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Case Report

Relative Preservation of Ocular Stability in a Severe Posterior Labyrinth Malformation Case

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Abbreviations Y.O: Year-Old; Cvemp: Cervical Vestibular Evoked Myogenic Potentials; LE: Left Ear; RE: Right Ear; vHIT: Video Head Impulse Test; VOR: Vestibular Ocular Reflex; MRI: Magnetic Resonance Imaging; SVV: Subjective Visual Vertical; oVEMP: Ocular Vestibular Evoked Myogenic Potential; COR: Cervico-Ocular Reflex

Abstract

We present the case of a patient with a malformation of the inner ear, proposing hypotheses to explain the relative preservation of ocular stability when exist a severe posterior labyrinth malformation.

Introduction

There have been reports of anomalies in the development of the inner ear, even with isolated anomalies of the vestibule [1]. In these cases, the auditory function could or could not be compromised, and although the vestibular tests were abnormal in some of them, the patients did not present alterations in their balance1. Little is known about the mechanisms that allow compensation for the lack of development of the lobby.

We present a case with isolated unilateral malformation of the vestibule and propose hypotheses about the compensatory mechanisms.

Case Presentation

The case of a female patient, 54 y.o, is presented here. 33 years before consultation, she suffered a vertigo episode that lasted approximately 20 hours, and which gradually improved until the patient was asymptomatic. For the last 30 years she has been suffering from a right side progressive auditory loss, currently without auditory function.

For the last 3 years she has been suffering from instability. She has no important pathological history.

Physical examination

Otologic examination:

- Ectoscopically normal
- Normal ears

Neurological examination:

- Normal ocular movements with full dynamic range, no gaze-evoked nystagmus is observed.
- Motor system with no peculiarities
- Normal cerebellar tests

Neuro-Otologic examination:

- Romberg test with right pulsion
- Positionaltests (Dix-Hallpike and Mc Clure): normal
- Head-shaking: Nystagmus to the left
- Head-heave: Positive to the right
- Absence of spontaneous nystagmus with and without ocular fixation

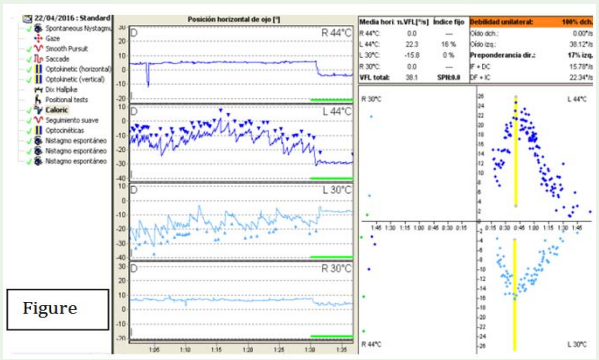

Figure 1: Caloric test.

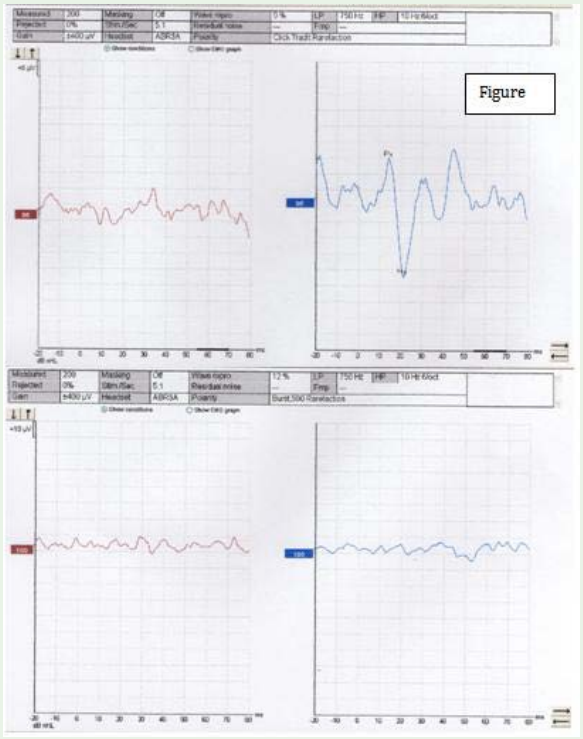

Figure 2: cVEMPs.

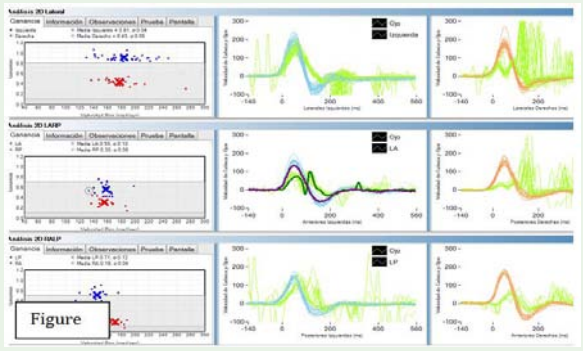

Figure 3: Video head impulse test.

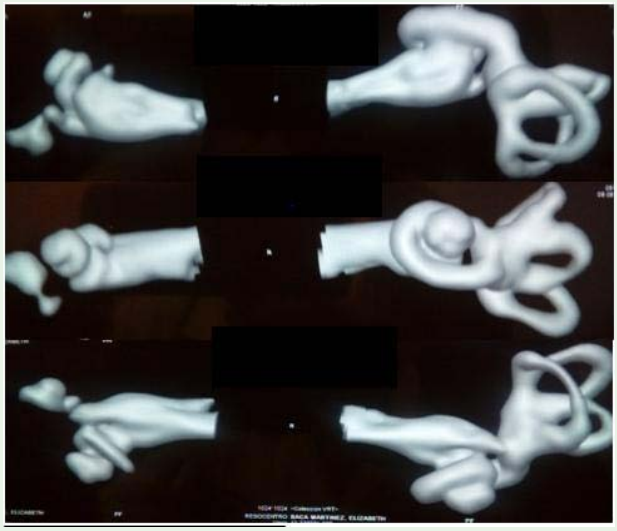

Figure 4: MRI.

Supplementary tests

- Caloric test: no response in right ear (Figure 1).
- Tonal audiometry: right ear: no hearing. Left ear: 20 dB.
- Subjective Visual Vertical (bucket test): leaning of the 3rd vertical towards the left.
- cVempwith normal response in LE and lack of response in RE and oVemp with no reproductive response in either ear are performed (Figure 2).
- vHIT: there is a markedasymmetryfor VOR gain in the six canals of the LE, with covert saccades; the gain is relatively preserved for the horizontal and anterior canals and it shows a larger drop in the posterior one (Figure 3).
- MRI: the image shows absence of the three semicircular canals in the LE (Figure 4).

Discussion

Our patient suffers from a severe malformation of the internal ear, with absence of all the semicircular canals; yet, the function of the anterior and mainly of the lateral canals indicate that there is enough function for the patient to be compensated. The posterior canal function, the utricular function (cVEMPs and SVV) and the sacular (oVEMPs) function are distinctly altered which rules out the possibility of ocular movements being compensated by these receptors.

The vestibular nuclei contains Type I neurons which increase their discharge rate by ipsilateral rotation and Type II neurons which do so by contralateral rotation; thus, Type I neurons of a vestibular cluster receive inputs from the ipsilateral lateral semicircular canal, and via the commisure they project to the neurons in the contralateral vestibular nucleous which, in time, receive an input for the corresponding lateral canal [2]. This system functionally inhibits the opposing labyrinth, so that the loss of ipsilateral function would reduce the inhibiting effect and induce an increase in the activity

of the contralateral nucleous. In this situation, the dynamic activity of vestibular neurons depends solely on the cephallics peed signals generated at the intact labyrinth [3]. After the loss of unilateral function, Type II neurons in the contralateral medial vestibular nucleous show an increase in their activity [4]. In our case, the contralateral head rotation would produce responses which, via the commisure and Type II neurons would lead to a VOR type response in the affected side.

Another explanationis that these findings indicate that the Cervico-Ocular-Reflex (COR) may be the compensating mechanism in this case. Several authors have demonstrated a COR gain increase in different kind of peripheral l and central lesions [5].

The history of vertigo as well as that of hearing loss indicates that this defective inner ear may have functioned in the past, but maybe with the help of a COR with increased gain, which would explain the quick compensation.

The new methods for neurophysiological study together with the new imaging techniques make it possible for us to have a profound understanding of the underlying mechanism behind the clinical manifestations of vestibular function.

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