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Sequential Bilateral Upper Extremity Lymphedema in Advanced ALS with Hypoxic Brain Injury

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Introduction

Lymphedema is a chronic, progressive condition caused by impaired lymphatic drainage leading to protein-rich interstitial fluid accumulation and subsequent tissue fibrosis. Secondary lymphedema most commonly develops after malignancy-related lymph node dissection or radiation therapy. In contrast, non-malignant causes of upper extremity lymphedema are uncommon, and bilateral involvement is particularly rare. Amyotrophic lateral sclerosis (ALS) is characterized by progressive muscular weakness, resulting in severe restriction of movement. Prolonged immobility may compromise the skeletal muscle pump mechanism, which plays an essential role in both venous and lymphatic return. However, clinically significant lymphedema has rarely been reported in patients with advanced ALS, especially in the absence of malignancy. We report a case of sequential bilateral upper extremity lymphedema in a patient with advanced ALS complicated by hypoxic brain injury and prolonged vegetative status.

Case Report

A male patient born in November 1946 developed progressive limb weakness in May 2016 and was diagnosed with ALS in September 2016. Noninvasive ventilation was initiated in November 2016 due to ventilatory failure. With progression of bulbar dysfunction, tracheostomy and transition to tracheostomy invasive ventilation were performed in May 2017. In December 2020, he was admitted with altered mental status and diagnosed with hypoxic brain injury. Thereafter, he remained in a vegetative state with complete immobility under 24-hour invasive ventilation. In 2023, swelling of the left upper extremity developed. There was no edema in other body regions. Lymphoscintigraphy demonstrated impaired lymphatic drainage in the left upper extremity (Fig. 1-A), while CT scans excluded venous obstruction and malignancy. Complete decongestive therapy, including manual lymphatic drainage and compression garment application, resulted in clinical improvement. In December 2025, similar swelling developed in the right upper extremity. On physical examination, a positive Stemmer sign and a peau d'orange appearance were observed (Fig. 2). Lymphoscintigraphy again confirmed lymphatic dysfunction on right upper extremity (Fig. 1-B), and symptoms improved with conservative treatment.

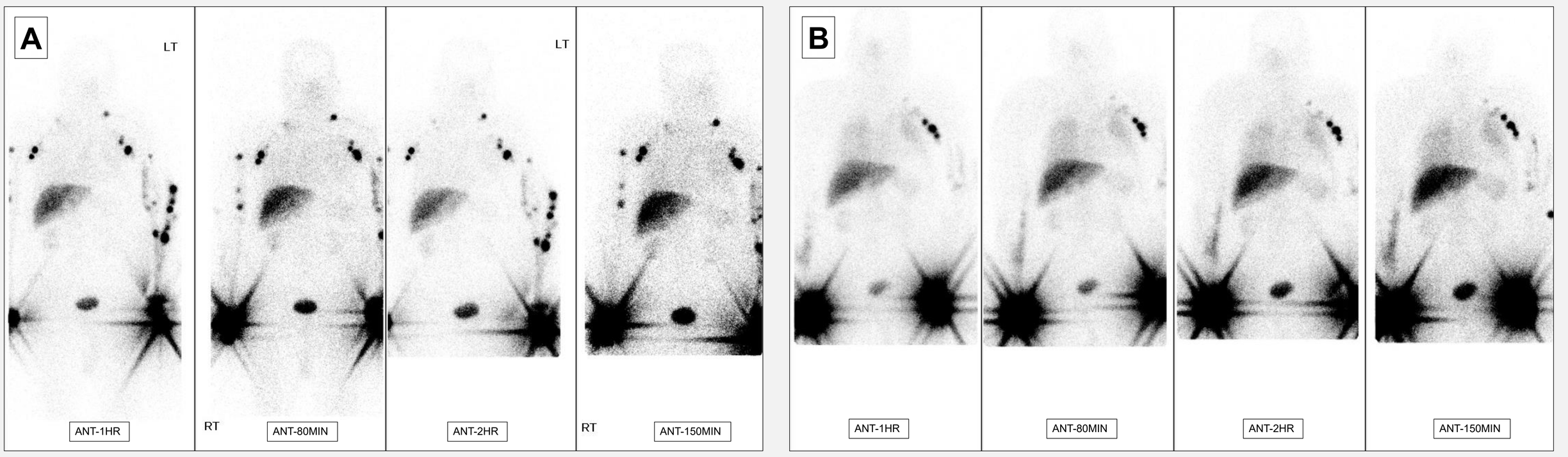

Fig. 1. Lymphoscintigraphies of the patient. In the image obtained in June 2023, lymphatic vessel uptake is increased in the left arm compared to the right. Multiple lymph nodes are observed around the elbow region. Dermal backflow is noted along the medial aspect of the left upper arm (A). In the study performed in December 2025, the main lymphatic vessel is not well visualized in the right upper extremity, and dermal backflow is observed in the forearm (B).

Fig. 2. Lymphedema of right upper extremity. Diffuse edema is observed in the right arm (A), with a peau d'orange appearance noted (B).

Conclusion

This case highlights that severe immobility and possible autonomic dysregulation in advanced neurologic disease may predispose patients to secondary lymphedema, even in the absence of malignancy or lymphatic surgery. Sequential bilateral involvement suggests progressive lymphatic insufficiency rather than simple dependent edema. Clinicians should consider lymphedema in chronically immobilized patients presenting with persistent limb swelling to ensure appropriate diagnosis and management.