Variable Presentations in Meniere's Syndrome.

Navin L Hirapandani, Hony Professor.

Dinaz Namdarian, Lecturer

Jyoti P Dabholkar, Professor.

Trupti Manjrekar, C.P. Rajeevan, Neelam Sathe. Senior Resident

Sameer Lambay. Junior Resident Department of ENT,

TNMC & BYL Nair Ch. Hospital Bombay - 400 008.

Introduction

One hundred cases of Meniere's Syndrome were studied for variable presentations. Vertigo can present as both acute and chronic manifestations. It is usually progressive in nature except for the periods of remissions. However, some people do have persistent vertigo over a long period of time. The severity of the vertigo does not necessarily depend on the duration of the disease.

As far as the deafness is concerned, it is of the sensorineural type and progressive in nature.

We have described Cochlear Meniere's where only deafness occurs in Meniere's Syndrome.

Recovery of vertigo can occur with treatment, but deafness does not improve. Tinnitus improves in above 30% of cases. Deafness and tinnitus are sometimes masked by the symptom of vertigo, which can be intractable.

Electronystagmogram [ENG] shows a characteristic peripheral response. Central and peripheral types of nystagmus can be differentiated, however localisation of the pathology and diagnosis should not be given with only ENG. It is necessary to consider Recruitment.

Phenomenon. Retrocochlear tests like Brainstem Evoked Response Audiometry [BERA] and the modern radiological investigations like

Computed Tomography Scan [CTScan] and Magnetic Resonance Imaging [MRI].

Discussion

Classical Meniere's Syndrome consists of Vertigo, Tinnitus and Deafness. It is a progressive disease characterized by attacks of Vertigo and periods of remissions. Caloric Test and ENG shows a characteristic Reduced response. Clinical Diagnosis is important as many of the cases can be diagnosed correctly and confirmed by the Caloric test and ENG.

Unilateral disease is more common but bilateral disease can occur. Atypical symptoms like imbalance, heaviness of the head, pressure sensation, etc., can occur.

Amongst the symptoms Vertigo is more typical. It also hampers the regular work. It causes considerable discomfort. It is also one of the debilitating symptoms of the body. The patient can not work but is disturbed and also can not do regular work like driving a car.

The vertigo is progressive and lasts from Seven weeks to Seven months, and occasionally more than a year. However as a variation the Meniere's Syndrome can present as a short duration disease which can be controlled with treatment in the early or late stage.

Vertigo can be controlled with treatment but the assessment is difficult due to the phases of remissions, i.e. the symptom free interval.

Although characteristic vertigo is common, the patient may have atypical vertigo, in which there is a sense of rotation of self rather than the surroundings. To and fro movements can occur. Vertigo can subside with time, esp. with adequate treatment.

However the termination of the symptoms is not always predictable and symptoms may subside in a particular stage of the disease. Long term vertigo is known to subside more easily probably due to dysfunction of the Sensori-aural Cells.

Tinnitus is often accompanied by vertigo. The symptom of tinnitus is masked by the symptom of vertigo. However, the patient complains subjectively when asked positively about the tinnitus. This is because the symptom of tinnitus is not noticeable in severe attack of vertigo. However, the patient may complain of associated vertigo occasionally.

The tinnitus may be of different types, like - whistling, ringing, sensation of flowing air, etc.

The tinnitus is noticed at night when the surroundings are quiet. It is more disturbing if the symptom of vertigo is not prominent. In this case also, the patient is unable to concentrate on his work and wants relief from the symptom. The treatment of tinnitus lies in the anti-labyrinthine drugs and only 50% chances of relief are expected. Occasionally the tinnitus subsides by itself. The tinnitus is often refractory to treatment.

We have done 50 cases of Vestibular Neurectomy through the Middle cranial Fossa Approach, but the tinnitus was persistent. This is unusual after a neurectomy, when the impulse from the nerves to the Central Nervous System [CNS] are cut off. Hence an alternative pathway does exist. Different authors have described different pathways; hence it is possible that an alternative access to the CNS exists.

The patient should not be given assurance about the relief from tinnitus. However, if the tinnitus subsides, it gives considerable relief to the patient. Tinnitus is often more annoying when it is the only symptom. However reassurance and anti-labyrinthine drugs provide certain relief.

Studies have been done to describe tinnitus at various frequencies, however no definite conclusions have been drawn regarding this symptom. Labyrinthectomy also does not give complete relief from this tinnitus.

Deafness is an important symptom at later stages. We have described Cochlear Meniere's where only deafness occurs. Paparella et al have said that the utricleoendolymphatic valve plays an important role in the pathogenesis of vestibular Meniere's as compared to Cochlear Meniere's. They also say that function of the valve is important in explaining patient's symptomatology.[3] The patient ignores the symptom at an early stage due to vertigo and tinnitus. Deafness is progressive as described and increases with time and at a later stage, severe deafness often occurs. At an early stage with deafness, the diagnosis of Meniere's can be difficult. At a later stage as the deafness progresses, diagnosis is confirmative.

During the late stages of deafness, the other symptoms of vertigo and tinnitus subside. This is already ascribed to the possible dysfunction of the sensorineural cells at a late stage.

As far as the relief of vertigo is concerned, in the early stages Medical line of treatment is given. This can be continued from several weeks to several months. Many of the cases show relief from vertigo. Vertigo is more easily controlled than other symptoms. Decompression of the sac can be done and relief of vertigo to the extent of 60% is seen. We have done 50 cases of Vestibular Neurectomy with 100% results. However since the operation is intracranial, it is reserved for intractable cases. Norre et al have said that training procedures improve the patients symptoms in a specific and genu-

ine way [2].

As far as the tinnitus is concerned, the treatment is more difficult. Although 30% results are seen with medical and surgical line of treatment, some patients get adjusted to the symptom and do not mind it in the long term and hence no special treatment is necessary.

As far as the deafness is concerned, it is progressive and does not improve with treatment. However the patient is often unconcerned about the deafness due to the severity of other symptoms. At a later stage, the deafness becomes more severe.

The positive finding of Recruitment, which is positive in typical cases, is diagnostic. Recruitment can be confirmed with Audiometry, and subsides with time.

Hyperacusis, which is an abnormal reaction to loud noises, occurs occasionally. This symptom is also diagnostic and can subside with time.

The recruitment may present as hypersensitivity to loud noises. But occasionally sensation of echo occurs in the ear.

As regards the evaluation of vertigo, caloric test and ENG are done. Classical Hallpike and Cairns test is done with caloric stimulation being given with cold and hot water at 30 degrees and 44 degrees Celsius resp., on each sides. The evaluation parameter in ENG is the maximum amplitude of nystagmus. This is done because comparison with all normal cases is facilitated. Parameters like number of beats of nystagmus are very variable and comparison with abnormal cases is difficult. Even the maximum number of beats does not take into consideration the amplitude of nystagmus. Hence evaluation of maximum amplitude of nystagmus gives a more accurate finding. Prognosis is given by repeating the ENG and seeing the results with treatment. In Meniere's syndrome, the classical slow and fast phase of nystagmus is seen and this phenomenon is also seen in the lesion of peripheral labyrinth.

Deafness is recorded by Audiometry. It shows mild to moderate or severe deafness depending on the severity of the disease. High frequency loss is recorded in a few cases with audiometric findings. Severe deafness is accompanied with relief of symptoms, possibly acting as Autolabyrinthectomy. As the deafness is of sensorineural type, it does not improve with medical line of treatment. This is unlike vertigo, which improves.

Tinnitus can be recorded at different frequencies with audiological evaluation, but is of mainly academic importance.

The test of Recruitment is done by Fowler's test when the difference between the two ears is minimum 30 decibels hearing loss. At low intensities the graph is flat. But at high intensities the patient hears better and there is a convergence of the graph at higher intensities due to better audition. Recruitment phenomenon is diagnostic if present.

Regarding the diagnosis with ENG, the central and peripheral types of nystagmus can be recorded. Specific diagnosis can not be given in reference to cortical and subcortical lesions in the cerebrum, nuclear lesion in the Vestibular apparatus or the peripheral nerve with ENG recordings. Except for identifying the central or peripheral types of nystagmus, specific localisation and diagnosis should not be given with ENG recordings. Babin has mentioned positional nystagmus in Meniere's Disease. We have found it to be a rare phenomenon. [1] We have not found a specific pattern pertaining to Meniere's Syndrome. Certainly central lesions like cerebellar lesions do show pendular tracings. However, in central lesions ENG response can be variable and diagnosis is not accurate as to the specific site of the lesion. Besides the patient of a central lesion most often lands up with a neurophysician. The ENT surgeon mainly deals with the peripheral lesions, viz. Meniere's Syndrome. Vascular lesion of peripheral labyrinth, etc these show characteristic peripheral types of nystagmus

with the slow and fast phase of recording. Hence, in the ENT department, majority of patients comes with peripheral lesions and diagnosis of a central lesion is a rarity. The only exception is Acoustic Neuroma of the VIII Nerve. Tos et al have mentioned that Acoustic Neuroma were estimated to represent 8-10% of all intracranial tumours [4]. Here also the principal diagnosis is done by the modern imaging techniques like the CT Scan and MRI. Severe reduction or absence of response in ENG may be suspicious but it is not diagnostic of acoustic Neuroma as it can occur in late stages of Meniere's Syndrome.

Summary and Conclusion

One hundred cases of Meniere's Syndrome have been described with variable presentations. Although Vertigo is a characteristic feature, the severity and duration of vertigo can vary. Characteristic remissions are not observed in some cases. Cochlear Meniere's is a phe-

nomenon, which has been described by us where only deafness occurs. Although uniform deafness occurs, high frequency loss occurs in a few cases.

Specific anatomical diagnosis with only ENG should be avoided and the battery of tests including Recruitment tests, SISI test, Audiogram and recent Radiological tests like CT Scan and MRI should be done to differentiate specially from Acoustic Neuroma.

Acknowledgement

I thank the Dean, Dr [Ms] K.D. Nihalani, TNMC & BYL Nair Ch. Hospital, for her permission to publish this article.

I also thank the Executive Director, Mr. C.G. Joshi and Medical Director Mr D.P. Vyas of Bombay Hospital for their help and co-operation.

I also thank the technician Ms N. Patil and the Residents who have helped me.

References

1. Babin Richard W., (1979): *Positional Nystagmus in Meniere's Disease*. *Annals of Otology Rhinology & Laryngology* 88:293.
2. Norre M.E. De Weerd W., (1980): *Treatment of Vertigo based on Habituation*. *Journal of Laryngology & Otology* 94:971.
3. Paparella Michael M., Goycoolea Marcos, (1981): *Endolymphatic Sac Enhancement Surgery for Meniere's Disease*. *Annals of Otology Rhinology & Laryngology* 90:610.
4. Tos Mirko, Thomsen Jens, (1984): *Epidemiology of Acoustic Neuromas* *Journal of Laryngology & Otology* 98:685.