

OSCILLUCUSIS AND SUDDEN DEAFNESS IN A MIGRAINE PATIENT

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ABSTRACT - Migraine is a complex disease that includes neurologic, gastrointestinal and autonomic symptoms, although headache is most common feature. In a portion of cases headache is preceded by focal neurologic symptoms termed auras. Auditory symptoms only rarely occur as part of an aura. We describe a patient whose 13-year migraine history that included the abnormal perception an oscillation of the intensity of ambient sounds (oscillucisus). During a migraine attack immediately after oscillucisus, the patient developed acute and permanent sudden deafness. Clinical and neurologic examinations revealed only profound hearing loss in her left ear. Audiometric testing confirmed the sensorineural nature of the hearing loss. The clinical aspects and physiopathology of auditory symptoms in this case and in patients with migraine is reviewed.

KEY WORDS: aura, headache, migraine, oscillucisus, sudden deafness.

Oscilucusia e perda auditiva em um paciente com migraena.

RESUMO - Migrânea é desordem complexa que inclui sintomas neurológicos, gastrointestinais e autonômicos, na qual a cefaléia é o achado predominante. Em uma parcela de pacientes o quadro álgico pode ser antecipado por sinais neurológicos focais conhecidos como aura. Descrevemos um paciente que iniciou sua história de migraena acompanhada por sensações de flutuações nos sons ambientes (oscillucisus) por treze anos. Durante uma crise de cefaléia imediatamente após a oscilucusia o paciente desenvolveu um quadro agudo e permanente de perda auditiva. Exames clínicos e neurológicos revelaram somente uma perda profunda da audição em ouvido esquerdo, cuja natureza sensorineural foi confirmada por uma avaliação audiométrica. Os aspectos clínicos e fisiopatológicos dos sintomas auditivos encontrados neste caso e em pacientes com migraena com aura são revisados e comentados.

PALAVRAS-CHAVE: aura, cefaléia, migraena, oscilucusia, perda súbita de audição.

Migraine is one of the most common neurologic diseases that occurs in the general population¹. The combinations of symptoms, including headache, neurologic, gastrointestinal and autonomic features¹, the focal neurological phenomena that precede or accompany an attack are known as aura. These neurological manifestations include visual, somato-sensory, olfactory and less frequently auditory symptoms.

The case of a migraine patient with auditory aura followed by hearing loss is revised, and the literature about auditory symptoms in migraine with aura patients are reviewed.

CASE

A 56-year-old woman started to present migraine episodes at age 40. An year later she perceived ambient sounds to be fluctuating in intensity (oscillucisus). This would last 50 minutes, and was followed by a unilateral

(right or left side), throbbing headache associated with nausea and vomiting. These episodes lasted 18 hours, occurred at twice a month and were commonly triggered by stress. At age 53, she had the sudden onset of profound hearing loss in her left ear that was unassociated with vertigo or any other neurologic symptoms. This symptom started 30 minutes after the oscillucisus and was followed by a migraine attack that lasted one day. The profound hearing loss in her left ear remained unchanged throughout the remainder of her life.

The patient's medical history was negative for hypertension, coronary artery disease, stroke, recent viral infection, and hypercoagulation syndromes (polycythemia and macroglobulinemia). Her family history revealed that her mother and daughter had migraine headaches. The general and neurologic examinations were normal except for the profound left-sided hearing loss. Complete hematologic and metabolic work-ups, including thyroid function studies, were normal. MRI and CT of the brain were nor-

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mal. Audiometric testing showed a severe sensorineural hearing loss in the left ear with discrete sensorineural hearing loss in the right ear. Vestibular function testing performed with rotation, in the dark, at multiple frequencies and peak velocities were normal. Auditory evoked potential of the brainstem was normal.

The patient was started on propranolol 40mg/day. This improved her headache, decreasing its frequency to one attack every three months. It also decreased the intensity and duration of her headaches. The migraine has been controlled with propranolol for three years; however the auditory disturbance remained unchanged.

DISCUSSION

Migraine is a common neurologic disorder characterized by attacks that consist of various combinations of headache and neurologic, gastrointestinal and autonomic symptoms¹. The clinical spectrum of this disorder is abundant and some patients will develop focal neurological symptoms that precede or accompany an attack of headache. These symptoms, called aura, consist of visual, sensory, or motor phenomena and may involve language or brain stem disturbances¹. Auditory symptoms are less common than vestibular symptoms²⁻⁴. Phonophobia is probably the most common auditory symptom associated with migraine, occurring at some time in more than two thirds of patients, usually in association with headache⁵. Auditory symptoms also include: auditory hallucinations⁶, oscilloclucis⁷, tinnitus⁸, fluctuating low-frequency hearing loss⁹, and sudden deafness¹⁰⁻¹².

Auditory hallucinations as aura in migraine patients (AHAM) without psychotic disease has been reported in different studies^{6,13}. The auditory hallucinations include voices accompanying the migraine attacks^{6,13}, the sense that ambient sounds were fluctuating in intensity (oscilloclucis)⁷, and tinnitus, which can occur in many conditions other than ototoxicity, such as AHAM⁸. In AHAM we need to eliminate other etiology, such as autosomal dominant partial epilepsy¹⁴ and thalamic and pontine auditory hallucinosis¹⁵.

Sudden hearing loss is defined as a severe to profound loss of hearing occurring over minutes to hours that is sensorineural in origin¹⁶. Sudden unilateral deafness occurring in a young patient is usually due to viral infection of the cochlea¹⁷⁻¹⁹, but the most common causes of sudden unilateral deafness are cerebrovascular diseases or hypercoagulation syndromes, such as polycythemia and macroglobulinemia²⁰⁻²². Viirre et al described the sudden hearing loss in 13 migraine patients¹¹, all of whom showed a sudden onset of hearing loss and other neurological

phenomena that could be attributed to vasospasm, including vertigo, amaurosis fugax, hemiplegia, facial pain, chest pain, and visual aura¹¹. Acute hearing loss can also be secondary to endolymphatic hydrops diseases²³⁻²⁴. In this situation, the acute stimulation of one of the otolith organs from the hydrops will produce drop attacks that may lead to confusion with vertebrobasilar insufficiency²⁵. Migraine patients have reported the abrupt onset of a profound hearing loss²⁶⁻²⁸. Although some gradual improvement may occur, they are often left with a severe, unilateral, or bilateral²⁷, sensorineural hearing loss. Some patients with sudden hearing loss report a previous history of fluctuating hearing in the same ear, and many develop persistent tinnitus²⁹.

Previous reports have observed that migraine can lead to permanent auditory and vestibular deficits^{11,26,30-31}. It is common that migraine-related transitory neurological symptoms lasting more than four and less than 60 minutes can be considered to be aura. The most common migraine aura is visual, and metabolic studies of the brain suggest a primary neuronal basis with secondary vascular changes, such as vasospasm or vasodilatation depending on the phase of migraine³²⁻³³. In retinal migraine this vasospasm may result in blindness³⁴⁻³⁵, illustrating vasospasm to have occurred at some time during the migraine attack^{12, 21,36}.

The pathophysiology of auditory symptoms is probably related with vasospasm of small arterioles within the cochlea and labyrinth, much as retinal migraine may involve only a subset of arterioles in the retina^{11,12,22,37}. However controversy exists regarding its role in the production of the symptoms^{29,37}. Another way to explain the heterogeneity of inherited migraine syndromes is to postulate a group of defects in genes that code for a family of protein³⁸. These proteins build the ion channels that, in migraine, produce an abnormal voltage-gated calcium-channels. The ion channels in the inner ear are critical for maintaining the potassium-rich endolymph and neuronal excitability. A defective ion channel shared by brain and inner ear could lead to reversible hair cell depolarization and auditory and vestibular symptoms²⁹.

In our case the presence of oscilloclucis before the headache phase of migraine suggests an auditory migraine aura. The etiology of this symptom could be related to transient vascular changes in the cochlea during migraine attacks. Our patient has had an association between migraine and auditory aura for thirteen years, when she developed sudden and permanent deafness. Although the etiology of these

symptoms remains to be determined, we believe that both oscillucis and sudden and permanent deafness in migraine patients reflect localized aura phenomena and cochlear ischaemia, respectively.

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