We describe eight idiopathic cases of red ear syndrome in seven children and one adult. All were migraineurs with a history of paroxysmally painful and red ear, unilateral or alternating, in isolation or associated with migraine attacks. The reported duration of these episodes varied from 30 minutes to 1 hour. Neurologic examination, brain MRI and CT scans, and x-rays of the cervical spine were normal. The close temporal relationship between the “red ear episodes” and migraine attacks suggests an association between the two conditions.

Key words: red ear syndrome, migraine, nervus trigeminus

Abbreviations: RES red ear syndrome, RE red ear

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frequency of RE episodes and migraine attacks, the patient was started on therapy with nimodipine and subsequently experienced a reduction in the frequency and intensity of both disturbances.

**Patient 2.**—An 8-year-old boy reported a history of migraine without aura. Forty days prior to presentation, he was involved in a motor vehicle accident and sustained a minor closed head injury. Subsequent to this, his headaches increased in frequency (at least two to three weekly attacks) and duration (from 2 to 4 hours to 4 to 24 hours). In addition, his previously typical frontotemporal pain now spread to the ipsilateral ear, which would become red and hot and painful to touch; the RE episodes usually lasted about 30 minutes and occurred three to four within a month.

Neurologic examination during these episodes was normal; there were no trigger points nor temporomandibular joint dysfunction. Electroencephalography and CT of the brain and cervical spine were normal.

**Patient 3.**—A 9-year-old boy reported a history of migraine without aura that extended over several years. At aged 11 years, he began to experience about six episodes per month of burning painful RE, usually on the left, lasting 1 hour, and occurring especially during the night. The episodes were not related to exercise or neck movement, but at times were associated with acute migraine attacks.

Neurologic examination was normal, as were CT of the brain and x-rays of the cervical spine.

**Patient 4.**—A 12-year-old girl reported episodes of migraine without aura (IHS 1.1) since aged 8. In the last year, she had begun to experience an episodic burning painful sensation usually localized to her left earlobe. The pain would build inexorably and in about 5 to 10 minutes become so severe as to require bed rest. These RE episodes had a duration of about 30 to 45 minutes, a frequency of two to three a month, and were not related to exercise or stress; on five occasions, they were associated with acute migraine attacks.

Neurologic examination, CT of the brain, and x-rays of the cervical spine were normal.

**Patient 5.**—A 5-year-old boy reported a 6-month history of migraine without aura. He had an average of two attacks per month lasting 2 hours (IHS 1.1). Migraine attacks often were associated with episodes of painful burning in his ears; the ear symptoms would alternatingly lateralize or be bilateral, last less than 30 minutes, and occur two to four times a month; twice they occurred independent of a migraine attack.

Neurologic examination during RES episodes was normal, as were an EEG, a CT scan of the brain, and x-rays of the cervical spine.

**Patient 6.**—A boy, aged 13, suffered episodes of migraine without aura for 3 years. For the last year, he had experienced episodes of painful burning RE, usually on the right. These episodes lasted 30 minutes, occurred four times per month, seemed to be triggered by school-related stress, and six or seven times occurred in association with an acute migraine attack.

Neurologic examination, CT of the brain, and x-rays of the cervical spine were normal.

**Patient 7.**—An 11-year-old boy reported a 2-year history of migraine without aura. During the previous 8 months, he had experienced episodes of painful burning in his right ear, lasting less than 30 minutes, and occurring three to five times per month; approximately half of these episodes were immediately followed by an acute migraine attack.

Neurologic examination, EEG, CT of the brain, and x-rays of the cervical spine were normal. The patient began prophylactic treatment with flunarizine and subsequently reported a reduction in the frequency of both migraine attacks and RES episodes.

**Patient 8.**—A 36-year-old woman reported episodes of migraine with and without aura since aged 15. Her headache attacks, at times, were preceded by a visual aura, primarily with negative features (typically scotomas) lasting 20 minutes. She had an average of one weekly attack, lasting 4 to 48 hours. Over the few years prior to evaluation, the migraine attacks often were associated with a burning and painful sensation in the left earlobe that resolved at the end of migraine attacks. The RE episodes, usually of 30 to 180 minutes’ duration and a frequency of one to two a month, were not associated with stress or abnormal neck movements.

Neurologic examination, MRI of the brain, and CT of the cervical spine were normal.
The clinical characteristics of these cases are shown in the Table.

**RESULTS AND COMMENTS**

These eight patients afflicted with migraine also suffer from RES. In this group, the RES episodes usually occurred in close temporal relationship with migraine attacks, but also could occur in isolation. Even with the latter, however, the RES episodes appeared to retain some migrainous characteristics, such as the alternating, unilateral (rarely bilateral) location of the pain.

Red ear syndrome was first described by Lance as acute episodes of unilateral ear pain, usually described as “burning,” with associated changes in cutaneous color and an increase in ear temperature. In 10 of the 12 patients initially reported by Lance, the RES was attributed to such diverse etiologies as cervical arachnoiditis, cervical spondylosis, atypical glossopharyngeal neuralgia, third cervical root neuralgia, temporomandibular joint dysfunction, thalamic syndrome, and a possible traction injury of the upper cervical roots. In 2 patients, both females, Lance did not find any plausible etiology and so classified these cases as “idiopathic red ear.” Neither patient clearly had migraine as well as RES.

Others recently have reported two patients with chronic paroxysmal hemicrania (CPH) and unilateral otalgia with the typical features of RES, and one patient with extratrigeminal episodic paroxysmal hemicrania (EPH) and RES, all responsive to indomethacin. These authors proposed that this symptom complex be included with the so-called trigemino-autonomic cephalalgias (TACs) recently described by Goadsby and Lipton (ie, short-lasting unilateral neuralgiform headache with conjunctival injection and tearing [SUNCT], hemicrania continua, CPH, EPH, and cluster headache). Goadsby and Lipton have postulated that both paroxysmal hemicrania and RES might be due to a common pathogenetic mechanism based on a brain stem connection between the trigeminal nerve and the facial parasympathetic outflow (ie, a trigemino-autonomic reflex).
Red ear episodes, then, could be mediated by a cervicovascular reflex due to either an upper cervical disorder, or directly by trigemino-autonomic stimulation via the auriculotemporal nerve, a branch of V3, that supplies the anterosuperior aspect of the ear. If this hypothesis is valid, the association between headache attacks and RE episodes in the idiopathic cases might be due to an intrinsic disregulation of the brain stem trigemino-autonomic circuits rather than a local axon reflex, as suggested by Lance.

In our reported RES cases, all but one (case 2) showed a clear association with migraine attacks, and in that patient the initiation of RES following head injury suggests a possible secondary origin from upper cervical irritation. Even so, the RES in this patient consistently occurred ipsilateral to the headache and was temporally related to migraine attacks, leading us to believe that the head injury was a precipitating factor but not the primary cause of his RES. We consequently consider all of our cases presented here to be “idiopathic.”

Unlike Lance’s idiopathic cases, however, our patients showed RE episodes with some typical features of migraine. First, these episodes were unilateral or alternating in side location (or both) and sometimes bilateral. Secondly, they were often temporally associated with a migraine episode. Lastly, they were not triggered by thermal or mechanical stimuli and started in the absence of any illness, and in one case (No. 3) during the night, as frequently happens for migraine attacks.

In our patients, the absence of a functional or anatomical cervical disorder and the absence of local mechanical factors as RE episode precipitants mitigate against cervical disorder as the pathophysiologic generator of their RE episodes. Their association with migraine, the alternative unilaterality common with the episodes, and their duration suggest instead that the RES episodes experienced by our patients were linked directly to migraine. This hypothesis is strengthened by Hirsch’s recent report of unilateral and bilateral RES episodes occurring in patients with “vascular” headaches. Current theories of migraine biogenesis generally invoke direct activation of the trigeminovascular system or secondary stimulation of the trigeminovascular reflex consequent to activation of certain cortical, subcortical, or brain stem structures. Whether primary or secondary, trigeminovascular activation may produce extracerebral vasodilation via the release of vasodilator substances (eg, nitric oxide, substance P, CGRP), and such extracranial vasodilation may underlie the symptoms of migraine-associated RES.

That trigeminovascular activation may produce pain that extends beyond the trigeminal domain may reflect overlap between neurons of the trigeminal nucleus caudalis and neurons located within the C1 and C2 dorsal horns of the upper cervical spinal cord. This anatomical convergence long has been considered a possible explanation for the neck pain that is prominent in some cases of migraine and may, in fact, paroxysmally occur independent of any migrainous headache and yet respond to migraine-specific therapies. If the analogy is valid, we similarly could consider the RE episodes of our patients that were not temporally linked to migraine to represent “atypical” (ie, acephalgic) migraine attacks.

Another potential pathophysiological mechanism for RES deserves mention. The trigeminal nerve directly innervates the anterior portion of the earlobe and also provides the sensory innervation of the external carotid artery, which, in turn, supplies blood to the ear. Theoretically, then, the trigeminal nuclei, activated by any trigger, could directly produce an antidromic discharge along the trigeminal sensory fibers innervating the ear, promoting release of vasodilator and pro-nociceptive substances and provoking the RE attacks.

In conclusion, RES should be considered a potential accompaniment to migraine. Rarely, RE symptoms conceivably may represent the only clinical manifestations of an acute migraine attack. Further studies are needed to confirm whether and why RE associated with migraine does occur more commonly in childhood, whereas the secondary forms prevail in later years.

REFERENCES