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"Frog Sign": Uncovered Condition of Long-term Non-invasive Positive Pressure Ventilation in Late-onset Pompe Disease

Junichiro Takahashi, Madoka Mori-Yoshimura* and Yuji Takahashi

Department of Neurology, National Center Hospital, National Center of Neurology and Psychiatry, Japan

***Corresponding author:** Madoka Mori-Yoshimura, Department of Neurology, National Center Hospital, National Center of Neurology and Psychiatry, 4-1-1 Ogawahigashi, Kodaira, Tokyo 187-8551, Japan. E-mail: yoshimur@ncnp.go.jp

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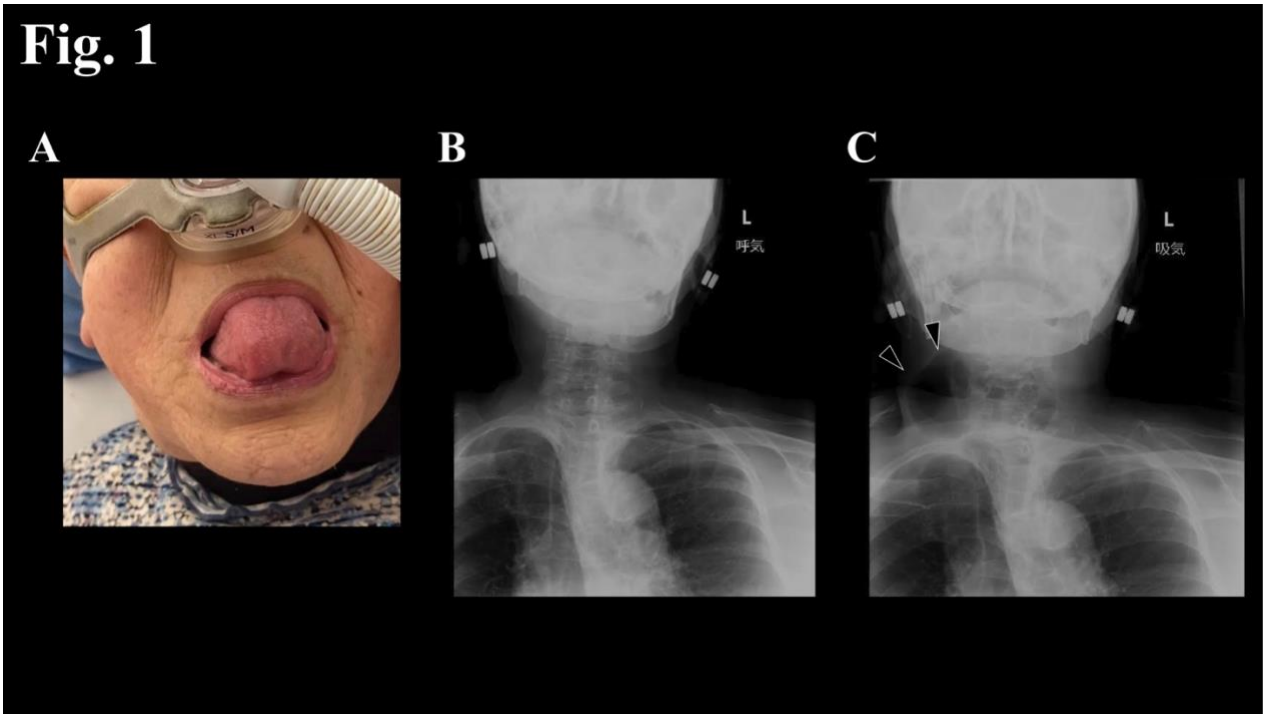

Figure 1 (A): Macroglossia in the patient; **(B):** Plain posteroanterior radiographs at expiratory phase, and **(C):** at inspiratory phase.

No abnormality was observed at expiratory phase (B), but an air-filled mass in the right, anterior, paramedian region was detected at inspiratory phase (C).

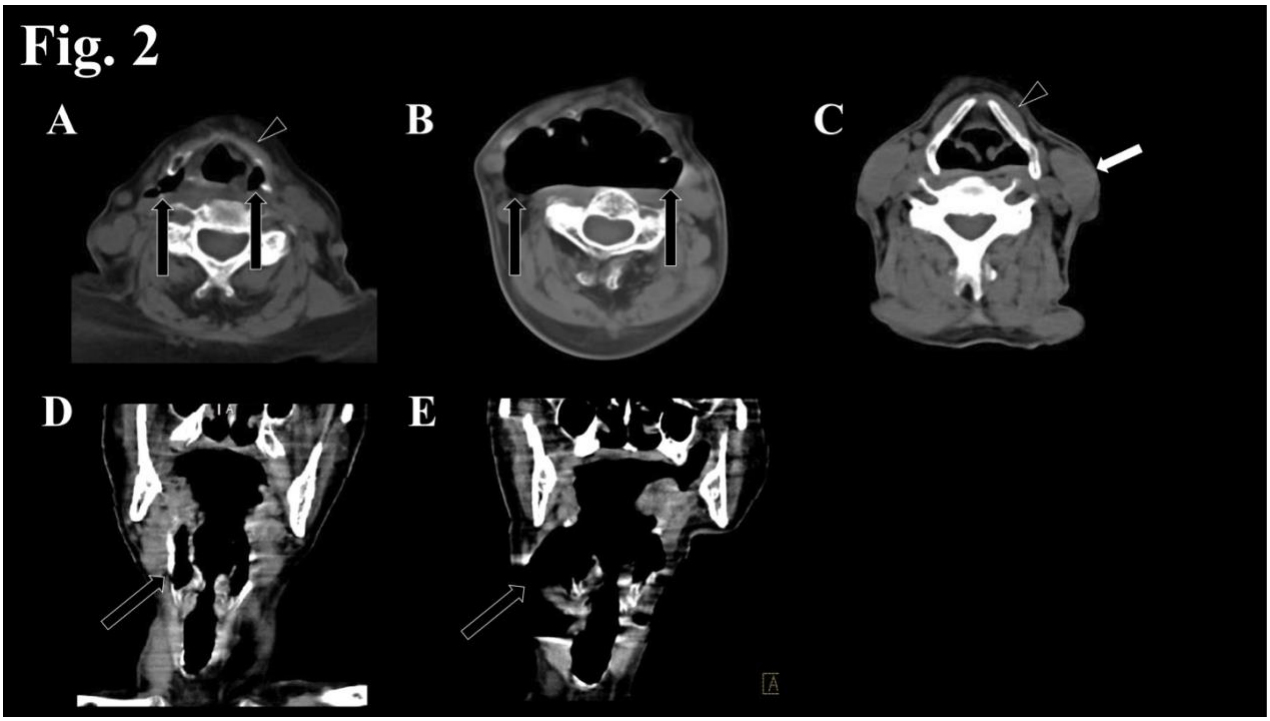

Figure 2 (A): Expiratory axial CT of the neck in the patient; **(B):** Inspiratory axial CT of the neck in the patient; **(C):** Expiratory axial CT of the neck in control patient; **(D):** Expiratory coronal CT of the neck in the patient; **(E):** Inspiratory coronal CT of the neck in the patient.

Neck CT at the expiratory phase showed dilation of both piriform fossae, more evident on the right side (A and D, black arrows), and further dilation at the inspiratory phase (B and E, black arrows). Compared to health control (C), neck muscles, such as sternocleidomastoid (C, white arrow), and infrahyoid muscles (A and C, black arrowheads) could not be detected due to severe atrophy in the patient.

Case Presentation

An 84-year-old female, genetically diagnosed with late-onset Pompe disease (LOPD), gradually presented with swelling on the right side of her neck during the inspiratory phase on non-invasive positive pressure ventilation (NPPV).

She required NPPV during day and night in her 70s with high end-expiratory positive airway pressure of 15 hPa, because of upper airway obstruction due to macroglossia (Figure 1A).

At her age of 82 years old, she noticed swelling on the right side of her neck during inspiratory phase on NPPV. Over two years, neck swelling was gradually increasing in size.

On physical examination, swelling on the right side of her neck was detected during inspiratory phase via nose, and rapidly returned at expiratory phase via mouth (Video 1), but slightly decreased in size via nose (Video 2).

Clinical Video: Videos are related to this article can be found online at:

https://www.literaturepublishers.org/archive/Frog-Sign:-Uncovered-Condition-of-Long-term-Non-invasive-Positive-Pressure-Ventilation-in-Late-onset-Pompe-Disease.html

Clinical Video 1: Inspiratory via nose, and expiatory via mouth. Swelling on the right side of her neck was detected during inspiratory phase, and disappeared at expiratory phase via mouth.

Clinical Video 2: Both of inspiratory and expiatory via nose. Similarly, swelling appeared on her neck during inspiratory phase, but slightly decreased in size and remained swollen even at expiratory phase via nose.

Plain posteroanterior radiographs at inspiratory and expiratory phase demonstrated an air-filled mass in the right, anterior, paramedian region (Figure 1B and 1C). Neck CT revealed severe atrophy of neck muscles, compared to control patient (Figure 2A and 2C). Even in expiratory phase, distended pyriform sinus with air-filled outpouching was observed (Figure 2A and 2D) and further extended across the thyrohyoid membrane inspiratory phase was detected (Figure 2B and 2E), compared to control (Figure 2C). A diagnosis of laryngocele, which is the dilation of the laryngeal saccule between false and true vocal cords, was confirmed.

The phenomenon was similar to how a frog expands its throat. Male frogs have a "vocal sac," a sound-resonating throat pouch. When they exhale with the mouth and nostrils closed, the vocal sac expands as its fills with air [1].

In the present case, the combination of dilated piriform fossae behaving like a "vocal sac" in frogs, increased elasticity of the throat wall with thinning tissue following severe muscle atrophy, and continuously high expiratory pressure due to upper airway obstruction due to macroglossia, may have contributed to this complication.

We named this finding the "Frog sign," and suggest including it among the list of complications that may occur in patients with LOPD receiving NPPV.

Keywords: Frog sign; Late-onset Pompe disease; Throat expansion; Non-invasive positive pressure ventilation

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Author's Contributions

- Junichiro Takahashi: Drafting/revision of the manuscript for content, including medical writing for content.
- Madoka Mori-Yoshimura: Drafting/revision of the manuscript for content, including medical writing for content.
- Yuji Takahashi: Drafting/revision of the manuscript for content, including medical writing for content.

Statement of Ethics

- Written informed consent was obtained from the patient for publication of this case report and any accompanying images.
- The study was approved by the National Center Hospital Ethics Committee (A2018-024).

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