Tinnitus and myoclonus

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Objective tinnitus with palatal myoclonus was already described more than a century ago by Bigelow and Holmes. Up to 1961 Pulec and Simonton (6) collected 170 cases from the literature.

Objective tinnitus: a noise, a murmur or clicking coming from a patient's ear and audible for the examiner, is relatively rare. Part of the case occurs with palatal myoclonus, sometimes called clonical spasm or palatal nystagmus. Another part is caused by murmurs of the arteriovenous anastomoses or aneurysms somewhere nearby the ear. Fowler (3) divides the tinnitus into vibratory and non-vibratory. To the vibratory form belong the muscular, vascular and mucous sounds. These are objectively audible. To the non-vibratory tinnitus belong those noises as found with otosclerosis, labyrinthitis, middle ear processes and irritations of the acoustic nerve. These are only subjectively audible and generally continuous. A tinnitus can be temporary, continuous, rhythmic, irregular, soft or loud. The characteristics of the vibratory tinnitus are dependent on the origin of the sound. The tinnitus that occurs with palatal myoclonus need not be equal in frequency to the number of contractions of the clonus. According to some authors this tinnitus is caused by muscular contraction (muscle sound) of the stapedius muscle, or of the tensor tympani muscle or of the bigger muscles of the Eustachian tube (tensor veli palatini muscle and levator palatini muscle).

Others are of the opinion that the tinnitus is dependent on the opening or closing of the Eustachian tube.

Hammelburg (4) published a case of objective tinnitus in a patient with a chronic otitis media. The sound was caused by air, penetrating in the middle ear due to perforation of the drum membrane, preceded by discharging exudate.

In 1954 Versteegh, Jr. (9) published a case where a patient showed an objective tinnitus without myoclonus after a barotrauma. On account of the fact that the tinnitus was bilateral and stopped when he was asleep or under anaesthetics, the diagnosis of neurosis was considered very likely.

A clonus, according to Dobson and Riley (2), is a spontaneous, pretty regularly recurring muscular contraction without opposition or counteraction from an antagonist. The clonus is not felt by the patient.

Records of elderly patients (60–80 years old), with tinnitus and myoclonus generally show a cerebral accident in the clinical history or (and) other neurological symptoms such as hemiplegia (paresis) spasm, speech

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defects (aphonia), nystagmus, hypertension, etc., but rarely an objective tinnitus.

In addition to clonus of the soft palate, the palatine arches, the posterior wall of the pharynx, the tongue, the larynx, and the vocal chords, the muscles of the floor of the mouth, the neck- and inter-costal muscles, and the diaphragm may also be affected. The lowest number of contractions per minute is 10 and the highest about 300. The cause of the myoclonus is often not to be found. In the literature many causes have been mentioned, e.g. trauma capitis (so-called Whiplash head injury), injuries in the fossa posterior, influenza, operative treatment in the ear, nose and throat region, multiple sclerosis, encephalitis, shock, psychogenic factors, brain tumours, aneurysms in the brain, etc.

Attempts at recording the clonus were made by Chadwick and Macbeth (1) and Puleck and Simonton (the latter two placed an electrode in the Eustachian tube and recorded outbursts of motor unit activity during $\frac{1}{3}$–$1\frac{1}{2}$ seconds with intervals of $\frac{1}{5}$–2 seconds). The click auscultated by means of a stethoscope was heard one second after the beginning of the electric activity. They also recorded the pressure fluctuations in a tiny balloon placed inside the Eustachian tube. Their conclusion was that the click was caused by collapsing of the wall of the tube.

As early as 1867 Schwartze (7) saw movements of the central and posterior part of the eardrum in a patient suffering from myoclonus, and attributed them to contractions of the tensor tympani muscle.

Between 1854 and 1904, in publications of 55 cases of palatal myoclonus with objective tinnitus, 8 times movements of the drum membrane were found, five of them in the form of retractions. Wirth (8) used the name of 'Trommelfell Nystagmus'.

Klein (1907) found in two autopsy cases of patients with objective tinnitus and palatal myoclonus changes in the region of the inferior olivary nucleus.

Nathanson (5) published 12 cases of palatal myoclonus without objective tinnitus. Autopsy performed in two cases showed changes in the cerebellum and brainstem.

Weinstein and Bender, according to Nathanson, in their experiments with monkeys, have been able to induce myoclonic contractions of the palate, the tongue, the lips, etc., by means of an electric stimulus in the region of the hilus of inferior olivary nucleus. Bender and Teng caused a typically persisting palatal clonus in the case of a monkey by destruction in the region of the inferior olivary nucleus.

Personal observations

Patient 1, Miss V. G., born 31 August 1914, was complaining for about a year of ticking in the right ear that started after syringing the ear. Two years before she consulted our out-patients department as she had suffered a cranial

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trauma when diving in a swimming-pool. Now she complained of a full feeling and an irregular ticking sound in the right ear, she had no tinnitus, no loss of hearing, and no mouth, nose, or throat troubles. Nose and nasal cavities, as well as the eardrum, were normal. There were no movements of the drum membranes to be seen, not even under the microscope. The audiogram was normal, the Rinne on both sides positive. The ticking sound was audible at a distance of two yards in a quiet room. A further examination showed a palatal myoclonus, myorhythmies of the palatine arches, twitches of the tongue, clonus of the floor of the mouth, and slight movements of the vocal chords. An X-ray examination showed no clonus of the diaphragm. The frequency of the clonus was between 50 and 60 contractions a minute.

The neurological examination (van Vliet) did not produce an organic genesis for these contractions. The laboratory tests were normal. Paracentesis, Politzer manœuvre and catheterization of the right tube had no effect on the symptoms. When pressing a finger on the right half of the soft palate in the direction of the opening of the tube, the sound disappeared whereas the muscle contractions could be normally felt. On 16 January 1969 middle ear aeration on the right was performed under general anaesthesia.

After the operation the patient had no longer subjective or objective tinnitus, the palatal myoclonus was no longer perceptible, the myorhythmies of the pharyngeal arches and the root of the tongue, though less strong, still existed. The clonus of the floor of the mouth was also decreased in frequency. After three months the drain-tube in the drum membrane was ejected. The patient at that time did not show any objective or subjective abnormalities. This condition has continued up till now.

Patient 2, Miss R., born 25 July 1946, was complaining for about three to four years of a regular clicking sound in both ears, that had started after a serious cold of some months' duration. She was also a little deaf at that time. By examination we found that the eardrums were undamaged, the reflexes normal. Under a microscope the drums showed pit-shaped retractive movements at the posterior half, whereas the anterior part, the handle of the hammer, and the flaccid part showed no movement whatsoever. The tube function right and left was normal. A movement of the drum membrane, a clicking sound and a contraction of the palate occurred at the same moment. The movement of the drum membrane was due to the simultaneous contraction of the tensor veli palatini and the levator veli palatini muscles. The tensor veli palatini muscle drew the tube walls apart; the levator veli palatini muscle, in the beginning of its contraction, acted synergistically, but with a powerful contraction its medial end would go up so far that the pharyngeal opening of the Eustachian tube was closed. The result was lowered pressure in the tube and tympanic cavity, which caused the retraction. As the clicking coincided with the retracting movement of the drum membrane, one may conclude that the sound was caused by the drawing apart of the E. tube walls. By this patient too the click stopped when a finger was pressed against the soft palate in the direction of the pharyngeal opening of the Eustachian tube. The tinnitus was audible at a distance of about a yard. This patient had only a palatal clonus. No myorhythmies were visible. The clonus was not regular, the frequency was about 70 per minute. The result

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of a neurological examination (van Vliet) was as follows: The medical history together with paraesthesia and difference in reflex of the arms indicated that the palatal myoclonus might be a result of an injury in the reticular substance of the medulla oblongata or pons, e.g. an encephalo-myelitis disseminata.

The routine laboratory tests did not show anything particular. This patient was also treated with middle ear aeration. On 21 October 1969, the right ear was operated on under general anaesthesia.

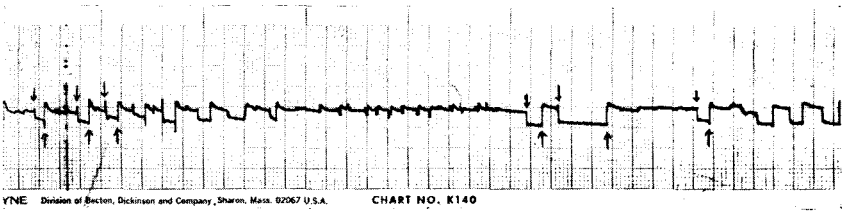


FIG. 1.

Tympanogram of the left ear of patient 2 made with the impedance-bridge. The space between an arrow downward and an arrow upward correlates with the retractive movement of the eardrum.

During the anaesthesia, the clonus, the movement of the eardrum and the sound stopped. After the operation the symptoms of the right ear were gone, whereas the palatal clonus and the symptoms of left ear remained unchanged. Two days later the sound in the right ear returned, though it was very irregular and lower in frequency than in the left ear. This continued up till 18 November 1969. On this day the left ear was operated (middle ear aeration). During the anaesthesia all symptoms were gone and did not return after the end of the operation.

Summary

One might divide the palatal myoclonus into groups according to the aetiology:

(*a*) the group, presumably resulting from intracranial inflammation, neurosis and tumours; in these cases often an objective tinnitus is found;

(*b*) the group, resulting mostly from circulation or vascular disturbances in cerebro. There is no objective tinnitus in these cases; the age is generally over 50.

Possibly palatal myoclonus occurs more often than is generally observed, as these movements sometimes stop when the mouth is opened.

Two patients, both women, ages 28 and 24 years, with objective tinnitus and myoclonus of the soft palate, were examined and treated. The first case had also myorhythmic movements of the pillars of the soft palate and the tongue and clonus of the floor of the mouth and vocal cords. The second case exhibited movements of the posterior part of both tympanic membranes. This movement is believed due to a negative

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pressure in the tympanic cavity and Eustachian tube caused by the closing of the tubal orifice by the musculus levator veli palatini, while the m.tensor opens the tube. The clicking sound must in this case be produced by the tube snapping open.

The patients were treated by middle ear aeration. Both patients were free of symptoms after the operation and stayed this way even after ejecting the grommet in the eardrums a few months later.

The results we have obtained by ventilation of the middle ear in our two cases may support the theory of Purves-Stewart (10) and we therefore also believe that one of the causes of this syndrome may be a hypersensitive reflex arch, caused by a lack of the regulating activity of the pyramidal tract. The stimulus of the reflex may have its origin in the nasopharynx, the Eustachian tube or middle ear.

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