Red Ear Syndrome

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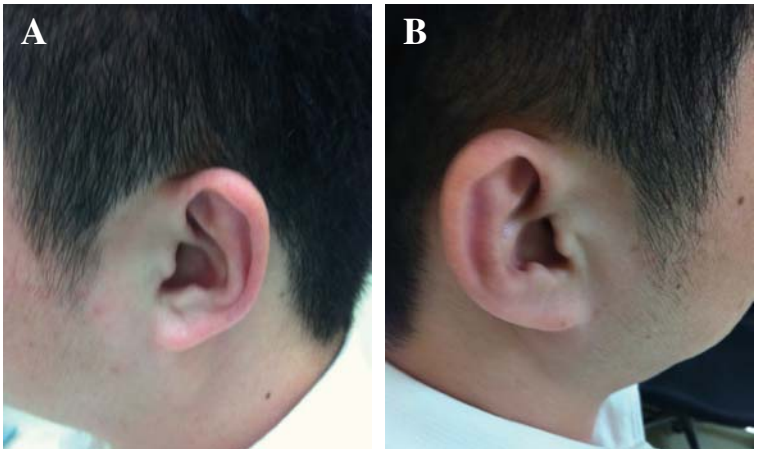

Figure 1. Erythema of left (A) and right (B) ears. The left ear seemed more severe than right ear e.g. the earlobe was involved only in the left side.

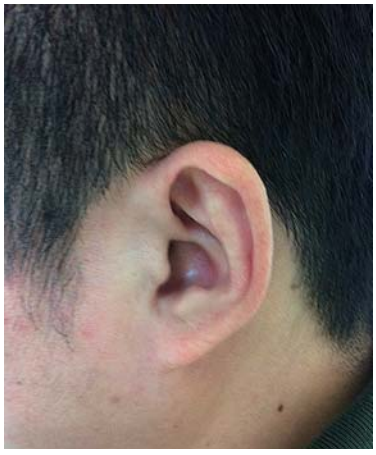

Figure 2. Regression of the erythema 2 days later.

An otherwise healthy 39-year-old man, who denied history of migraine or chronic headache, presented with recurrent left ear pain. The first attack was several months ago, lasting for seconds only. The second attack persisted for 20 minutes, followed by the third episode several hours later. The patient attributed the last attack to be triggered by a violent movement of jaw while he opened his mouth. It lasted for about 2 hours this time which drove him to seek medical help immediately. The patient described the pain as burning-like and the intensity was 8–9 on a 0–10 numerical rating scale. The pain got worse with the ear being touched. Physical examination showed erythema of not only left but also right ear (Figure 1). Neurological examination was unremarkable. No evidence of cranial autonomic involvement were noted, such as tearing, conjunctival injection, rhinorrhea, nasal congestion, forehead sweating, ect. He was given symptomatic treatments with non-steroidal anti-inflammatory drug (NSAID). Two days later, he came back without further attacks and his ears were normal (Figure 2). In February 2014, about 30 months after his first attack, we contacted him by telephone. The patient still had similar attacks with the frequency of once every 1–2 months and the duration of several hours. Except the pain, he denied any neurological complications or sequelae. Thus, he did not ask for further investigation or treatment.

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In this report, we described a patient with red ear syndrome (RES). RES is a rare headache first characterized in 1996[1]. There are approximately 100 cases in the medical literature[2]. RES is a chronic disorder with episodic manifestations of ear pain and erythema. Our patient had left ear pain but both ears erythema. The presence of ear discoloration without pain had been reported[3]. The character of pain is typically burning-like, of moderate intensity. The frequency ranges from once a month to 20 times per day and the duration varies from seconds to 4 hours in most cases. About 62% of RES patients have unilateral involvement, followed by bilateral (31%) and both[2]. Most patients have both spontaneous and triggered attacks. Triggers include heat, rubbing the ear, touching the surrounding skin or hair, physical exercise, neck and jaw movement, etc. The mechanism of RES remains unclear. The dysfunction of cervical spinal nerves (especially C3) and the dysregulation of brainstem trigemino-autonomic circuits have been speculated. Except the primary RES, about a quarter of RES patients have secondary causes[2]. The most common conditions are upper cervical spine lesions (spondylosis, injury, Chiari I malformation, etc.), temporomandibular joint dysfunction, cranial and cervical neuralgia and so on[2]. RES is in general considered as refractory. Medications for neuralgia and headache have been tried, such as NSAIDs, gabapentin, amitripyline, flunarizine, propranolol, and greater auricular nerve blockade[2].

Key Words: chronic disorder with episodic manifestations, headache, neuralgia, red ear syndrome

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REFERENCE

1. Lance JW. The red ear syndrome. Neurology 1996;47:613-620.
2. Lambru G, Miller S, Matharu MS. The red ear syndrome. J Headache Pain 2013;14:83.
3. Hirsch AR. Red ear syndrome. Neurology 1997;49:1190.