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THE MENIERE SYMPTOM-COMPLEX

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To the instructive studies of Meniere, for many years director of the Paris Institution for Deaf Mutes, we owe our first intimate knowledge of what may be termed aural vertigo. Earlier writers had called attention to the relationship between otitis and nerve symptoms. Meniere's series of cases published in 1861 brought to the attention of the profession the symptom-complex, which has since been recognized as a clinical entity.

Let me briefly recall a few points relative to the anatomy and physiology of the internal ear. The function of hearing is but poorly understood. The sense of hearing is the last to awaken in the developmental period of infant life. Since the labyrinth is conceded by most authori-

ties as affected in Meniere disease, a word as to its anatomy is warranted. The three semi-circular canals, with their ampullæ, constitute an essential part of the labyrinth. It will be recalled that the bony canals enclose the thin wall structure of the same contour known as the membranous semi-circular canals. Each of these semi-circular tubes begins as an elliptical enlargement, the osseous ampulla, and each re-enters the vestibule after making an entire circular turn. The very delicate membranous semi-circular canal, enclosed in its compact osseous outer wall, contains endolymph. The membranous portion, less in diameter than its osseous covering, is attached to the endosteum of the latter by fibrous bands. The membranous portion, consisting of a system of cavities, receives the terminal endings of the auditory nerve. The semi-circular canals and the vestibule are organs of equilibration. The eighth, or auditory nerve, is composed of two parts, each having its own termination and central nuclei. The one portion, the cochlear, is the nerve of hearing. The other division is the vestibular part, and is the nerve of

equilibration. The latter division has its origin in the ganglion of Scarpa, and thus from the semi-lunar canals and utricle of the labyrinth and passes with the cochlear division to the pons varolii. The nuclei of the auditory nerve have direct connections with other parts of the central nervous system, particularly the cerebellum and third, fourth and sixth cranial nerves. These associations are of manifest importance in considering the phenomena of the Meniere disease.

As Starr says: "It is evident that the semi-lunar canals, in which impulses are received whose object is to make us aware of our position in space and to direct the whole system of movements by which our equilibrium is preserved, have a most widespread connection with the cerebral nervous system and an especially close connection with the cerebellum."

Let us now consider the disease under review. Meniere's original studies referred to what is now classed as the apoplectic type of the affection as true Meniere disease is now called—that form which is sudden—apoplectiform in its onset and based solely upon an acute condi-

tion. Pollitzer and Knapp regard those cases presenting the symptom-complex—deafness and the triad vertigo, tinnitus, vomiting—based upon a previously existing otitis, as quite distinct from the true de novo affection—the clinical entity—Meniere's disease.

The Meniere symptom-complex consists of the more or less sudden onset of deafness associated with the characteristic triad vertigo, tinnitus and vomiting. Together with these classic symptoms are often added subjective pressure sensations of the head, a varying degree of cerebellar ataxia and a true mystagmus. One or more of these associate manifestations may be present. Diarrhœa, often intense and serous, is not so infrequent.

In presenting a classification of the special forms of Meniere's symptom-complex and the conditions under which it develops, I have found Frankl-Hochwart's grouping the most comprehensive. We may consider first those cases coming on suddenly in patients who have not previously suffered from any form of ear affection. Among these is that class including the true apoplectiform cases,

whose anatomical basis is found to be hæmorrhage in the auditory labyrinth; we also find in these infiltration of the auditory nerve. The apoplectiform onset of deafness, partial or complete, tinnitus, vertigo and vomiting may appear in those individuals free from ear trouble and previously also in good general health. Again, the classic triad may come on in those of normal hearing apparatus but otherwise predisposed by active syphilis or its consequential following, particularly tabes dorsalis; by general arterial disease; nephritis; by leukæmia.

In another class we may place those cases determined by trauma such as result from violent blows upon the head, cerebral concussion, direct injury to the labyrinth, or from Caisson disease.

In the above cases we have presupposed freedom from previous acute or chronic ear disease. Not uncommonly, however, the Meniere symptom-complex develops in those with either acute ear affection or with a chronic form of otitis. The latter, the chronic ear cases, are by far more frequently associated with the Meniere triad than the acute. However, in acute laby-

rinthine involvement, the typical deafness and triad of symptoms is not uncommonly a form in which the Meniere appears. The so-called abortive forms of cerebro-spinal meningitis, beginning with deafness and vertigo in children, probably belong to this class. The toxic forms of the affection have a similar origin. Among the latter as factors are coffee, quinine and the salicylates. These cases are, of course, of the mild form of the Meniere symptom-complex, and are purely symptomatic. Another class of the same type are those with chronic disease of the eighth cranial nerve, the acusticus or its root, and among them are the tabetic cases and those seen in basal brain tumor.

Frankl-Hochwart has described a symptom-complex developing acutely with fever, herpes, facial paralysis and nerve deafness combined with the Meniere triad. This he calls polyneuritis centralis Menierformis, and regards its etiology due to probable developing basal brain tumor.

Of interest also is the affection described by Gertier of Switzerland and

known as "vertige paralysant." It is seen also in Japan and called Kubisagari. The disease is characterized by a series of sudden attacks of short duration, consisting of vertigo, somnolence, ptosis, pains in the cervical region and palsy of the neck muscles, causing the head to drop forward. There may be general weakness and paralysis, with peripheral speech disturbance; consciousness is not lost. The seizures last a few minutes, occur only in hot weather and may be repeated every few hours. The patients may be quite well in the interim. The onset, duration and symptoms suggest the Meniere disease.

Voltolini has described an affection which he looks upon as primary labyrinthine, and which strongly resembles Meniere. Its onset is with high temperature, general cerebral symptoms, with vertigo and vomiting. There is obscuration of consciousness. In a few days the symptoms subside, leaving a staggering gait, vertigo and deafness. The latter remains in some degree permanent. A plastic destructive exudate has been found in the semi-circular canals.

The Meniere triad is likewise observed as a symptom-complex in which the transitory vertigo is the marked feature, as a result of manipulation such as Eustachian tube catheterization, head galvanization, and even ear douching.

We have another category, which includes paroxysmal attacks—the so-called pseudo-Meniere seizure—observed in neurotics; in epileptics and during hysterical seizures; rarely, but at times, in the course of true neurasthenia and hemicrania.

In true Meniere disease the pathological basis is to be found in some form of labyrinthine disturbance. Hæmorrhage, inflammatory processes, intense hyperæmia, syphilitic disease, etc., are described. Local circulatory disturbance of the labyrinthine structure is emphasized by Oppenheim.

The several features of the disease are best understood by a brief analysis of the essential symptoms independently considered.

The deafness may affect one or both ears, and varies from slight impairment of hearing to absolute loss.

The vertigo is of variable intensity, though usually severe and most annoying. Its character is variously described. A common feeling among these patients is as though they were rapidly revolved first upon one axis, then on another; or again, as the sensation experienced upon a rapidly-revolving carousal or upon a ship tossed in a heavy sea. In some a true revolving movement, so-called circus motions, are seen.

The tinnitus likewise varies in its intensity. Many patients complain of this disturbance becoming rapidly more intense toward the approach of a paroxysm, and other are entirely free during intervals. The typical paroxysm in many cases begins with the tinnitus as a kind of aura, thus giving warning of an approaching seizure, much as the case in epileptic attacks. The subjective nature of the tinnitus varies. It may be of a buzzing character; like a violent windstorm; as though a large seashell were held close to the ear; the roaring of an immense waterfall, etc.

As is the case with all forms of subjective ear noise, more particularly in the

Meniere symptom patients, the aural sounds have been the developmental element in determining true hallucinatory impressions. I do not doubt, however, that these were cases established upon a paranoid soil.

The vomiting of the Meniere type is sometimes represented by a feeling of severe nausea, and only most exceptionally are all gastric manifestations absent. Though occasionally an early symptom, the vomiting appears as a rule at the height of the paroxysm. Again it may be the signal for the termination of the attack. The amount ejected and the presence or absence of gastric pain are variable. Typical projectile or cerebral vomiting with no nausea is observed. Imperative serous diarrhœa may attend an attack.

The degree of violence and suddenness of onset is very variable. The attack may be so immediate as to lay the patient prostrate without warning and with the momentum of a blow. In this respect it reminds one further of the epileptic seizure. The approach may be somewhat gradual, enabling the victim to gain support or

lie down. The direction in which the patient falls may be uniform, but generally is not. As the violence of the seizure abates and the symptoms subside incoordination and ataxia of the lower limbs become evident. Indeed, the Romberg symptom and varying degrees of ataxia are commonly present as permanent symptoms, and are valuable diagnostic factors in interparoxysmal periods. Careful examination with proper tests will almost invariably reveal some degree of motor disturbance in the interim between the acute attacks. Far less frequently is some incoordination in the upper extremities found. A case is described of a physician whose handwriting was markedly changed by the disease.

The ocular symptoms are of interest. Nystagmus, double vision, narrowing of the visual fields, subjective sensations, such as zigzag figures, are often observed, and should be sought for. Closing of the eyes, fixing upon a near object, tightly closing the lids or compressing the eyeballs sometimes favorably influences the vertigo.

Consciousness is very rarely lost, and

only transitory disturbances are described. Vasomotor phenomena are among the accompanying manifestations. The pulse is accelerated, very rarely retarded. A nervous chill or tremor may initiate the attack, and a sudden congestion of the face is frequently seen.

A severe degree of true neurasthenia often develops as the result of frequent attacks of Meniere symptoms. It is, however, a fact that the victims of this disease do not belong to the recognized neurotic type.

As might be expected from the variety of forms and because of a want of unanimity of opinion as to what constitutes true Meniere disease, the pathological findings have not been uniform. In the generally accepted form of Meniere, however, hæmorrhage or inflammatory labyrinthine disease has been regularly found.

Experimental investigations in lost function of the labyrinth in lower animals has produced a picture in many respects analogous to true Meniere disturbance in man. Admitting that clinical observation and experimental control point strongly to labyrinthine disturbance as

the seat of lesion, I incline to the belief that the occasional remarkable manifestations seen in Meniere disease indicate a pathological condition of more extensive involvement. It is, however, not improbable when one considers the nerve association between this part of the auditory apparatus and centers of important function in the central nervous system that the peripheral lesion may induce the severe train of symptoms seen in Meniere's disease.

As to the prognosis of these cases of Meniere's disease, experience varies. The individual symptoms as a rule become ameliorated after some years. The nervous system, however, is invariably left with some traces of exhaustion, and a varying degree of neurasthenia remains. The outlook in the apoplectiform cases has been less favorable, many having died soon after the initial attack. As a rule, the defect of hearing is in some degree permanent. The tinnitus becomes less severe in subsequent attacks. The vertigo also diminishes in intensity. The nature of the seizures often changes, the symptoms individually becoming milder.

With degeneration of the auditory nerve the paroxysms disappear. The average duration of the active disease is said to be about three years. Long periods of remission are frequent.

A brief sketch written by a sufferer of the disease so well describes her symptoms, which are quite typical, that I quote it here as an excellent illustration. The case was reported in 1898 in a paper by J. M. Brown and Judson Deland of Philadelphia. The patient wrote:

"I went to bed on the night of January 6 feeling well, but extremely tired, and the first premonition of approaching illness was on being awakened at 8 o'clock on the morning of the 7th, when I experienced the most violent noises in my head, like the pounding of iron. I had no dizziness or nausea during the afternoon, but on going downstairs received a slight physical shock as if the steps wavered and came in contact with the feet too suddenly. This continued all the morning, and was accompanied by a feeling of great lassitude. I ate heartily at luncheon, had no pain in my head or elsewhere, although the noises continued

without interruption; if anything, increasing in violence. A loud noise, such as the water rushing in the toilet-room, seemed to aggravate this condition, filling the head with noise. At about 1.30 P. M. I was seized with the acute attack of vertigo. The room suddenly and with great violence seemed to turn directly upside down, the floor approaching me giving the sensation of an actual shock as if caused by an earthquake. I staggered to the door with my feet wide apart like an intoxicated person and called for assistance, and fell into a chair, and life seemed to leave me for an instant. A glass of wine restored me temporarily, and I reached the bed without assistance and lay down. Upon moving my feet on the bed I experienced a slight sensation of the bodily physical shock before produced on going downstairs to be conveniently near some member of the family. About an hour later I was seized with the second attack of vomiting, this time accompanied by the most awful dizziness imaginable. The doctor likens this condition to seasickness, but I am sure that no seasickness could be half so terrible.

The nausea, vertigo and vomiting continued during the afternoon, and were intensified by the slightest movement of any part of my body, the room seeming to spin around and around with great velocity. I was utterly unable to rise to return to my room, and was finally carried there in great distress from the disturbance and change of position. Doctor called again at 7 o'clock in the evening and ordered absolute quiet, and I retired and slept heavily until morning. Upon awakening all my symptoms returned with increased force, the slightest movement of any portion of my body, excepting my hands, producing the most deathly nausea and vertigo, together with a feeling as if I were falling to the affected side (the left), while the room would seem to revolve rapidly in the same direction and the floor or objects under or below me seemed to rise up violently against me.

"These symptoms, as before, were increased by any movement of my extremities, even the simple movement of the eyeballs being sufficient to produce it, and it was only by maintaining an im-

movable position on my left side with my eyes closed that I enjoyed the least immunity from these distressing feelings. They continued without abatement for a week or eight days, then gradual amelioration took place, though several days elapsed before I was enabled to actually change my position on the bed; this was done by keeping the upper part of my body thoroughly rigid and horizontal, the least raising of the head or trunk instantly producing the feeling of shock and dizziness, yet without such violent recurrence of nausea. The inhalation of ammonia salts during these efforts at change of posture seemed to partially restrain the distress, especialy the nausea. After the first week the vomiting ceased, and in the next few days some abatement in the violence of the shock recurred as if the bed rose violently against me. This lasted during the entire three weeks I was in bed. In disappearing it seemed to leave the various parts of my body by degrees, beginning at the extremities and approaching my head, the last place to subside being the affected side between shoulder and hip, where I still periodic-

ally notice it. On moving across it the bed seemed to rise like the ocean billows carrying me on their crest. As the vertigo grew milder the violence of this motion and the excursion of distant objects seen through my windows first grew less and less; nearer objects, as the confines of my room and furniture, later, and finally my bed on which I rested. When convalescence had proceeded far enough and I could get up, I staggered considerably and pitched forward to the right or unaffected side. I was obliged to help myself by the bed and furniture, fearing to trust myself to others.

"At the present time, six months since I was stricken, the noise continues with varying intensity, ranging from those heard on a quiet summer night to those heard in a foundry. It was much less when lying on the affected side; and when lying on the other side, no matter what they may have been during the day or when up and about, they resemble the chiming of bells, and sometimes like the shifting of sand or rubbing of sandpaper. External noises when near me sound abnormally loud, as, for instance, persons

coughing or the tearing of paper, which are almost painful and startling. I am unable to distinguish the ticking of a watch when held close to the affected ear."