Expert Opinion

The Red Ear Syndrome: An Auriculo-Autonomic Cephalgia

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"The mystery of one red ear" might have been the title of a Sherlock Holmes novel by Dr. Doyle rather than a real life medical mystery uncovered and solved by a neurological sleuth, Prof. Lance.

CLINICAL HISTORY

A 54-year-old white female was seen with a 10-year history of episodes of a burning sensation of the left ear. The episodes are preceded by nausea and a hot feeling for about 15 seconds and then the left ear becomes visibly red for an average of about 1 hour, with a range from about 30 minutes to 2 hours. About once every 2 years, she would have a flurry of episodes occurring over about a 1-month period during which she would average about five episodes with a range of 1 to 6.

There was also an 18-year history of migraine without aura occurring about once a year. At the age of 36 years, she developed left-sided pulsatile tinnitus. A cerebral arteriogram revealed a proximal left internal carotid artery occlusion of uncertain etiology after extensive testing. An MRI scan at the age of 45 years was normal. Neurological examination was normal. A carotid ultrasound study demonstrated complete occlusion of the left internal carotid artery and a normal right.

Question.—What is the diagnosis?

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EXPERT COMMENTARY

In 1995, I encountered three patients with the complaint of an intermittent burning pain in one ear accompanied by flushing of the earlobe. I presented my findings to the Annual Scientific Meeting of the Australian Association of Neurologists with the plea to my colleagues "lend me your ears."[1] In response to my request, I was given information about nine other patients with this problem so that I could analyze their case histories and postulate the mechanism. My definitive publication[2] included seven patients with a disturbance of the upper cervical spine, two of whom also had pain of glossopharyngeal distribution. Two patients had temporomandibular joint dysfunction, one had a thalamic syndrome with secondary migrainous features, one developed migraine headaches after her right ear and cheek turned bright red "as though they are on fire" in response to heat, and the 12th patient had the red burning ear response to heat without a migraine headache following.

From this series, I deduced that the condition was caused by an antidromic discharge of nerve impulses in the third cervical root and greater auricular nerve in response to some local pain—producing lesion in the upper neck or trigeminal areas of innervation.

In response to this report, Hirsch[3] added the description of five headache patients who described frequent bright red discoloration of the ears independent of headache, not associated with pain. Raieli et al[4] described eight patients, seven children and one adult aged 36 years, with the problem. They all suffered from migraine. The episodes were associated with migraine headache at times but recurred independently on other

occasions. Raieli and his colleagues postulated that this placed the red ear syndrome in the group of trigemino-autonomic mediated by the auriculo-temporal nerve. The problem with this is that the relevant part of the earlobe is innervated by the greater auricular nerve arising from the third cervical root. I would be prepared to settle for the term "auriculo-autonomic cephalgia."

The flushing must be secondary to pain as facial flushing by itself, as in Harlequin syndrome,[5] is not painful. Burning pain is characteristic of small afferent fiber (C fiber) discharge. I suggested that the condition could be caused by antidromic discharge of the impulses in such fibers causing vasodilation in the area to which pain was referred by discharge of vasodilator peptides.

This mechanism is thought to account for the facial flush seen after stabbing pains of tic douloureux or after thermocoagulation of the trigeminal ganglion, when it is confined to the skin areas innervated by the appropriate division or divisions of the trigeminal nerve.[6] Similar flushing is seen overlying the painful parts of the face in cluster headache and other trigemino-autonomic cephalgias such as chronic paroxysmal hemicrania.

The parasympathetic overflow through the sphenopalatine ganglion is responsible for flushing in the nose and cheek and is least apparent in the ear.[7] On the other hand, sympathetic innervation contains both constrictor and vasodilator fibers[8] and blockade of the greater auricular nerve by local anesthetic increases earlobe temperature by more than 7°C.[9] Even after such a block, heating increases temperature still further.[7] Whether ear temperature in the red ear syndrome increases by localized sympathetic dysfunction or antidromic discharge remains uncertain.

In summary, the red ear syndrome can be primary, commonly in association with migraine, or secondary, commonly in association with an upper cervical abnormality. The pain is clearly primary and the localized vasodilator secondary, although the precise mechanism is still obscure.

The patient described in your case history falls into the migraine equivalent category and I do not think that her left internal carotid artery occlusion is relevant to the cause of her red ear syndrome but sounds a cautionary note about the use of β-blockers as vasoconstrictive agents in the treatment of her symptoms.

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