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Teaching Video NeuroImage: Alternating Skew Deviation as a Manifestation of Anti-GAD65–Associated Cerebellitis

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Article

A 53-year-old woman presented with ataxia, dysarthria, and vertical binocular diplopia on left and right gaze but absent in primary gaze. Her symptoms were progressive over several months without clear inciting event. Initial neuro-ophthalmology showed subtle downbeat nystagmus in primary gaze that increased in horizontal gaze, consistent with 'side-pocket' phenomenon.[1] On cross-cover testing she was found to have an alternating skew deviation, raising question of cerebellar localization (Video 1). Multiple etiologies can lead to ASD including autoimmune, ischemic and paraneoplastic entities. Prior structural imaging was unrevealing. Serum studies revealed elevated glutamic acid decarboxylase antibody (anti-GAD65) levels (>250.0 IU/mL, normal range 0-5.0 IU/mL). She was subsequently diagnosed with anti-GAD65 cerebellitis. The patient was not screened for a neoplasm as anti-GAD65 is rarely paraneoplastic in nature. Anti-GAD65 interferes with the production of GABA, thereby disrupting supranuclear pathways and has been associated with autoimmune epilepsy and stiff-person syndrome. Intravenous immunoglobulin may improve outcome.[2]

Video: Alternating Skew Deviation in anti-GAD65 Cerebellitis

This patient has an alternating skew deviation with 8 prism diopters right hypertropia in right gaze and 8 prism diopters left hypertropia in left gaze.

http://links.lww.com/WNL/C557

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