Dysphagia in hereditary spastic paraplegia (HSP)

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Abstract

Dysphagia is a frequent symptom in many neurological diseases. Neurogenic dysphagia is associated with reduced quality of life, malnutrition, aspiration pneumonia and poor outcome. Therefore, early detection and adequate management of swallowing impairment is mandatory. In hereditary spastic paraplegia (HSP) dysphagia seems to be a rare and often mild symptom. Swallowing disorder has been reported in about 5% of pure and up to 30% of complicated HSP patients. Dysphagia occurs preferably in SPG5-, SPG7- and SPG11-HSP. In cases with genetically unconfirmed HSP and prominent dysphagia other causes should be considered by clinicians such as amyotrophic lateral sclerosis, primary lateral sclerosis or spinocerebellar ataxia type 3.

Fiberoptic endoscopic evaluation of swallowing (FEES) is a valuable tool for objective swallowing assessment. By using FEES we were able to detect different forms of HSP related dysphagia. In a 22-year-old-female suffering from SPG11-HSP dysphagia was due to pharyngeal spasticity leading to mild to moderate residues in the valleculae and base of tongue. In contrast, FEES examinations in two siblings (33-year-old-female and her 43-year-old-brother) with SPG7-HSP revealed dysphagia that was characterized by leakage to the pyriforms before the swallow was initiated resulting in an increased risk of predeglutitive aspiration. This form of dysphagia was probably caused by cognitive impairment, i.e. dysexecutive syndrome, similar to swallowing disturbance described in patients with frontotemporal dementia. In conclusion, FEES should be used to assess the nature of dysphagia in HSP patients and to guide appropriate treatment strategies.