RED EAR SYNDROME – CHRONIC PAIN SYNDROME OF LOW PREVALENCE

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SUMMARY – A case is described of a 72-year-old woman that presented to our outpatient neurology clinic complaining of relapsing attacks of severe pain in and around the left ear and ipsilateral side of the face, accompanied by objectively visible cutaneous erythema with burning sensation throughout the attack. There was no alteration in her neurologic status. Comprehensive tests were done to exclude the possible primary diseases. The findings were within the limits of physiological values. Attempts at treating the pain with various analgesics and a combination of analgesics and first generation anticonvulsants did not prove effective. The pain was slightly relieved with pregabalin in maximum dosage. Because of various potential pathophysiological mechanisms, literature does not indicate any unique potential treatment approach, and some patients are resistant to any kind of therapy.

Key words: *Ear diseases – complications; Ear diseases – physiopathology; Erythema – etiology; Erythema – pathology; Erythema – therapy; Syndrome*

Introduction

Since 1994, when "the mystery of one red ear" was originally described in the literature[1], and 1996, when the term 'red ear syndrome (RES)[2] was coined, about seventy cases have been described. The syndrome is characterized by the attacks of pain of variable length in the area of the auricle and around it, accompanied by autonomous symptoms, of which cutaneous erythema with burning sensation ipsilateral to the pain as the most pronounced one is observable to the patient and the examiner during the attack[3]. The etiology is insufficiently known. It is well-known that it can result from the 3rd cervical root irritation, cervical arachnoiditis, spondylosis of cervical facet joint, temporomandibular joint dysfunction, symptom of glossopharyngeal or trigeminal neuralgia, or part of thalamic syndrome[2], part of trigeminal autonomic cephalgia, and may be, even without obvious structural cause, precipitated by touch, movements, exposure to cold or warmth. It is more common in people suffering from migraine attacks[5].

Case Report

A 72-year-old woman presented to our outpatient neurology clinic for relapsing attacks of burning pain in and around the left ear and ipsilateral side of the face, accompanied by objectively visible redness of the skin with burning sensation throughout the attack (Fig. 1). The pain had been present for three years, becoming more intensive and more frequent in the last year. The patient reported no serious illness in the past, did not suffer from migraine attacks, and was taking a small dose of an ACE inhibitor for hypertension. Comprehensive work-up was done, including hematologic and immunologic tests, brain and cervical spine magnetic resonance imaging in order to exclude the possible

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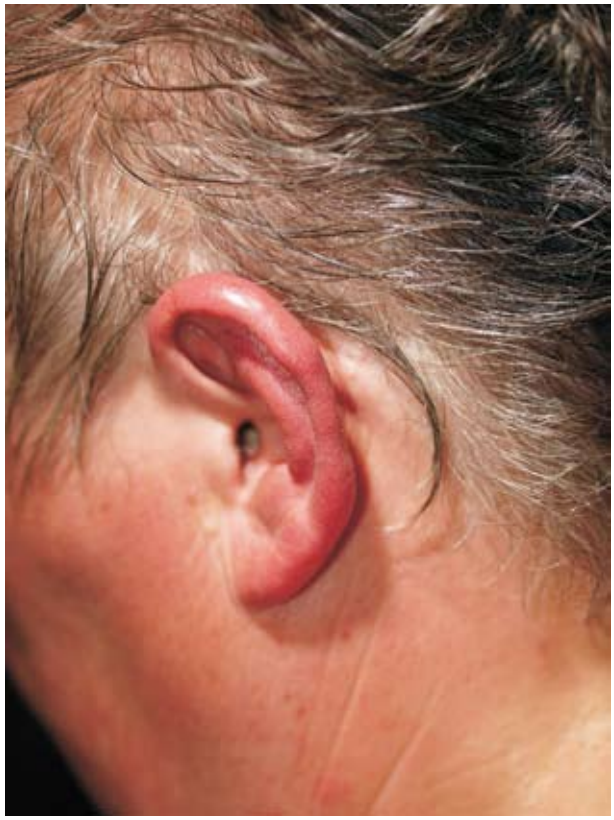

Fig. 1. Red and swollen left ear in a 72-year-old female patient.

underlying disorder. Test results were within the limits of physiological values. Attempts at treatment with acetylsalicylic acid, diclofenac, ketoprofen, ibuprofen, a combination of paracetamol and tramadol, indomethacin, tricyclic antidepressants, a combination of some analgesics and carbamazepine in high dose, or gabapentin with periodical subcutaneous infiltration of local anesthetic, or local application of lidocaine patch as well as transdermal fentanyl showed partial and short-term effectiveness in this case. Pregabalin titrated up to 300 mg twice a day caused slight reduction of pain. The improvement lasted for several months, and then the pain became bilateral. Upon two attacks of more intensive pain, bilateral occipital block of bupivacaine and dexamethasone was applied with acute effectiveness.

Discussion

According to literature data published to date, RES is most frequently unilateral, but it can also be bilateral or change sides. It is characterized by objectively visible erythema of the skin in and around the ear, with burning sensation ipsilateral to the pain and attacks of pain in the area of the auricle and external auditory canal, which can spread to the surrounding anatomical structures. An attack of pain can occur spontaneously, but it can also be caused by chewing, touching, exposure to cold or warmth, as well as by ingestion of spicy food or alcohol. Cases of erythema in and around the ear have been described in which the pain component was not present[6]. In Lance's report, the syndrome was most frequently connected with irritation of C3 root, spondylosis of cervical facet joints or temporomandibular joint dysfunction[1]. Other cases described were related to cervical arachnoiditis, Chiari I malformation, congenital malformations of cervical spine, herpes zoster in the area of C3-C6 dermatomes, and compression of cerebellar tonsils induced by exercise[7]. In Donnet and Valade's work, two types of RES were shown: the first, idiopathic, more frequent in younger population and more often associated with migraine, and the second, more common in adults, associated with disorders of cervical spine[8]. An increased comorbidity was noticed accompanying migraine, chronic paroxysmal hemicrania, SUNCT syndrome[9], and in several cases the etiology of problems was not explained. Erythromelalgia is a similar disorder, which most frequently affects lower extremities, and less frequently upper extremities. Since RES and erythromelalgia have several diagnostic criteria in common, i.e. erythema, elevated temperature, burning pain, pain aggravated by warmth, and pain relieved by coldness[10], there is an opinion that erythromelalgia is a component of RES, and it is possible that RES is the auricular variant of erythromelalgia[11]. A similar syndrome has been described in the area of scrotum, successfully treated with gabapentin[12].

Conclusion

Observation of the cases described so far leads to a conclusion that primary RES can exist as primary trigeminal or cervical autonomic cephalgia, or coexist with other rare trigeminal autonomic cephalgias, thus suggesting a similar pathophysiological basis. The secondary form, predominantly associated with changes in cervical spine joints or in temporomandibular joint, has different pathophysiological basis

and is much more resistant to therapy[13]. Due to various potential pathophysiological mechanisms, there is no unique treatment approach recommended, and many patients are resistant to any kind of therapy. In the literature, there are descriptions of cases in which patient condition was partly improved with the introduction of acetylsalicylic acid, ibuprofen, propranolol, tricyclic antidepressants, flunarizine, surgical section of C3 root, pregabalin and subcutaneous or transdermal application of lidocaine.

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Sažetak

SINDROM CRVENOG UHA – KRONIČNI BOLNI SINDROM NISKE UČESTALOSTI

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Opisuje se slučaj 72-godišnje žene koja se javila neurologu zbog ponavljajućih napadaja bolova jakog intenziteta i različitog trajanja u području lijeve ušne školjke, vanjskog slušnog hodnika i ipsilateralne strane lica praćenih objektivno vidljivim eritemom kože uz osjećaj žarenja tijekom cijelog napadaja. Neurološki status je bio uredan. Provedena je opsežna dijagnostička obrada u svrhu isključenja moguće primarne bolesti. Rezultati učinjenih pretraga bili su u granicama fizioloških vrijednosti. Pokušaji liječenja bola različitim analgeticima i kombinacijom analgetika i antikonvulziva starije generacije nisu se pokazali učinkovitima. Do blažeg smanjenja tegoba došlo je uvođenjem pregabalina u maksimalnoj dozi. Zbog raznolikih potencijalnih patofizioloških mehanizama u literaturi se ne navodi jedinstvena terapijska preporuka, a dio oboljelih otporan je na bilo koji oblik terapije.

Ključne riječi: *Bolesti uha – komplikacije; Bolesti uha – fiziopatologija; Eritem – etiologija; Eritem – patologija; Eritem – terapija; Sindrom*