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Coexistence of Migraine Headache and Red Ear Syndrome

Kırmızı Kulak Sendromu ile Birliktelik Gösteren Migren Baş Ağrısı

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Dear Editor,

Red Ear Syndrome (RES) is a rare condition originally described by Lance (1). RES episodes are characterised by unilateral or bilateral attacks of paroxysmal burning sensations and reddening of the external ear. The duration of these episodes ranges from a few seconds to several hours. The attacks occur with a frequency ranging from several daily to a few per year.

An 11-year-old boy reported a 3-year history of migraine without aura. During the previous 6 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. He had no family history of any migraine or other neurological disorders. Investigations including blood counts, serum electrolytes, and biochemistry were all within normal limits. Except for the reddening of the right external ear (Figure 1), the physical and neurological examinations were normal, as well as electroencephalography and brain-cervical spine magnetic resonance imaging. The patient was given prophylactic treatment with amitriptyline and subsequently reported a reduction in the frequency of both migraine attacks and RES episodes.

Two different forms of RES can be distinguished: Primary early-onset idiopathic RES seems to be associated with migraine, whereas primary late-onset idiopathic forms have been reported in association with trigeminal autonomic cephalalgias (2,3). Secondary forms of RES occur with upper cervical spine disorders or temporo-mandibular joint disorder (4).

Aetiology, pathophysiology and treatment of this syndrome have not been clearly defined. Episodes can occur spontaneously or be triggered, most frequently by rubbing or touching the ear, heat or cold, chewing, brushing of the hair, neck movements or exertion. The pathophysiology of RES is still unclear but several hypotheses involving peripheral or central nervous system mechanisms have been proposed (5). RES is regarded refractory to medical treatments, although some migraine preventative treatments have shown moderate benefit mainly in patients with migraine-related attacks. Open-label trials have suggested a beneficial effect with in some cases with the use of gabapentin, amitriptyline, imipramine, flunarizine, propranolol, verapamil and pregabalin (6).

RES could be considered a potential accompaniment to migraine. Rarely, RES symptoms conceivably may represent the only clinical manifestations of an acute migraine attack (2). RES associated with migraine occurs more commonly in childhood, whereas the secondary forms prevail in later years. Amitriptyline treatment was found to be efficient in our case, compatible with the literature. Hence, amitriptyline can be preferred for treatment in migraine headache patients with RES.

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Figure 1. A spontaneous red ear syndrome attack, in our patient diagnosed with migraine

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