

[CASE REPORT]

Brain Abscess Presenting as Prolonged Headache in a Patient with Amyotrophic Lateral Sclerosis under Mechanical Ventilation

Yoya Ono¹, Nobuaki Yoshikura¹, Akira Takekoshi¹, Naoyuki Ohe², Hisamitsu Hayashi³,
Megumi Yamada¹, Yuichi Hayashi¹, Akio Kimura¹ and Takayoshi Shimohata¹

Abstract:

A 57-year-old woman with amyotrophic lateral sclerosis (ALS) receiving mechanical ventilation developed intractable right temporal headache. She was diagnosed with brain abscess secondary to chronic suppurative otitis media. In this case, the otitis media was caused by nasopharyngeal reflux associated with eustachian tube muscle weakness and a supine position. In addition, ALS patients under mechanical ventilation have a limited ability to convey their pain. Their complaints are often overlooked because many physicians do not know that pain is common in ALS. Physicians should recognize brain abscess as a severe complication of ALS and listen to the complaints of these patients.

Key words: amyotrophic lateral sclerosis, brain abscess, chronic suppurative otitis media, intractable headache, eustachian tube, communication

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Introduction

Brain abscess is a life-threatening infection due to the intracerebral collection of pus. The most frequent clinical manifestation is headache, followed by focal neurological deficits, a fever and nausea (1).

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that causes the degeneration of the upper and lower motor neurons. The clinical presentation and complications of ALS include limb weakness, dysphagia, speech disturbance, weight loss, pain, muscle wasting, dyspnea, and pneumonia (2). Impaired communication due to severe speech disturbance can reduce the quality of life for patients with ALS and lead to use of communication devices (3). However, even using these devices, patients cannot always convey their feelings in the same way as orally.

We herein report a patient with ALS under mechanical ventilation who developed intractable right temporal pain

due to a brain abscess.

Case Report

A 57-year-old woman was admitted to our hospital with right temporal headache. Six years before this admission, she had first noticed muscle weakness in her left leg and been diagnosed with probable ALS using the revised El Escorial Criteria (4). Four years before this admission, percutaneous endoscopic gastrostomy and tracheotomy had been performed, and mechanical ventilation had been started. Her voluntary movements became highly restricted, except for her eyes, and she managed to communicate using eye blinks or a communication board.

Three years before the current admission, she started to have recurrent episodes of bilateral otalgia. She was diagnosed with otitis media, and oral antibiotics and analgesics were prescribed, but her pain did not completely resolve. Two months before this admission, she began to complain of right temporal pain. Since her pain persisted and became

¹Department of Neurology, Gifu University Graduate School of Medicine, Japan, ²Department of Neurosurgery, Gifu University Graduate School of Medicine, Japan and ³Department of Otolaryngology, Gifu University Graduate School of Medicine, Japan

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Correspondence to Dr. Takayoshi Shimohata, shimohata@gmail.com

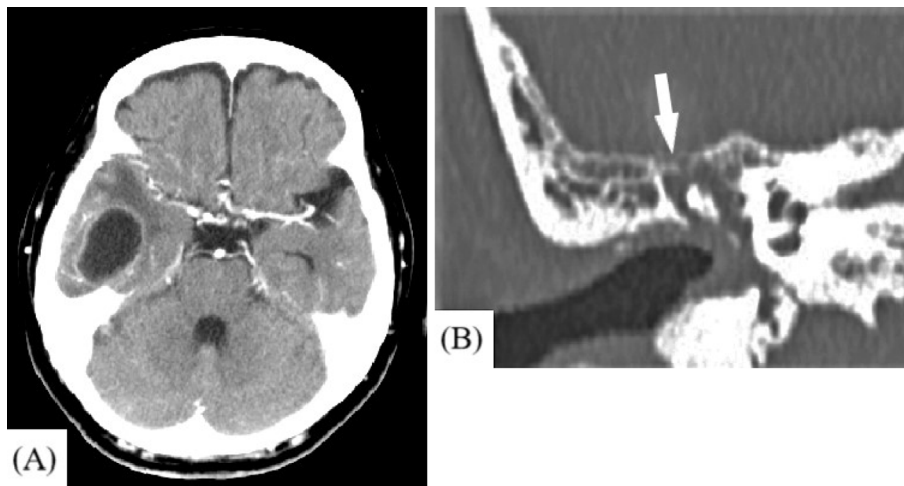


Figure. (A) Contrast-enhanced computed tomography (CT) of the brain on admission. (B) CT of the middle ear on admission (coronal section). Soft-tissue density and thinning of the tegmen tympani (arrow) in the right middle ear cavity.

intolerable, she was brought to our hospital in an ambulance.

On admission, she was alert and afebrile. Her right tympanic membrane was erythematic and accompanied by otorrhea. A blood test revealed an elevated inflammatory reaction (white blood cell level, 22,560 mg/dL, and C-reactive protein level, 38.51 mg/dL). Contrast-enhanced computed tomography (CT) of the brain revealed a hypodense area of 3 cm in diameter with ring enhancement surrounded by edema in the right temporal lobe (Figure A). CT of the middle ear showed soft-tissue density and thinning of the tegmen tympani in the right middle ear cavity (Figure B). Based on these findings, she was diagnosed with brain abscess secondary to suppurative otitis media.

She was treated with intravenous meropenem (3 g/day divided into 2 equal doses). Twelve days after admission, brain abscess drainage and tympanoplasty on the right ear were performed simultaneously. Blood culture, brain abscess culture, and ear drainage culture were negative. After eight weeks of intravenous antibiotics, her temporal headache improved. Contrast-enhanced CT of the brain at 81 days after admission showed a significant reduction in abscess size. Finally, the patient was discharged from the hospital 89 days after her admission.

Discussion

To our knowledge, this is the first case report of a patient with ALS developing a brain abscess that spread from chronic suppurative otitis media. In the previous report, the major cause of death in ALS patients with tracheostomy-positive pressure ventilation was respiratory failure from pneumonia/bronchopneumonia and sepsis associated with stones in the urinary or biliary tract (5). Brain abscess should also be recognized as a fatal complication in the patients. Two factors associated with ALS were considered to affect brain abscess development.

The first was eustachian tube dysfunction, which is attributed to muscle weakness and a long-term supine position due to ALS. In general, tensor veli palatini and levator tensor palatini open the eustachian tube to ventilate the middle ear (6). These muscles also help the eustachian tube drain effusion from the middle ear by contraction and relaxation (muscular pumping) (6, 7). The eustachian tube itself protects against pathogens with its narrow and long lumen (6). As muscle weakness progresses, however, the middle ear pressure becomes highly negative because gas continues to be absorbed by the mucous membrane of the middle ear (6). Occasional ventilation at a highly negative pressure combined with an impaired pump function promote aspiration of nasopharyngeal secretions into the middle ear and ultimately cause acute otitis media (8). Furthermore, patients with ALS under mechanical ventilation are in a supine position almost all day, which also enhances the reflux of saliva or nasal discharge (6). A brain abscess is formed by hematogenous dissemination from a systemic source of infection (e.g. endocarditis, bacteremia, etc.) as well as direct spread from contiguous infection sites (e.g. otitis media, sinusitis) (1). Therefore, if ALS patients under mechanical ventilation develop sepsis due to infection, bacteria can spread to the brain hematogenously and cause brain abscess.

Another factor that must be considered is the delayed diagnosis due to impaired communication between patients with ALS and physicians. It is difficult for physicians to understand the severity of pain in patients with ALS under mechanical ventilation. Furthermore, pain in patients with ALS is often overlooked because physicians misunderstand ALS as a purely motor disease and have little interest in pain (9). However, the reported frequency of pain in patients with ALS ranges from <15% up to 85% (9), indicating that pain is a common symptom in ALS. Pain in the extremities, back, shoulders and neck are frequently observed in patients with ALS. The pain is generally nociceptive because of the reduced mobility and skin pressure (10). In the present case,

the patient had complained of pain around her neck. This might have masked the right temporal pain and delayed the diagnosis of the brain abscess. A similar diagnostic delay can occur in other conditions, including urolithiasis and cholelithiasis, which can lead to fatal infection. The most effective way of diagnosing these diseases in ALS patients under mechanical ventilation is to spend time listening to their complaints and understand the characteristics of their pain.

In conclusion, brain abscess secondary to chronic suppurative otitis media should be recognized as a complication of ALS under mechanical ventilation. Brain abscess should be suspected when ALS patients under mechanical ventilation suffering from chronic otitis media present with a fever or prolonged headache. Because impaired communication related to ALS might delay the diagnosis of a critical complication, physicians must pay careful attention to complaints and conditions of patients with ALS.

The authors state that they have no Conflict of Interest (COI).

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