EJR European Journal of Rheumatology

DOI:10.5152/eurjrheum.2025.24104

Case Report

Unveiling the Uncommon: Systemic Lupus Erythematosus with Dual Peculiarities—Vocal Fold Paralysis and a Winged Scapula

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Abstract

This case report highlights 2 uncommon presentations of systemic lupus erythematosus (SLE): winged scapula and unilateral vocal fold paralysis. A 44-year-old Emirati gentleman diagnosed with SLE presented with a 3-month history of gradual loss of voice and weakness in the right upper extremity. Clinical examination unveiled incomplete glottic closure due to right vocal fold paralysis. Additionally, there was restricted movement in the right arm and noticeable winging of the right scapula. Subsequent investigations indicated that the involvement of the recurrent laryngeal nerve and long thoracic nerve was attributed to SLE. The treatment plan included administering intravenous immunoglobulin at a dosage of 2 mg/kg over 3 days, followed by 2 doses of rituximab at 1 g each, spaced 2 weeks apart. These interventions were complemented by the patient's baseline SLE treatment regimen, including mycophenolate mofetil at 1 g daily and hydroxychloroquine at 200 mg daily. Following 3 months of initiating immunosuppressive therapy, substantial compensation was observed in the right vocal fold paralysis, demonstrating full glottic closure and only minor limitations in full abduction at the right shoulder. This case underscores the rarity of peripheral nervous system involvement in SLE, revealing a significant clinical challenge in diagnosing and managing affected patients. It underscores the imperative for more precise diagnostic criteria to effectively guide clinical decisions, particularly in light of the complexity of these presentations.

Keywords: SLE, vocal cord paralysis, winged scapula

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Cite this article as: Namas R, Al Qassimi S, Jamal Teir H, et al. Unveiling the uncommon: systemic lupus erythematosus with dual peculiarities—vocal fold paralysis and a winged scapula. *Eur J Rheumatol.* 2025, 12(3), 0104, doi: 10.5152/eurjrheum.2025.24104.

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Received: October 22, 2024
Revision Requested: November 9, 2024
Last Revision Received: November 26, 2024
Accepted: December 8, 2024
Publication Date: October 30, 2025

Introduction

Systemic lupus erythematosus (SLE) is a multifaceted autoimmune disease that is primarily caused by the dysregulation of the immune system, leading to the development of autoantibodies and immune complex deposition.[1,2] The 1999 diagnostic criteria published by the American College of Rheumatology classified neuropsychiatric systemic lupus erythematosus (NPSLE) into 19 neuropsychiatric syndromes, with 12 of these affecting the central nervous system (CNS) and 7 of these affecting the peripheral nervous system (PNS). According to a meta-analysis conducted by Unterman et al,[3] neuropsychiatric symptoms are prevalent in more than 50% of patients with SLE. Moreover, data from the Arab Gulf region shows variability in the frequency of NPSLE, ranging from 15.6% to 19%, with Dubai, United Arab Emirates, specifically reporting a frequency of 15.6%.[4,5] Studies indicate that CNS manifestations are common in SLE. The most frequently observed symptoms in NPSLE include depression, headaches, and cognitive dysfunction.[6-8] In contrast, PNS manifestations are notably infrequent. A recent international cohort study revealed a precise prevalence of 7.6% for PNS manifestations among individuals with SLE.[9] The most frequently occurring PNS syndromes include peripheral neuropathy, mononeuropathy, cranial neuropathy, and Guillain-Barré syndrome.[9] Here, the authors present a case of a patient with a longstanding history of SLE who developed unilateral vocal fold paralysis and winging of the scapula, both symptoms associated with the PNS.

Case Presentation

A 44-year-old Emirati man was diagnosed with SLE in 2008 following a prolonged episode of recurrent fever lasting over a month, accompanied by lymphadenopathy for which he underwent a lymph node biopsy that confirmed necrotizing lymphadenitis. Initial diagnostic laboratory investigations showed a positive IF-ANA with a speckled pattern and a titer of 1:1280, anti-dsDNA, anti-Smith, anti-SSA, and RF with low C3 and C4. Since his diagnosis, he was found to have renal involvement after the development

of proteinuria with a 24-hour urine protein of 2113.2 mg/24 hours, hematuria, and acute kidney injury with a creatinine of 1.4 mg/dL. He declined undergoing a renal biopsy at the time. His disease was subsequently well controlled on mycophenolate mofetil, hydroxychloroquine, and prednisolone. A few years after his diagnosis, he presented to an external hospital seeking medical attention due to a gradual onset of aphonia, dysphagia to solids and liquids, and weakness in his right upper extremity over the past 3 months. Concurrently, he appeared to have a significant flare of his SLE in the form of recurrent malar rash, oral ulcers, arthritis, proteinuria, elevated anti-dsDNA, and low complement levels with elevated inflammatory markers. He was hospitalized at an external facility for 3 weeks, during which he received intravenous (IV) corticosteroids. During his hospitalization, he underwent an otorhinolaryngology (ENT) evaluation, including laryngoscopy, which detected a polyp on the right vocal cord. Initially scheduled for biopsy, a subsequent laryngoscopy showed marked improvement in the polyp, likely due to corticosteroid treatment. Consequently, it was deemed that a biopsy was no longer strongly indicated. Moreover, there was a modest improvement in the range of motion of his right upper extremity, although he still faced challenges when attempting to raise it upwards. After discharge, he was advised to follow up with ENT and neurology as an outpatient, and his oral glucocorticoid dosage was gradually reduced. He is a known case of diabetes mellitus type II, hypertension, and hyperuricemia. He has a history of smoking for 15 years but has not traveled recently, experienced recent illness or Coronavirus disease 2019 (COVID-19) infection, and has not received the COVID-19 vaccination.

Following discharge, he sought a second opinion. The musculoskeletal examination revealed a limited range of motion at the right shoulder, up to 90°, along with winging of the right scapula and muscle loss in the posterior deltoid fibers, supraspinatus, and serratus anterior muscle groups (Figure 1). The left shoulder demonstrated a full range of motion, while both elbows showed a flexion contracture of approximately 10°, more prominent on the left. The wrists displayed unrestricted motion without evidence of joint effusion or tenderness along the joint lines. Laryngeal stroboscopy showed incomplete glottic closure due to right vocal fold paralysis in a paramedian position.

After a month of receiving hyaluronic acid injection laryngoplasty, his voice and dysphagia with solids showed noticeable improvement. However, he persisted with right vocal fold paralysis, occasional dysphagia with liquids, and weakness in his right upper extremity. As a result, referrals were initiated for evaluations by neurology and gastroenterology. Upper endoscopy identified deformity of the corniculate cartilage, while a modified barium swallow study revealed deficits in the pharyngeal stage, including pharyngeal residue and a wet voice after swallowing.

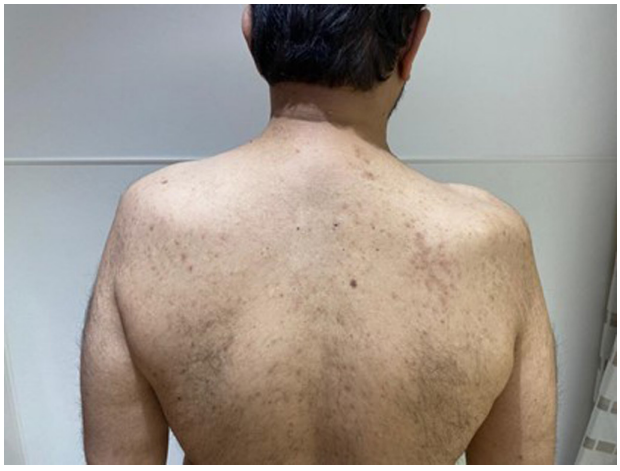

Figure 1. Winging of the right scapula and muscle loss in the posterior deltoid fibers, supraspinatus, and serratus anterior muscle groups.

From a neurological perspective, he underwent a nerve conduction study and electromyogram, which detected active and chronic neurogenic changes in muscles innervated by the right suprascapular, dorsal scapular, and long thoracic nerves. These findings suggested possible C5/C6 nerve root disease, as well as patchy upper trunk brachial plexopathy or plexitis. Magnetic resonance imaging of the brachial plexus did not indicate active brachial plexitis. Biopsy was not feasible due to the location of the nerves involved. Based on his clinical presentation, examination, and imaging results, the diagnosis of recurrent laryngeal nerve and long thoracic nerve involvement in the context of SLE was favored. Considering his clinical improvement, it was decided that a combination of physical therapy and management of SLE would be the initial therapeutic approach.

Due to the rare occurrence of SLE with involvement of the vocal folds and upper trunk of the brachial plexus, he initiated treatment with intravenous immunoglobulin (IVIG) at a dose of 2 g/kg over 3 days, followed by 2 doses of rituximab at 1 g each. These treatments were administered alongside his ongoing SLE regimen, which included mycophenolate mofetil at 1 g daily and hydroxychloroquine at 200 mg daily. He tolerated the therapy well, and subsequently, the rituximab doses were scheduled every 6 months. During the follow-up appointment 3 months later, significant improvement was noted in his right vocal fold paralysis, with complete glottic closure and only mild restriction observed in full abduction of the right shoulder (Figure 2). Written informed consent was obtained from the patient directly.

Discussion

Due to the rarity of PNS manifestations in SLE, it is essential to increase awareness of potential symptoms to enable early diagnosis and treatment. This was exemplified in the case of our patient, who presented with aphonia and dysphagia when consuming solid foods, along with weakness in his right arm, rendering him unable to raise it. Further diagnostic evaluations, including laryngoscopy, confirmed paralysis of the right vocal cord. Additionally, electromyography revealed patchy upper trunk brachial plexopathy and C5/C6 nerve root disease, leading to active and chronic neurogenic changes in the muscles supplied by the right suprascapular, dorsal scapular, and long thoracic nerves. The collective presentation of signs and symptoms described prompted significant concern regarding the possibility of mononeuritis multiplex affecting the brachial plexus. Mononeuritis multiplex typically manifests as asymmetrical sensory and motor deficits in at least 2 peripheral nerve areas.[10] The pathogenesis is believed to involve immune-mediated vasculitic damage to the vasa nervorum, leading to Wallerian degeneration.[11] In this case, the suspected mononeuritis impacted the brachial plexus, leading to the development of winged scapula. Similarly, cranial neuropathy involving the vagus nerve contributed to his dysphagia due to right vocal cord paralysis. Cranial neuropathy involving

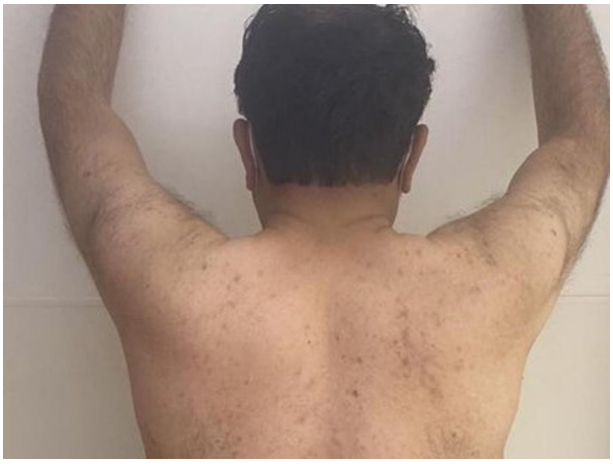

Figure 2. Complete resolution of previously noted winging of the right scapula.

the vagus nerve is rare compared to more frequent involvement of the vestibulocochlear nerve, oculomotor set (oculomotor, trochlear, and abducens nerves), trigeminal nerve, and facial nerve.[12] However, due to the challenges in obtaining a biopsy from the nerve's location, a definitive diagnosis could not be established. There is a dearth of studies exploring the relationship between SLE-related PNS with disease activity and laboratory markers, and this is likely due to small sample size and heterogeneity of PNS manifestations in SLE.[13] However, associations have been reported with disease activity and CNS contribution. Florica et al reported higher disease activity (i.e., median SLEDAI of 11) and a higher frequency of mononeuritis multiplex in their cohort of patients with SLE-associated neuropathies.[14]

Based on the probable diagnosis of mononeuritis multiplex, prompt initiation of symptomatic treatment was implemented for the patient. As per standard practice, the primary approach involves the early administration of high-dose corticosteroids and immunosuppressants,[1,15] both of which were prescribed for the patient. Corticosteroids effectively relieved his dysphagia and hoarseness of voice. Furthermore, the patient received IVIG and rituximab, which improved weakness in his right arm and enhanced his range of motion. Immediate initiation of physiotherapy was recommended to aid in restoring strength in his trapezius and serratus anterior muscles. Literature highlights the pivotal role of physiotherapy in management, especially since vasculitic neuropathy and demyelination show favorable responses compared to axonal loss.[15] His clinical improvement is likely a result of intense immunosuppressive therapies, hyaluronic injections, and physical therapy. Given the chronic nature of SLE, the formation of a multidisciplinary team dedicated to patient monitoring, education, and treatment optimization is crucial. This collaborative approach is instrumental in reducing morbidity and mortality rates and enhancing the patient's quality of life.

In summary, we describe a case of SLE presenting with rare peripheral nervous system manifestations, including vocal cord paralysis due to recurrent laryngeal nerve palsy and winging of the scapula from C5/C6 root involvement. The patient's symptoms markedly improved with treatment involving immunosuppressants and early initiation of physiotherapy. Due to the infrequency of PNS syndromes in SLE, it is crucial for healthcare providers to maintain heightened awareness for timely recognition and appropriate management.

Data Availability Statement: Data is available to be reviewed for any audits.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – R.N., S.AQ., F.E., R.N.; Design – R.N., S.AQ., F.E., R.N.; Supervision – R.N., S.AQ., F.E., R.N.; Resources – R.N., S.AQ., F.E., R.N.; Materials – R.N., S.AQ., F.E., R.N.; Data Collection and/or Processing – R.N., S.AQ., F.E., R.N., H.J.T., A.H., P.K.; Analysis and/or Interpretation – R.N., S.AQ., F.E., R.N., H.J.T., A.H., P.K.; Literature Search – R.N., S.AQ., F.E., R.N., H.J.T., A.H., P.K.; Writing – R.N., S.AQ., F.E., R.N., H.J.T., A.H., P.K.; Critical Review – R.N., S.AQ., F.E., R.N., H.J.T., A.H., P.K.

Acknowledgements: The authors would like to acknowledge our patient's trust and confidence in us participating in their care.

Declaration of Interests: The authors have no conflict of interest to declare.

Funding: The authors declared that this study has received no financial support.

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