CLINICAL VIGNETTE

ACHOO Syndrome

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An 85-year-old male with a history of uncontrollable consecutive sneezing episodes, presents with recent worsening bouts. Symptoms are episodic, occurring about once daily, and characterized by sneezing in continuous chain up to fifty times. He is only able to stop the sneezing by totally blocking all light from view and laying down. While doing this with his eyes closed, he will see a red-orange light that fades into a light blue color, then white, and finally to black. Only after this does the sneezing finally stop. He also has no pain nor fever or any other upper respiratory infection symptoms. His past medical history is otherwise negative for any pulmonary or cardiovascular conditions and he has no allergies. He finds the episodes exhausting and annoying. With the current COVID-19 pandemic, he is especially concerned about having an episode in public and causing alarm. His physical exam includes normal vital signs with BP 129/73, HR 60, T 97.3F, O2 98%. He is well developed and well nourished, and in no distress. HEENT findings include presence of right hearing aid and glasses. Lungs are clear to auscultation.

Discussion

The act or noise of sneezing, or *sternutation*, is the expulsion of air from the lungs through the nose and mouth, most commonly caused by the irritation of the nasal mucosa.[1] It can be triggered by sudden exposure to bright light, a full stomach, or physical stimulants of the trigeminal nerve. The sneeze reflex may be divided into two phases. The first is a nasal or sensitive phase, following stimulation of the nasal mucosa by a chemical or physical irritant. The signal is transmitted via branches of the trigeminal nerve to the trigeminal ganglion, with stimuli reaching the sneezing center in the lateral medulla. The second phase begins once a critical number of inspiratory and expiratory neurons has been recruited, which consists of eye closing, deep inspiration and then a forced expiration with initial closing of the glottis, and increasing intrapulmonary pressure. The subsequent dilatation of the glottis gives rise to an explosive exit of air through the mouse and nose, eliminating the mucosal irritant.[1] It is estimated that approximately 40,000 particles are expelled during a forceful sneeze, the particle size ranging from 0.5 to 5 microns.[1]

Autosomal Cholinergic Helio-Ophthalmic Outburst, or *ACHOO* Syndrome is characterized by an episode of uncontrollable sneezing in response to the sudden exposure to any bright light, such as but not limited to intense sunlight.[2] The sneezes generally occur in bursts of 1 to 10 sneezes, followed by a refractory period. This type of sneezing is also known as *photic sneeze reflex*. It is shown to have an autosomal dominant inheritance pattern and is assumed to affect 17 – 35% of the world population. Approximately one of every four individuals will have this response to bright light exposure, eliciting a sneeze or prickling sensation.[2] The reflex can only be triggered after the first exposure to light, and not on repetitive stimulation, and many reports cite a refractory period before the reflex can be elicited suggesting that a polysynaptic pathway is involved. The cause is not clearly understood, but may involve an over-excitability of the visual cortex in response to light, leading to a stronger activation of the secondary somatosensory areas.[2] This theory suggests the stimulation of the optic nerve triggers the trigeminal nerve. The afferent impulses of the pupillary light reflex are transmitted via the optic nerve while the efferent impulses are transmitted via the oculomotor nerve. An indirect impulse is transmitted to the ophthalmic division of the trigeminal nerve, and this impulse generates the nasal stimulation causing the sneezing by affecting the maxillary division of the trigeminal nerve. Another hypothesis is that there is a "crossing" of pathways in the brain between the pupillary light reflex arc and the sneezing reflex arc.[1] Light falling on the retina stimulates afferent fibers to the pretectal nuclei, which then send interneurons to the Edinger-Westphal nuclei. The parasympathetic fibers of the Edinger-Westphal nuclei and the trigeminal afferent fibers from the cornea both pass through the ciliary ganglion, where they may participate in transmission. Parasympathetic generalization may also contribute to photic sneeze. Stimuli that excites primarily one branch of the parasympathetic nervous system activates other branches. Thus, the parasympathetic branches of the oculomotor nerve activated to generate pupillary constriction against the bright light cause secretion and congestion in the nasal mucosa by triggering the parasympathetic activation by the pterygopalatine ganglion and a sneeze is triggered.[3]

Recommendation

There are no specific recommendations to manage photic sneeze reflex. Antihistamines used to treat rhinitis in seasonal allergies may also reduce the occurrence of photic sneezes in patients affected by both conditions. A military report demonstrated that interference light filters were ineffective, suggesting that photic sneeze reflex appears to be mediated by changes in light intensity rather than by specific wavelength of light. That being said, those affected by photic sneezing may find relief by shielding eyes and/or faces with hats, scarves, and sunglasses. One technique to try, offered by patient at a suburban ophthalmology practice is to use the Philtral Pressure Technique.[4] This

involves firm digital pressure applied by the patients index finger transversely to the skin of the sub-philtral region, directed posterosuperiorly onto the maxilla. This was successful in preventing photic sneeze reflex.

REFERENCES

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