

Cerebellar ataxia with auditory neuropathy and vestibular areflexia. A new variant of CANVAS syndrome?

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Abstract

Cerebellar ataxia with neuropathy and vestibular areflexia syndrome (CANVAS) is a rare neurological syndrome of unknown etiology. The main clinical features include bilateral vestibulopathy, cerebellar ataxia and sensory neuropathy with normal auditory function. An abnormal visually enhanced vestibulo-ocular reflex is the hallmark of the disease. We present a case of 35-year-old male patient who has demonstrated bilateral rapidly progressive hearing loss that eventually ended in total hearing loss in both ears within few months. On audiologic evaluation the type of hearing loss was due to auditory neuropathy (abnormal auditory pathway with intact cochlear function). On examination ataxia of gait, normal knee and ankle reflexes, gaze-evoked downbeat nystagmus and abnormal visually enhanced vestibulo-ocular reflex were found. Brain magnetic resonance imaging revealed no abnormality. Vestibular function testing no response to ice caloric stimulation. Nerve conduction studies was completely normal. The course of illness was progressive with ataxic gait and unsteadiness as the most disabling symptoms.

Biography

Elsaeid M Alieldin is a scientist, audiovestibular medicine consultant, and academic leader. He is the Director of Cochlear implant advisory board at KAMC, in the holy Capitol Makkah, Saudi Arabia, a position he has held since January 2018. Prior to this position, he served as a Professor of Audiological Medicine, at the Audiology unit, Department of Otorhinolaryngology–Head and Neck Surgery in the Mansoura University School of Medicine and Audiologist-in-chief of The Mansoura University Hospital. During his 15-year tenure, he expanded annual research funding by more than half and increased clinical activity by more than 30 percent, while strengthening teaching efforts and student training. With more than 60 published articles and chapters, he is an expert in balance and hearing disorders.



15th International Conference on Otorhinolaryngology: ENT Surgery, December 09-10, 2021

Citation: Elsaeid M Alieldin, Cerebellar ataxia with auditory neuropathy and vestibular areflexia. A new variant of CANVAS syndrome?, Otorhinolaryngology 2021, 15th International Conference on Otorhinolaryngology: ENT Surgery, December 9th, 2021, Page-04