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孤発性脊髄小脳変性症の病型による嚥下造影所見の比較

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Characteristics of the videofluorography of patients with sporadic forms of spinocerebellar degeneration (SCD) – Comparison among SCD subtypes –

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要旨

脊髄小脳変性症（SCD）には，摂食・嚥下障害が高率に発症し，療養生活の中で対応に難渋する症状の一つである．しかし，その病態について十分な検討を行った報告は少ない．特に病型による摂食・嚥下障害の病態の違いについての検討はなされていない．本研究では，孤発性 SCD のうち，SCD 重症度がほぼ同程度の晩発性小脳皮質萎縮症 3 例（LCCA）（年齢 57 歳～81 歳），オリーブ橋小脳萎縮症 3 例（MSA-C）（年齢 54 歳～64 歳），線条体黒質変性症 3 例（MSA-P）（年齢 57 歳～75 歳）に対し，摂食・嚥下機能評価について比較検討を行った．本研究ではビデオ嚥下造影検査（VF）における定性的評価として，口腔内保持不良，口腔内残留，喉頭侵入および誤嚥，咽頭残留の有無を評価し，定量的評価として，時相解析の口腔移送時間，嚥下反射開始時間，咽頭通過時間，舌骨挙上時間と嚥下時の舌骨移動距離を測定した．定性的評価では，MSA-P において口腔内保持不良，咽頭残留，喉頭侵入および誤嚥が最もよくみられた．定量的評価では，LCCA，MSA-C，MSA-P の順に嚥下反射開始時間，咽頭通過時間が延長し，MSA-P では口腔移送時間が最も延長していた．舌骨移動距離では，LCCA，MSA-C，MSA-P の順に前方移動距離が短縮していた．少数例ではあるが，SCD 重症度がほぼ同程度でも病型によって嚥下動態の重症度が異なり，3 病型の中では，MAS-P が最も重症で，LCCA が嚥下障害の軽度な病型であることが，客観的評価により示された．病態の特徴として，嚥下惹起時間が延長し，舌骨の前方移動が短縮していることが，食塊の咽頭残留を引き起こすと考えられた．鳥取臨床科学 2(1), 51-62, 2009

Abstract

In patients having spinocerebellar degeneration (SCD), the disturbance of their swallowing, referred to as

“dysphagia”, is frequently observed. This is one of the manifestations causing difficulties in care for their daily living. However, few studies have revealed the pathological mechanisms underlying dysphagia in SCD patients, and almost no studies on the differences among sporadic forms of SCD patients with three subtypes: late cortical cerebellar atrophy, and the olivopontocerebellar atrophy subtype and striatonigral degeneration subtype of multiple system atrophy (MSA), referred to as LCCA, MSA-C and MSA-P, respectively. In the present study, we compared the characteristics of the impaired swallowing function in three cases each of the three subtypes of SCD patients. Patients with LCCA, MSA-C and MSA-P were aged 57 – 81, 54 – 64, and 57 – 75, respectively. The severity of their cerebellar and parkinsonian features was almost the same among the three subtypes, although LCCA patients exhibited no parkinsonian features. Their swallowing function was investigated using videofluorography (VF). Function was shown qualitatively with VF as impaired holding and residue of a food bolus within the oral cavity; inspiratory sucking of a food bolus into the vestibulum laryngis, and beyond the vocal cord, referred to as penetration and aspiration, respectively; and a residual bolus in the pharynx. Quantitative measures of function using VF consisted of temporal analyses, including the oral transit duration (OTD), stage transition duration (STD), pharyngeal transit duration (PTD), and pharyngeal response duration (PRD); and anterior and superior displacement of the hyoid bone. With qualitative evaluation, impaired holding of a food bolus within the oral cavity, penetration and aspiration of a food bolus, and a residual bolus in the pharynx were more frequently observed in patients with MSA-P than in he other two subtypes. Quantitative assessment demonstrated the values of OTD and PTD in the three subtypes of SCD patients, in the following order: MSA-P>MSA-C>LCCA; and the highest value of OTD in MSA-P patients. It also showed the distance of anterior displacement of the hyoid bone in the three subtypes of SCD patients, in the following order: LCCA>MSA-C>MSA-P. This indicated that the anterior displacement in MSA-P patients was shorter than the other two subtypes. Although we studied a small number of SCD cases, our study demonstrated the different severity and characteristics of the impaired swallowing function among the three subtypes of SCD patients, even if the severity of cerebellar features was not different. Among the three subtypes, patients with MSA-P exhibited the most severe impairment, while those with the LCCA had the mildest impairment in the swallowing function. Taken together with our data, particularly in MSA-P cases, the delayed STD and shortened anterior displacement of the hyoid bone may induce a residual food bolus in the pharynx. *Tottori J. Clin. Res.* 2(1), 51-62, 2009

Key words: 脊髄小脳変性症, 多系統萎縮症, 晩発性小脳皮質萎縮症, 嚥下障害, 嚥下造影, 嚥下惹起時間, 舌骨移動距離; spinocerebellar degeneration (SCD), multiple system atrophy (MSA), late cortical cerebellar atrophy (LCCA), dysphagia, videofluorography, stage transition duration (STD, time for triggering swallowing reflex), anterior displacement of the hyoid bone

I. 緒言

日本での脊髄小脳変性症（以下 SCD）は 10 万人あたり 7〜10 人とされており，運動失調を主要症候とする原因不明の神経変性疾患の総称である．全体の約 40%が遺伝性で，残りの約 60%が遺伝性のない孤発性とされている [1]．孤発性は純粋小脳失調症型の晩発性小脳皮質萎縮症（以下 LCCA）と多系統障害型（以下 MSA）に大別され，MSA はオリーブ橋小脳萎縮症（以下 MSA-C），線条体黒質変性症（以下 MSA-P），Shy-Drager 症候群に分類されている．

SCD には，運動失調だけでなく，摂食・嚥下障害が頻発であり，療養生活の中で患者・家族の quality of life（QOL）を著しく低下させる．金