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Case report

Red ear syndrome – Case report and review of literature



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ABSTRACT

Red ear syndrome is characterized by: paroxysmal, unilateral, recurrent pain, redness and discomfort of the ear lobe accompanied by a burning sensation. The duration and frequency of red ear syndrome attacks is very various and the episodes, usually occur spontaneously. The pathophysiology is still unknown and also there are no medications with approved efficacy. The goal of this brief report is to present a 11-year old girls whose symptoms of red ear syndrome preceded migraine without aura and the signs of redness of the ear occurred in clusters. The occurrence of symptoms of our case may have confirmed the observation that red ear syndrome is associated with primary headaches particularly migraine and cluster headaches. The literature on this case report of pediatric idiopathic red ear syndrome has been reviewed.

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1. Introduction

The first publication describing the red ear syndrome appeared in 1994 by Lance. The same author two years later described a larger group of 12 patients and their common symptoms were termed red ear syndrome (RES). He characterized RES by unilateral painful attacks during which the involved ear becomes red and burning. Ten out of twelve of his patients, had symptoms of RES caused by upper cervical

arachnoiditis, cervical root traction, cervical facet joint spondylosis, glossopharyngeal and trigeminal neuralgia, temporomandibular joint dysfunction and thalamic syndrome. No associated pathology was detected in the other two of his patients [1]. Since then about 90 cases of RES have been published about 60% of the cases are in women and 40% are in men [2,3]. We present a case report of 11-year old girls whose symptoms of idiopathic RES preceded migraine without aura and the signs of redness of the ear occurred in clusters.

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Abbreviations: RES, red ear syndrome; ICHD-III, The International Classification of Headache Disorders, 3 edition beta; CPH, chronic paroxysmal hemicranias; HC, hemicrania continua; SUNCT, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing.

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2. Case report

An 11-year-old girl was admitted to the Chair and Department of Child Neurology in Poznan in February 2011 because of paroxysmal pain of the left ear. The patient's symptoms occurred 2 years before admission to hospital and were characterized by sudden onset of isolated left ear burning pain associated with redness and swelling. These episodes happened in clusters which remained up to 3 months, twice a year, particularly in the winter and in the autumn. During the cluster the episodes appeared usually up to 10 times per day at different times and lasted about 10–20 min. The typical signs of the episode are shown in Fig. 1. The attacks were sometimes induced by the heat. At the beginning RES were not associated with the headache, but one year later the episodes started to be usually associated with a migraine without aura. Before the appearance of RES, the patient did not suffer from migraine. The headaches began simultaneously with RES and were on the same side as RES and lasted three to four hours, and characterized by pulsating quality. Migraines were severe and accompanied by photophobia and phonophobia. The patient's perinatal history and childhood development were reported normal. Patient and her parents did not report family history of RES or headaches, other serious illnesses, head or neck injuries



Fig. 1 – Red and swollen left ear in a 11-year old patient during the episode.

in the past, or taking prescribed or not-prescribed medication. The treatment included administration during the episodes of non-steroidal analgesics (ibuprofen and ketoprofen) without improvement. The neurological and pediatric examination was normal. There was no deviation from the norm in laboratory tests and in the brain MRI with the angio program. The MRI of the cervical spinal cord found a protrusion at the height of the intervertebral tarsus C6-C7 nerve roots without oppression. These changes were not assessed by the radiologist as clinically insignificant. Other causes of RES were excluded and based on the patient's history, clinical patterns, and unremarkable diagnostics, the idiopathic RES was diagnosed. Discharge orders included carbamazepine (100 mg/ twice daily). Follow-up calls revealed that after 2 months the patient stopped carbamazepine because of reported lack of improvement, abdominal pain and throbbing of the heart.

3. Discussion

RES is characterized by: paroxysmal, unilateral (rare bilateral) recurrent pain and redness of the ear lobe discomfort accompanied by burning sensation. In the literature there are descriptions of patients who presented redness of the ear on both sides, although there are cases in which the redness of the ear was not accompanied by pain or redness spread on parts of the face or neck [4,5]. The nosological place of this syndrome is still unidentified in the ICHD-III. The prevalence of RES is also unknown, although it is thought that idiopathic RES is more common in children, while the secondary RES is probably more frequent in adults [6]. There are some articles that draw attention toward RES being associated with primary headache disorders, including migraine, chronic paroxysmal hemicranias (CPH), hemicrania continua (HC) and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) [7,8]. It is interesting that in our patient's case, the migraine without aura occurred about one year later after RES and usually migraine and RES appeared simultaneously on the same side of head. This coexistence was proved by Raieli, who showed in her study that in the pediatric age group the RES is a rather frequent clinical sign during headache and is more frequent during migraine [7]. It should be noted that the symptoms of RES do not have to be associated with a headache. Usually the episodes were spontaneous but not so rarely were precipitated by exertion, heat or cold, stress, touch, neck movements, eating or drinking or other common daily activities. The duration and frequency of RES attacks is very various, an attack usually lasts from a few seconds to several hours. Sometimes, the attacks occur only in clusters, lasting up to 2–3 months, and then the frequency of attacks seizures may be several times a day. In our case RES occurred in clusters, lasting up to 3 months, only in autumn and in winter. The occurrence of symptoms of RES in clusters and coexistence of migraine with RES may support the association of RES with primary headache disorders, particularly with migraine and cluster headaches. On the basis of the available literature on RES, the differential diagnosis should be taken into account: upper cervical spine, trigeminal neuralgia, glossopharyngeal neuralgia, CPH, temporomandibular joint dysfunction,

thalamic syndrome, migraine, otic zoster, herpes zoster virus infection of the C3-C6 dermatomes, Chiari type 1 malformation, SUNCT, erysipelas and exercise-induced compression of the cerebellar tonsils [1,7,8]. In our patient's case the differential diagnosis were excluded.

The exact pathophysiology of RES is unknown but an understanding of a possible pathophysiological hypothesis requires an understanding of the neural and vascular supply of the ear. The earlobe and the external auditory canal are innervated by C2 and C3 roots which form a sensory auriculotemporal nerve, the great auricular nerve and the auricular branches of the vagus nerve. The auriculotemporal nerve is delivered from the third branch of the trigeminal nerve. The ear lobe receives its blood supply from the external carotid artery through an anastomosis between the anterior auricular branches of the middle temporal artery and the posterior auricular artery. These vessels are supplied by sensory fibers from the trigeminal nerve. The vasostenosis and the vasodilator fibers of ear include an autonomic innervation. Flare in the RES is a result of activation of sympathetic vasodilation or inhibition of sympathetic vasoconstriction [9]. The electrical stimulation of the third cervical root shows vasodilatation of the ear and the part of the cheek nearby to the ear. It seems that the major role of this mechanism function is the inhibition of sympathetic activation with vasodilatation, because parasympathetic activation does not arise on the ear [9]. RES often coexists with other primary headaches, especially with migraine, CPH, HC and SUNCT what can be explained by the excitation of the trigemino-autonomy [8]. For this reason some authors suggested that RES may be part of the spectrum of the trigeminal autonomic cephalalgias. Kumar and Swanson believed that the primary RES form probably being a migrainous phenomenon and the secondary RES form being a neuralgiform radiculopathy involving the C3 root with release of vasodilator peptides or nitric oxide [9]. Moreover it is postulated that this syndrome is the result of abnormal neuronal connections in the brain stem between the roots of the C2-C3 core, trigeminal nerve and parasympathetic fiber innervating the skin of face [10]. This hypothesis is supported by Maranhão-Filho and Vincent who postulated that a brainstem dysfunction activates the trigemino-vascular system bilaterally, leading to sensory antidromic and parasympathetic reflex firing [5]. The conception described by Lance and the others authors that the RES should be classified as a new group, called auriculo-autonomous headaches, because the auriculotemporal nerve pathogenesis plays a major role, has not gained an approval of the contemporary authors dealing with this syndrome [1].

There are no studies and recommendations what are the most effective drugs for the treatment of RES. It is very important that at the beginning secondary RES must be excluded, because in that case the treatment is dependent on the cause of that RES. Patients diagnosed with secondary RES are usually drug-resistant. Furthermore, the patient should be informed how to avoid triggers or factors provoking the attacks of RES. Tricyclic antidepressants (imipramine and amitriptyline), flunarizine, β -blocker (propranolol), and calcium antagonists (verapamil), have been reported in concurrent migraine to be incompletely effective. Data on treatment with ibuprofen, indomethacin, and gabapentin in the literature is contradictory, but there are more cases describing the efficacy of these drugs,

particularly when RES coexists with other primary headaches [10]. There is a description in the literature of a similar syndrome on the scrotum area which was effectively treated with gabapentin [11]. There is one case report with slight reduction of pain after administration of pregabalin [12]. In our case the carbamazepine was not efficient, but more time may be required to be able to evaluate the effectiveness of this drug. In one case, the injections subcutaneously along the pinna and a C2-facet injection of bupivacaine and corticosteroids were ineffective [10]. Lance described that local anesthetic block of the C2 and C3 roots abolished flushing in one patient but the blockade of the auriculotemporal and posterior auricular nerves did not show efficacy [1]. The local anesthetic blockades of great auricular nerve by prilocaine mixed with methyl-prednisolone or methyl-prednisolone acetate showed efficacy [6]. In the literature, there is a case report that third local anesthetic blockades of great auricular nerve by 40 mg of methylprednisolone acetate completely subsided [13].

4. Conclusion

RES is an underdiagnosed disorder because it is a relatively new disease, and thus rather unknown. In our case report the symptoms of RES preceded migraine without aura, however the symptoms of RES have not always been associated with headaches. Furthermore, it is interesting that in our case report signs of redness of the ear occurred in clusters. The occurrence of such symptoms may have confirmed the observation that RES is associated with primary headaches. In RES, there are still many unsolved issues requiring further, careful observation and research. We describe our case, because we believe that the symptoms of RES in our patient can contribute to a greater understanding of RES.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; Uniform Requirements for manuscripts submitted to Biomedical journals.

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