, TMJ dysfunction, thalamic syndrome, and exercise-induced compression of cerebellar tonsils [2–5]. A number of cases also suggest a relationship to primary headache disorders such as chronic paroxysmal hemicranias (CPH), hemicranias continua, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome, and most commonly with migraine [6–11]. RES has also been described in a number of cases with unknown etiology [2].

Theories surrounding the etiology of RES involve three nerves in particular. The great auricular nerve, arises from the second and third cervical roots, and provides innervation to the earlobe. The greater occipital nerve arises from the second spinal nerve, and innervates the posterior part of the scalp and the skin over the ear and parotid gland. The auriculotemporal nerve, a branch of the mandibular division (V3) of the trigeminal nerve, innervates the TMJ and anterosuperior aspect of the ear.

Lance proposed the term "auriculo-autonomic cephalgia", a potential mechanism which suggests that irritation of the C3 root or great auricular nerve leads to antidromic release of vasodilator peptides from afferent nerve terminals [2]. Goadsby and Lipton proposed the term "cervico-autonomic reflex" to imply a functional connection between cervical somatic afferents and parasympathetic brainstem efferents [12]. They suggested that stimulation of the greater occipital nerve also led to local release of vasodilator peptides [12]. Durham et al. suggested that the erythema may be secondary to inhibition of sympathetic vasoconstriction or activation of parasympathetic vasodilator fibers by the trigeminal autonomic reflex [13]. Activation or irritation of the auriculotemporal nerve causes pain, as well as stimulation of facial parasympathetic nerves which cause erythema [13].

According to Lance, cases of glossopharyngeal neuralgia may be explained by the fact that pain afferents from the glossopharyngeal nerve join the spinal tract of the trigeminal nerve which converges with third cervical root afferents on second-order neurons in the upper cervical cord [2]. Lance also suggests that TMJ cases might depend upon a local axon reflex, possibly due to abnormal discharge in C-fibers (peripheral nerves of the somatic sensory system) responding to stimuli [2].

Recently other authors have suggested that RES may be part of the spectrum of trigeminal autonomic cephalgias (TACs) due to the association of several cases with primary headache disorders [5,7,10–17]. Goadsby and Lipton proposed the term TAC to encompass a variety of short-lasting primary headache syndromes which are associated with autonomic activation [12]. There