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Case Report

# Swallowing Therapy for Systemic Amyloidosis-induced Dysphagia: A Case Report

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Amyloidosis is defined as the extracellular deposition of fibrillar proteins in different organs and tissues. The clinical features of amyloidosis include cardiomyopathy, hepatomegaly, proteinuria, macroglossia, and dysphagia. We report the case of a 52-year-old man with myeloma-associated amyloidosis who complained of progressive dysphagia, dysarthria, and macroglossia. Videofluoroscopic swallowing study revealed severe reduction in tongue movements, aspiration after swallowing, and retention in the valleculae and pyriform sinuses. The patient underwent swallowing therapy for 6 months; the therapy included compensatory neck flexion, vocal cord adduction exercises, supraglottic swallow, and the Mendelsohn maneuver. The swallowing therapy enabled the patient to swallow liquid and soft solids without choking. Thus, swallowing therapy is helpful and improves the quality of life of patients with severe dysphagia induced by amyloidosis. ( Tw J Phys Med Rehabil 2011; 39(1): 25 - 29 )

**Key Words:** amyloidosis, macroglossia, dysphagia, swallowing therapy

## INTRODUCTION

Amyloidosis is defined as the extracellular deposition of fibrillar proteins (amyloid) in different organs and tissues.[1,2] Amyloid deposits stained with Congo red appear red under normal light and apple-green under polarized light;[1,3] this distinction is the gold standard for the diagnosis of amyloid deposits.[4] Amyloidosis is classified on the basis of the type of amyloid-precursor proteins that form the deposition. There are two main types: systemic and localized. The systemic amyloidosis is subclassified as primary and secondary.[1]

Primary (amyloid light chain, AL) amyloidosis is the most common form of amyloidosis. Excessive light chains are deposited systemically. Multiple myeloma is present in 15% of the patients with primary amyloidosis.[2]

Secondary (amyloid associated, AA) amyloidosis is associated with infectious, inflammatory, or less commonly neoplastic disorders.[2]

Upper aerodigestive tract involvement is observed in about 10-20% of amyloidosis patients. Amyloid deposits in the upper aerodigestive tract can lead to severe dysphagia,[2] which may induce many complications, including aspiration pneumonia, malnutrition, and dehydration.[5] We report the case of a man with amyloidosis who developed severe dysphagia and showed an improvement in swallowing function after swallowing therapy.

## CASE REPORT

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A 52-year-old man was diagnosed with multiple myeloma, and he had been receiving melphalan and prednisone treatment during the last 3 years. However, subsequent to the diagnosis of systemic amyloidosis, which was confirmed by Congo red staining (Figure 1), he developed progressive dysphagia while swallowing solid foods, dysarthria with changes in voice, and macroglossia. Weight loss of approximately 5 kilograms was noted. He also had a history of restrictive cardiomyopathy with myocardial infarction, hepatitis, chronic renal insufficiency and hypothyroidism without medical control.

The masticatory muscles were firm with cramps and the mouth opening was limited (less than 4 cm). Evaluation of speech and swallowing showed that the patient had an enlarged firm tongue (macroglossia) (Figure 2), slow and weak lingual movements, poor oral secretory function, and hoarseness with poor articulation. The patient showed delayed swallowing reflex with frequent choking. The laryngeal elevation was reduced and some residual food was observed in the oral cavity and pharynx. The dysphagia was classified as level 2 on the basis of the Dysphagia Outcome and Severity Scale (DOSS).[6] Nasopharyngoscopy revealed smooth and intact nasopharyngeal mucosa but incomplete glottic closure.

Videofluoroscopic swallowing study (VFSS) revealed severe reduction in tongue movements and poor ability to retain liquids in the oral cavity. The following abnormalities were observed during the pharyngeal phase: absence of swallowing reflex, nasal reflux, and retention of foods in the valleculae and pyriform sinuses (Figure 3). The anterior laryngeal elevation and pharyngeal peristalsis were reduced. Aspiration of thin liquids and solids (Figure 4) was also showed. Throat clearing was achieved by chin-tuck posture in VFSS.

The patient was referred to our clinic for swallowing therapy for malnutrition and frequent choking. He refused nasogastric tube placement and was fed under close supervision. The therapeutical goals were to increase oromotor function and prevent aspiration. The strategies of swallowing therapy included oromotor exercise, chin-tuck posture, supraglottic swallow and the Mendelsohn maneuver. Initially, we attempted to improve the tongue movement and increase the extent of mouth opening by oral massage and oromotor exercise. Upper airway aspiration was overcome by ensuring that the patient maintained the chin-tuck posture, resulting in decreased choking during swallowing. Vocal cord adduction exercises and supraglottic swallow were also introduced to ensure safe swallowing, and the laryngeal elevation was improved by the Mendelsohn maneuver.

After 6 months of swallowing therapy, muscle spasm and trismus improved (mouth opening increased about 5 mm). We observed an improvement in the tongue movement and a reduction of residue in the oral cavity. The patient was able to swallow liquids and soft solids without choking (DOSS, level 4). Clinically, no aspiration pneumonia occurred. Body weight gain (about 2 kg) was reported. However, 2 months later, the patient died from heart failure due to his underlying cardiomyopathy.

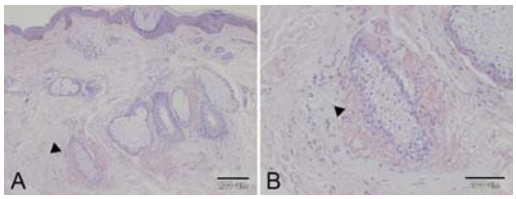


Figure 1. Congo red staining of the patient's skin biopsy. Amyloid appears as pink fibrils within the dermis.

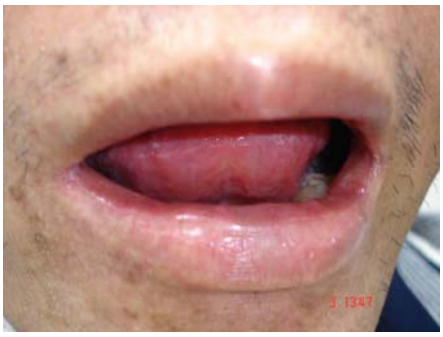

Figure 2. Macroglossia with limited mouth opening and tongue protrusion.

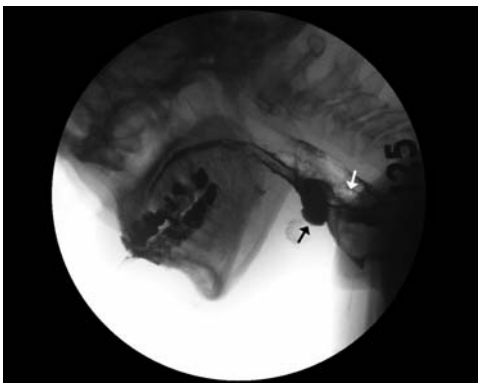

Figure 3. VFSS image showing significant food residue in the valleculae (black arrow) and pyriform sinuses (white arrow).

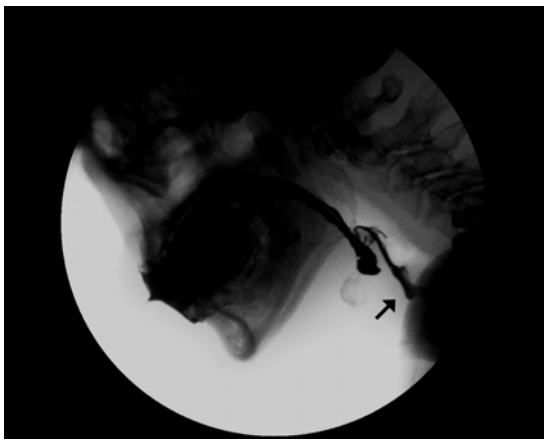

Figure 4. VFSS image showing aspiration, with food bolus landing in the open airway (black arrow).

## DISCUSSION

Cardiac involvement is present in 90% of the patients with multiple myeloma-associated amyloidosis.[7] The clinical manifestations of cardiac infiltration with amyloid deposits are myocardial dysfunction, conduction disturbances, and coronary vascular insufficiency. Chronic congestive heart failure occurs in up to 40% of the patients with multiple myeloma-associated amyloidosis. The median survival is only 6 months in patients with symptomatic heart involvement.[1] Death is often sudden or occurs due to rapidly progressing heart failure.[1] Our patient died due to cardiomyopathy-related heart failure but not due to the comorbidities associated with dysphagia.

Clevens et al[8] noted amyloidosis deposition throughout the upper aerodigestive tract, included to larynx, trachea and subglottis, base of tongue, oral cavity/ oropharynx and nose/nasopharynx. Stegemann et al[9] found an amyloid deposit eroding the vagus nerve. On pathology findings, Lavergne et al[10] reported the abnormalities of the motility of the digestive tract can be explained by either the muscular coat or the nervous plexus involvement by amyloid deposits. These findings may explain hypokinesis of esophagus and abnormality in the upper aerodigestive tract noted in VFSS. Unfortunately, no biopsy from the pharynx or the esophagus was performed to confirm the involvement of digestive tract by amyloid deposition in this case.

Amyloid infiltration of the upper aerodigestive tract leads to macroglossia, hardening of soft tissue, hoarseness, and decreased contractility of the esophagus, which causes speech difficulties and dysphagia.[2] In our patient, poor oromotor function with trismus, aspiration to liquids and solids and impaired laryngeal elevation with stasis were presented with chronic dysphagia. Swallowing therapy may play an important role in relieving this dysphagia.

Swallowing therapy can be administered via direct and indirect methods. Direct methods include tongue or larynx exercises and attempts to restore voluntary orofacial and laryngeal motor activity.[11,12] Indirect methods include compensatory mechanisms and the Mendelsohn maneuver, which are designed to overcome the consequences of functional impairment.[12]

According to the clinical evaluation and VFSS, the techniques of chin-tuck posture, supraglottic swallow and Mendelsohn maneuver were used to ensure safe swallowing and reduce stasis.

The position of the neck has a major influence on swallowing. Maintenance of the chin tuck posture before swallowing may be useful for patients with poor apposition of the soft palate to the tongue. Flexing the neck may also reduce aspiration by enlarging the valleculae and narrowing the laryngeal vestibule.[13] In our patient, the chin tuck posture was effective not only for nasal reflux before swallowing but also for aspiration after swallowing.

Supraglottic swallowing is another helpful maneuver for reducing aspiration. The patient inhales and holds his or her breath before putting food in the mouth and then ingests the food. After swallowing, the patient exhales forcefully and clears the larynx. This maneuver is very useful for patients with impaired laryngeal closure.[13] Our patient reported decreased choking during swallowing if supraglottic swallowing was used.

The suprahyoid muscles in healthy individuals show strong but brief contraction during swallowing. In patients with dysphagia, the Mendelsohn maneuver facilitates the elevation of the hyolaryngeal complex and maintains the pharyngoesophageal sphincter open by sustained voluntary contraction of the suprahyoid muscles.[13] Subjectively, reduction in sense of residual food in the pharynx was reported by our patient. No aspiration pneumonia occurred during the swallowing training course.

Our patient had received chemotherapy for multiple

myeloma-associated amyloidosis for 3 years and underwent swallowing therapy at our clinic for 6 months. Thus, DOSS was improved mainly by the swallowing therapy.

## CONCLUSION

In patients with amyloidosis-induced macroglossia and dysphagia, detailed pretreatment assessment may confirm the cause of dysphagia. Subsequently, individualized swallowing therapy should be developed to improve the swallowing function, nutrition, and quality of life of these patients.

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# 類澱粉沉積症併發吞嚥困難之吞嚥治療：病例報告

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類澱粉沉積症是一種 fibrillar protein 沉積在細胞外的疾病，會影響全身的器官與組織。臨床表現為心肌病變、肝腫大、蛋白尿、巨舌症及吞嚥困難等症狀。本病例報告一位診斷為類澱粉沉積症併有多發性骨髓癌的 52 歲男性，導致巨舌症及日見嚴重的吞嚥困難和發聲困難，轉介到復健科尋求治療。經 X 光透視螢光錄影吞嚥檢查發現，舌頭動作協調性差、食糰殘留在會厭鼓竇(valleculae)及梨狀竇(pyriform sinus)、明顯的嗆食(aspiration)現象。病人接受了六個月吞嚥訓練，包括擺位練習，加強聲帶閉合，上聲門吞嚥法(supraglottic swallow)及孟德森吞嚥法(Mendelsohn maneuver)等治療方式，已經可自行吃流質或軟質食物而沒有嗆到的情形。針對此病例若能完整評估吞嚥功能及給予適當的吞嚥訓練，應有助於改善吞嚥功能及生活品質。（台灣復健醫誌 2011；39(1)：25 - 29）

**關鍵詞：**類澱粉沉積症(amyloidosis)，巨舌症(macroglossia)，吞嚥困難(dysphagia)，吞嚥治療(swallowing therapy)

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