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Citation	The Laryngoscope, 118(11), pp.2057-2058; 2008
Issue Date	2008-11
URL	http://hdl.handle.net/10069/23070
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This document is downloaded at: 2012-05-17T04:07:17Z

**The Patulous Eustachian Tube Complicated with Amyotrophic Lateral Sclerosis:
A Video Clip Demonstration**

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Abstract

We report a 60-year-old case diagnosed as the amyotrophic lateral sclerosis (ALS) with patulous eustachian tube (ET). To our knowledge, this is the first case report of the ALS complicating patulous ET in the literature, demonstrating the movement of the tympanic membrane, the pharyngeal orifice of the ET and abnormal movement of the uvula due to paralysis of the soft palate on video.

Key words: amyotrophic lateral sclerosis, levator veli palatini muscles, patulous eustachian tube, tensor veli palatini muscle, tympanic membrane, video

Introduction

Amyotrophic lateral sclerosis (ALS) is a devastating, progressive motor-neuron disease characterized by the involvement of both upper and lower motor neurons. Upper motor neuron findings consist of spasticity and hyperreflexia, while lower motor neuron abnormalities include flaccidity, hyporeflexia, and muscle atrophy or fasciculation (1). When we, otolaryngologists, see the patients with ALS, we tend to pay attention only to their complaints of bulbar symptoms (2-4), but not to their ear symptoms. In the present report, we demonstrate the patients diagnosed as the patulous ET associated with ALS and review the literature.

Case Report

A 60-year-old man visited our outpatient clinic with a two year history of dysphagia. He was already diagnosed as an ALS by neurologists in our hospital one and half years ago. Because oral food intake became intolerable due to progressive aspiration, percutaneous endoscopic gastrostomy had been undertaken one month ago. He also complained of aural fullness and a flapping noise in his right ear

for the past several months.

On the physical examinations, fasciculation of the tongue and poor palatal movement on the right side were revealed. Nasopharyngeal endoscopy demonstrated insufficient elevation of the soft palate and inadequate closure of the nasopharynx on the right side when swallowing (velopharyngeal insufficiency, Video Clip 1), hence suggesting the atrophy of the levator veli palatini muscles (LVPM). A forty-five degree lateral-angle endoscopy showed that a pharyngeal orifice of the eustachian tube (ET) was wider on the right side than on the left side, plus the movement of the lateral wall when swallowing was poorer on the right side than on the left side, suggesting the atrophy of the tensor veli palatini muscles (TVPM) as well as the LVPM (Video Clip 2). Otoscopy revealed inward-and-outward movements of his tympanic membrane synchronized with his nasal respiration only on the right side (Video Clip 3). We diagnosed it as the patulous ET of the right side. We treated it by putting a thin chitin sheet on the postero-superior quadrant of his tympanic membrane and his discomfort in the right ear was reduced (5).

Discussion

ALS is a devastating illness for patients, and is also one of the most puzzling diseases in medicine in terms of our understanding of its pathogenesis (1).

Although otolaryngologists frequently evaluate patients with voice and/or swallowing troubles, ALS is infrequently encountered. When we see a patient with ALS, we tend to pay attention only to their complaints of bulbar symptoms such as dysarthria, dysphagia, and dysphonia (2-4), but not to their ear symptoms. In fact, there were few reports of ALS complicated with ear diseases in the literature (6).

On the other hand, the ET plays an important role in the ventilation, clearance, and protection of the middle ear (7). Among the muscles of the palate, the TVPM is known to open the ET (8). It is located laterally near the ET lumen, and attaches the lateral lamina of the ET cartilage and the anterolateral membranous wall of the ET lumen. The LVPM, originating from the inferior aspect of the petrous apex and forming a muscle sling at the soft palate, runs parallel and inferior to the ET lumen and cartilage. There has been a diversity of opinion concerning the role of the LVPM for ET function (9). In the present case, poor movement of the lateral wall of the right ET

suggesting the possible atrophy of the TVPM, and almost absent palatal movement suggesting the paralysis of the LVPM and possible subsequent atrophy of the LVPM were observed on the right side. From the fact that the patulous ET was revealed on the same side, it is interesting to speculate that the atrophy of the both TVPM and LVPM might be related to the cause of the patulous ET in the present case.

To our knowledge, there has been no report of the patulous ET associated with ALS. It is known that most patients with ALS undergo a phase of reactive depression after their diagnosis (1, 10, 11), and that the autophony on the patulous ET often leads to major depression and suicide also (12). Thus, the combination of these two diseases would be extremely painful for the present patient. Since only early precise diagnosis and treatment of this condition should lead to relief of his discomfort, we, clinicians, have to keep this rare combination of ALS and patulous ET in mind.

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Legend

Video Clip 1 shows that the endoscopic view of poor palatal movement on the right side and, the nasopharyngeal endoscopic view of the insufficient elevation of the soft palate and inadequate closure of the nasopharynx on the right side during swallowing.

Video Clip 2 demonstrates the forty-five degree lateral-angle endoscopic view of the right and left pharyngeal orifice of the eustachian tube (ET). The ET lumen on the right side was wider than on the left side, and the movement of the lateral wall of the ET lumen during swallowing on the right side is poorer than on the left side.

Video Clip 3 demonstrates the inward-and-outward movements of his tympanic membrane synchronized with his nasal respiration on the right side.