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LERMOYEZ SYNDROME: A LITERATURE REVIEW

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ABSTRACT

Introduction: Lermoyez Syndrome is considered a rare pathology of unknown etiology, whose clinical picture includes hypoacusis, tinnitus and vertigo, similar to Ménière's Disease, differing from the latter due to the improvement in auditory acuity during the vertigo crisis. GOAL: To review the most up-to-date articles on Lermoyez Syndrome, given its relevance for a full understanding of otoneurological complaints. **Methods**: Literature review of articles published in the last 30 years, in English, Portuguese and Spanish, using the keyword “Lermoyez syndrome” in the MEDLINE, PUBMED and Scielo databases. **Results**: Four articles were screened for full analysis. Three inferred that, although similar, there are intrinsic aspects in these patients, which distinguish their pathophysiology from those with Ménière's Disease. Audiometric tests seem to overlap the vestibular ones in terms of positive predictive value, both being widely detailed in the studies, highlighting the finding of improvement in the auditory threshold, after the vertigo episode, at lower frequencies. **Conclusion**: Although the clinical picture and audiological and vestibular findings of Lermoyez Syndrome are well described, the scarcity of studies about this disease makes it difficult to fully understand this pathology as well as its proper management.

Keywords: Lermoyez Syndrome. Vertigo. Otorhinolaryngology.

RESUMO

Introdução: A Síndrome de Lermoyez é considerada uma patologia rara e de etiologia desconhecida, cujo quadro clínico inclui hipoacusia, zumbido e vertigem, à semelhança da Doença de Ménière, diferenciando-se deste último pela melhora da acuidade auditiva durante a crise vertiginosa. **Objetivos**: Revisar os artigos mais atualizados sobre a Síndrome de Lermoyez, dada sua relevância para a plena compreensão das queixas otoneurológicas. **Métodos**: Revisão de literatura de artigos publicados nos últimos 30 anos, em inglês, português e espanhol, mediante busca com descritor "Lermoyez syndrome" nas bases de dados MEDLINE, PUBMED e Scielo. **Resultados**: Quatro artigos foram triados para a análise na íntegra. Três inferiram que, embora similar, há aspectos intrínsecos nestes pacientes, que distinguem sua fisiopatologia daqueles com Doença de Ménière. Testes audiométricos parecem sobrepor os vestibulares em termos de valor preditivo positivo, sendo ambos amplamente detalhados nos trabalhos, destacando-se o achado da melhora no limiar auditivo, após o episódio de vertigem, em menores frequências. **Conclusão**: Apesar do quadro clínico e dos achados audiológicos e vestibulares da Síndrome de Lermoyez serem bem descritos, a escassez de estudos acerca desta doença dificulta a plena compreensão dessa patologia bem como seu adequado manejo.

Palavras-chaves: Síndrome de Lermoyez. Vertigem. Otorrinolaringologia.

INTRODUCTION

Lermoyez Syndrome was initially described by the French researcher and otorhinolaryngologist Marcel Lermoyez in 1919 in which the patient reported a reduction in hearing acuity preceding the vertigo crisis, but with improvement or complete recovery of hearing during the vertigo crisis[1,2]. Therefore, despite being similar to Ménière's Syndrome, it presents a paradoxical audiological finding[3,4].

Despite this reverse pattern of the patient's symptoms, as we still have few studies on this topic, this syndrome is considered a variant of Ménière's disease[5,6]. Thus, two mechanisms are proposed for this disease: a spasm of the internal auditory artery, proposed by Marcel Lermoyez himself 1, and obstruction of the ductus reuniens, communication channel between the saccule and the cochlea — the latter being currently more accepted, although still requires further studies for its complete pathophysiological elucidation[7,8,9].

The clinical presentation is very rare, it is believed that around 0.2% of people with Ménière's disease have Lermoyez[5]. Despite the statistical limitation, this peculiar prevalence should encourage the possibility of diagnosis, as advances on this topic are published. Therefore, this work seeks to review the most up-to-date articles on Lermoyez Syndrome, given its relevance for a wider comprehension of patients with otoneurological diseases.

METHODOLOGY

This is a literature review of articles published on the topic of Lermoyez syndrome in the MEDLINE, PUBMED and SciELO Citation Index10,11 databases, based on the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) guidelines[12].

In the research strategy, “Lermoyez syndrome” was outlined for a broad approach to this rare pathology. The search was carried out on May 20th, 2023, with a total of 21 articles found. Then, the following filters were selected: “last 30 years”; "English"; "Portuguese”; and “Spanish". Only case series studies, clinical trials, reviews and meta-analyses were analyzed.

Duplicate articles and studies that only evaluated case reports were excluded. We did not apply any exclusion criteria regarding study duration or sample size.

The articles were selected by two reviewers who worked independently, in two stages. Initially, titles and abstracts were read, including publications that suggested the presence of original results on Lermoyez Syndrome. In case of disagreement between the two reviewers, the report was included at this stage.

In the second stage, based on the selected abstracts, the full articles were reviewed by the two reviewers. Conflicts of disagreement regarding the inclusion of articles were then resolved by exposing the two conflicting decisions to a third reviewer, who decided on inclusion or exclusion.

Data extraction was carried out by two evaluators, with analysis of the studies' descriptive and methodological data, such as year of publication, country, population and study design, topics for clinical practice and results. Subsequently, the data were compared and discussed.

RESULTS AND DISCUSSION

After inserting “Lermoyez syndrome” in the aforementioned electronic databases, 21 studies were identified. At the end of the filter selection, four articles were included for this systematic review, as described in the flowchart that demonstrates the selection process (Fig. 1).

Regarding the type of study, two articles are case reports while the other two are case series. All studies were published in the last 13 years, the most recent being in 2020. It is noticeable that the two most recent publications are case series, given the growth in cases over the years.

The country with the largest number of studies was China (n=2), although collaborations were found from countries such as Australia, the United States, Italy, Sweden and Taiwan. No selected study was carried out by Brazilians.

From the four studies included, it was possible to group the main findings on Lermoyez Syndrome into four categories: “Epidemiology and pathogenesis”, “Clinical manifestations and physical examination” and “Complementary examinations”.

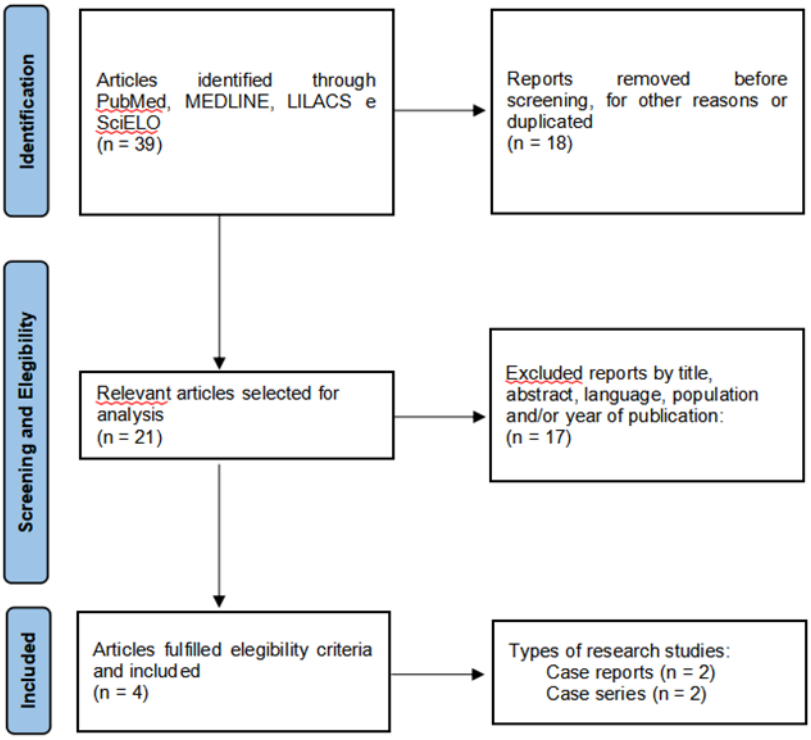

Figure 1: Literature search and selection

Epidemiology and pathogenesis

A restrição da dieta é decorrente do tipo de anestesia, da doença e do tipo de procedimento cirúrgico que será realizado. Sob anestesia geral, o jejum pré-operatório deve ser de oito horas, para evitar broncoaspiração durante a indução anestésica ou a intubação orotraqueal. Pacientes obesos, gestantes, portadores de hérnia hiatal, ou com grandes tumores intra-abdominais, têm maior risco de broncoaspiração e devem sempre fazer jejum de 12 horas[10].

The Taiwanese report published in 2018 by Shen et al.[13] describes seven patients with Lermoyez Syndrome, five male and two female, selected between 1992 and 2017, among 4096 cases of Ménière's Disease. Such prevalence was similar to that found by Schmidt et al[5].

The Chinese work by Zhou et al.[14], in turn, included two women and seven men with Lermoyez Syndrome, whose first symptoms occurred between the ages of 23 and 58. The authors believe that the different prevalence, since Ménière's disease is more common in women, and the lower intensity of cochlear lymphatic hydrops in Lermoyez syndrome, compared to Ménière's disease, indicate the existence of an intrinsic difference in the pathophysiology between the two diseases[14].

Meanwhile, the study by Zhang et al.[15] describes the case of a 49-year-old male patient, without comorbidities, who presented with Ménière's condition in the left ear and Lermoyez in the right ear. According to the authors, this case indicates a pathophysiological similarity between these syndromes, associated with different outcomes, however.

Additionally, Shen et al.[13], observed that vestibular tests of patients with Lermoyez Syndrome, such as the caloric test and the ocular and cervical Vestibular Evoked Myogenic Potential (VEMP), did not change significantly after the vertiginous crisis. These findings reinforce that Lermoyez Syndrome more significantly affects the cochlear portion of the inner ear, when compared to the vestibular portion — sharing a resemblance to the initial stages of Ménière's Disease. In addition to this hypothesis, Takeda et al.[16] in a 1996 study also indicate the existence of distinct pathogenesis between the uni and bilateral forms of Lermoyez Syndrome.

Clinical manifestations and physical examination

Lermoyez Syndrome presents symptoms similar to Ménière's disease, progressing with acute episodes of vertigo lasting minutes to hours, cochlear symptoms such as ear fullness, hypoacusis and tinnitus, as complained by the patient described by Zhang et al. 15 However, unlike Ménière's disease, there is improvement in hearing during episodes of vertigo[17].

Vertigo was reported by all patients in the Shen et al.[13] research, which alongside hypoacusis and tinnitus (both 86%) represent the Ménière's triad. Another characteristic in common between the two diseases is the presence of the intercrisis period, in which the patient is asymptomatic.

In the physical examination, emphasis should be placed on the otoneurological examination, identifying the presence of spontaneous and induced nystagmus, performing the headshake and the Dix-Hallpike maneuver. During the intercrisis period, otoneurological tests will be negative[17]. During the acute stage, Shen et al.[13] observed auricular fullness and positional nystagmus, both in 71% of patients, in addition to spontaneous nystagmus (57%) and nausea or vomiting (43%).

Complementary exams

The measurement of labyrinthine function in patients with Ménière's Disease and Lermoyez Syndrome, whether in the intercrisis or acute period, is not frequent. However, there is already evidence of changes in complementary exams for these pathologies, which can help in the diagnosis and management of cases[18]. The evaluation can be carried out by tests such as the (1) VEMP, which detects the function of the saccule and utricle; by the (2) Video Head Impulse Test (vHIT), which evaluates the semicircular canals; and by the (3) caloric test, which studies the function of the lateral semicircular canal.

In the cases tested by Shen et al.[13], vestibular tests showed a low percentile of abnormalities, with cervical VEMP (cVEMP) being positive in 43% of patients with Lermoyez Syndrome, while ocular VEMP (oVEMP) was not altered in any of the patients.

VEMP is included in the complementary workup for Ménière's Disease[19], demonstrating changes before VHIT, in the initial stages, since in the natural history of this disease the otolith organs are affected before the semicircular canals[18]. In Lermoyez Syndrome, both in the acute attack and in the intercrisis period, Manzari et al.[17] did not observe any change in amplitude in the cVEMP, which is a more commonly used test due to its easier capture. Although the stimulus in this test is sound, it is the vestibular system that is being evaluated. Therefore, even in cases of profound sensorineural hearing loss, such as Lermoyez Syndrome, the test can be used[18].

Regarding their patient, Manzari et al.[17] adds that oVEMP may show asymmetry above the reference value and decreased N10 amplitude in both ears, suggesting decreased utricular function. In Ménière's disease, the amplitude of the N10 below the eye ipsilateral to the healthy ear is generally increased during the vertigo crisis. The cross-response observed in the oVEMP implies that in patients with Ménière's disease, utricular function in the affected ear is increased during acute periods.

In addition to VEMP, there is vHIT, which can present asymmetry between the ears, due to the displacement of the dome. The gain of the vestibulo-oculomotor reflex (VOR) for rotations to the affected ear in Lermoyez Syndrome remains unchanged during the preacute stage, while there is a significant decrease in the gain of RVO for rotations to the healthy ear, according to the report from Manzari et al.[17] This reduced inhibitory response following amplifugal movement, due to endolymphatic hydrops, results in a smaller disinhibitory contribution of the affected ear to head rotations towards the healthy side, which explains the asymmetry seen in VHIT.

Regarding the caloric reflex test in Lermoyez Syndrome, the case report by Manzari et al.[17] was within normal limits, while the case series prepared by Shen et al.[13] found changes in only 38% of patients. It is important to reiterate that such an exam is uncomfortable for the patient and can lead to intense nausea and vomiting, which compromises the full evaluation of the exam.

When investigating auditory function, tonal and vocal audiometry with immittance testing is the gold standard test. It is also possible to evaluate stapedial reflexes which, in the patient studied by Manzari et al.[17], were normal. At the time of the acute vertigo attack, there was an improvement in hearing through the air, with a reduction in the threshold to 250 Hz by approximately 30 dB. The air-bone gap for low frequencies found in the intercrisis period disappears, which represents that in Lermoyez Syndrom, not only there is an improvement in low-frequency hearing, but the air-bone gap may also be abolished[17].

In the case series observed by Shen et al.[13], only one patient had bilateral involvement, while the others had unilateral involvement, totaling eight ears tested. Of these, only four (50%) had a hearing threshold above 25 dB in pure tone audiometry at 4 frequencies (500, 1000, 2000 and 3000 Hz). However, when analyzing global frequencies (125 to 8000 Hz), all patients had a hearing threshold above 25 dB, configuring hearing loss. The authors recognize that this test does not provide good parameters in the audiological evaluation of patients with Lermoyez Syndrome in general[13].

This same report attested to a statistically significant reduction ($p < 0.05$) in patients' hearing thresholds before and after the vertigo crisis, from an average of 36 dB (± 24) to 23 dB (± 23 dB), mainly in low frequencies and medium, between 250 and 1000 Hz.

The case described by Zhang et al.[15] also showed that auditory thresholds, especially in low frequencies, rose rapidly in the first months after the onset of the disease, showing slow progression in the following months. Tests during vertigo episodes were not performed.

Zhou et al.[14] also found a more significant hearing improvement in the lower frequencies of the participants, whose hearing thresholds before the assessment were, on average, 45.7 dB (± 16.7), while after the onset of vertigo were around 32.3 (± 11.6) dB. At intermediate and high frequencies, the variation was from 42.9 dB (±16.7) to 31.8 dB (± 11.5) and from 50.7 dB (± 10.6) to 45.0 dB (± 14.4), respectively.

In order to expand the investigation on vertigo attacks, imaging tests can also be performed, such as magnetic resonance imaging (MRI) of the posterior cranial fossa with contrast and high resolution computed tomography (CT) of the mastoid bones. Manzari et al.[17] additionally evaluated, in their case report, the eighth cranial nerve, middle ear and labyrinthine capsule of the inner ear. Nonetheless, MRI did not demonstrate abnormal findings. In contrast, some degree of endolymphatic hydrops was noted in all affected ears of the nine patients in the Chinese study by Zhou et al.[14], who also underwent MRI with contrast.

CONCLUSION

Both the clinical condition and the findings of audiological and vestibular exams in Lermoyez Syndrome, although well described in the literature, still lack full understanding due to the low prevalence of this disease. This limitation implies the absence of studies aimed at standardizing therapeutic management, which remains undefined.

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