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Tinnitus as a Presenting Symptom in Secondary Neuropathy: a Case Report

Marina Savastano, M.D., Raffaele Bottin, M.D., Claudio Andreoli, M.D., *Michela Marcon, M.D.

*ENT Clinic and *Neurologic Clinic, Padua University, Padua, Italy*

Abstract: The authors describe a case of neuropathy characterized by involvement of most of the right cranial nerves, the initial symptom of which was tinnitus. The most likely etiopathogenetic hypothesis seems to be the paraneoplastic syndrome in which a primitive neurogenic axon lesion is more likely, although a myelinopathic lesion cannot be ruled out. It is feasible to suggest that the tinnitus was caused by a central neuron demyelination associated with concomitant peripheral nervous system involvement. Observation of present case allows the authors to point out that tinnitus may be the early sign of neurological disease which can evolve into the central and/or peripheral neurologic disease.

Key Words: Tinnitus, onset symptom, secondary neuropathy.

INTRODUCTION

Tinnitus is a symptom stemming from auditory nerve fiber activity in the absence of any sound.[1] It may be the presenting sign of an otoneurological pathology, such as schwannoma of the auditory nerve in which it is frequently the symptom that leads to diagnosis.[2] As Eggermont explained, all levels of the nervous system are variously involved in the onset of tinnitus, so different neurovascular alterations may be responsible for the symptom.

This paper describes a disease case of the central and peripheral nervous system mainly involving myelinopathic lesion, with the presenting symptom of tinnitus.

CASE REPORT

G.L., 66 years old, was referred to the neurology department with pain and paresthesia in the region of the right fifth cranial nerve, associated with double vision looking toward the right. At least a month before the onset of these symptoms the patient had complained of severe and persistent tinnitus in the right ear which subsequently developed slowly into a progressive deterioration in ipsilateral hearing function. The symptoms worsened, spreading caudally due to the successive involvement of the majority of the right cranial nerve (i.e., the first, third, fourth, fifth, sixth, seventh, eighth, ninth and tenth) and interfering with lower limb motility. Four months after the onset of tinnitus the patient's condition rapidly deteriorated with a progressive loss of consciousness and ultimate exitus. At the time of presentation, the ENT examination revealed an absence of velum platinum motility on the right side, no objective otologic signs; liminar audiometry demonstrated moderate-severe right sensorineural hearing loss and measurement of the characteristics of tinnitus indicated a frequency of 2500 Hz and an intensity of 55 dB; ABR showed signs of retrocochlear damage.

The hypoacusia progressively deteriorated over two months into anacusia (Fig. 1). The most significant clinical investigations performed were the CT scan of

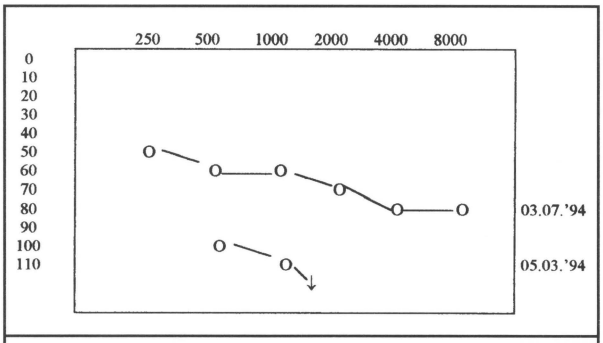

Fig. 1. Liminar audiometry of right ear performed at the time of observation and two months later

Reprint requests: Marina Savastano, M.D., Via Monegario 6/c, ENT Clinic, Padua University, 35127 Padova, Italy.

the brain with and without contrast (revealing a lesion in the paramedial frontal region between the two frontal horns); examination of the cerebrospinal fluid (showing severe barrier damage, normal pressure and immunocytological evidence of gross atypical cells: the outcome of cytology for neoplastic cells was dubious); electronystagmography (EMG), (showing signs of lower limb neuropathy of a myelinopathic nature, with extensive peripheral denervation); MRI of the brain with and without gadolinium (confirming the frontal lesion and also a small dubious area of alternated signals in the right lateropontine and mesoencephalic regions); total-body CT (images of solid nodules about 1 cm in diameter in the left lung with numerous enlarged lymph nodes); and raised humoral neoplastic markers.

CONCLUSION

It is common knowledge that tinnitus can be caused by central and peripheral neurological alterations and by the complications of cardiac or vascular malformations, metabolic disorders, hyperdynamic circulation, high endocranial pressure or tumors.[3] It has been demonstrated that this system coincides with an increased spontaneous activity of the auditory cortex,[4,5] which this symptom may correlate with a phenomenon originating in the inferior colliculus,[6] where the mechanism may be explained by reduced inter-neuron inhibition leading to a lower excitability threshold, demonstrated by an increased maximal neuronal discharge. Moreover, as Eggermont suggests, this pathological condition may be due to the excitation of a nerve fiber by other aberrant nerve fibers, with the synchronization of the various synapses. This condition is found in cases of auditory neuron myelin sheath lesions.

In the case described here, the two lesions detected by CT and MRI of the brain were not consistent with the patient's clinical picture. It is feasible to suggest, that the tinnitus was caused by a "central" neuron demyelination, associated with concomitant peripheral nervous system involvement, as demonstrated by the EMG.

The origin of this demyelination unfortunately remains to be explained, since no autopsy was performed to clarify the various hypotheses considered. The suggestion, that the successive cranial nerve damage should be secondary to neoplastic or inflammatory disease of the base of the skull (as in Garcin's syndrome), was not demonstrated by radiological investigations (which are, in any case not necessarily capable of providing a final diagnosis). The most likely hypothesis is paraneoplastic syndrome, in which a primitive neurogenic axon lesion is more likely, although an original myelinopathic lesion cannot be ruled out. In the patient considered here, a pulmonary lesion typical of such pathologies is also detected, but it is difficult to interpret these findings in this light, because of the unilateral cranial nerve involvement.

Nonetheless, it is worthy to emphasize that tinnitus may be the initial symptom not only of essentially peripheral otorhinolaryngoiatric pathologies, but also an early deceptive sign of neurological disease that can evolve into the central and/or peripheral nervous system.

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