Content available at: https://www.ipinnovative.com/open-access-journals

IP Indian Journal of Immunology and Respiratory Medicine

Journal homepage: https://www.ijirm.org/

Case Report

Primary sjögren's syndrome in an adolescent: A rare presentation with ascending motor neuropathy mimicking CIDP

Mohd Aateer Shaikh[1], Gyanshankar P Mishra[1]*, Sanjay M Gour[1], Radha Munje[1]

[1]Dept. of Respiratory Medicine, Indira Gandhi Government Medical College, Nagpur, Maharashtra, India

Abstract

We report the case of an 18-year-old female presenting with a 5-month history of fever, progressive limb weakness, and generalized swelling. Initially diagnosed with Guillain-Barré syndrome (GBS), the patient's clinical course evolved into chronic inflammatory demyelinating polyneuropathy (CIDP). Further investigation, including serological testing and a lip biopsy, revealed positive autoimmune markers (anti-SSA (Ro) and anti-SSB (La)) and confirmed the diagnosis of Sjögren's syndrome-associated autoimmune neuropathy. The patient was managed with intravenous steroids, antibiotics, diuretics, and physiotherapy, resulting in substantial clinical improvement. This case underscores the importance of a thorough diagnostic approach and individualized treatment strategies in managing autoimmune-related neuropathies.

Keywords: Case report, Sjögren's syndrome, Pediatric Sjögren's syndrome, Guillain-Barré syndrome, Chronic inflammatory demyelinating polyneuropathy, Autoimmune neuropathy, Plasmapheresis, Autoimmune diseases.

Received: 09-02-2025; **Accepted:** 31-03-2025; **Available Online**: 29-04-2025

1. Introduction

Sjögren's syndrome is a chronic autoimmune disorder marked by lacrimal and salivary gland dysfunction, causing dry eyes (xerophthalmia) and mouth (xerostomia). While typically diagnosed in adults over 40, pediatric cases are and often lack classic sicca symptoms, making early diagnosis challenging. Beyond glandular involvement, the disease may present with extra-glandular features—especially neurological manifestations such as peripheral sensory neuropathy—which can precede sicca symptoms occur in up to 70% of cases.[1] However, motor impairments are underreported, particularly in pediatric patients.[2] The 2016 ACR-EULAR Classification Criteria provide objective guidelines for the diagnosis of primary Sjögren's syndrome, aiding recognition even in atypical presentations (

Table 1).[3] This case describes an uncommon presentation of primary Sjögren's syndrome in an adolescent with isolated limb weakness, emphasizing the disease's broad and diverse clinical spectrum. To our knowledge, this is the first reported case from India of an adolescent presenting with isolated limb weakness as the primary neurological manifestation of Sjögren's syndrome.

2. Case Presentation

2.1. History

An 18-year-old female was admitted to our hospital on 01/02/2024 with a 5-month history of low-grade fever, which escalated to high-grade fever over the past 15 days. The fever was intermittent and responsive to antipyretics. In addition, the patient reported progressive breathlessness for the past 15 days (MMRC grade 4), accompanied by orthopnea, though without paroxysmal nocturnal dyspnea. She also presented with progressive limb weakness, initially affecting the lower limbs and gradually involving all four limbs, consistent with ascending paralysis. Furthermore, the patient had a facial rash (**Figure 1**) and generalized body swelling for one month, which was pitting in nature and improved with limb

*Corresponding author: Gyanshankar P Mishra
Email: gpmishra81@gmail.com

http://doi.org/10.18231/j.ijirm.2025.006

elevation. Over the last 15 days, she also experienced non-bilious vomiting, containing food particles, typically postprandial, without diarrhea or hematemesis.

The patient had a history of hospitalization three months prior for similar symptoms. Previously, the patient had been hospitalized for fever, vomiting, and limb weakness, initially diagnosed as Guillain-Barré syndrome (GBS). GBS was confirmed through MRI and nerve conduction studies, which demonstrated motor-predominant demyelinating polyradiculoneuropathy. She was treated with intravenous steroids and plasmapheresis, achieving partial clinical improvement.

2.2. Examination

On admission, the patient's vital signs were notable for a pulse rate of 130 beats per minute, blood pressure of 80/54 mmHg, respiratory rate of 40 breaths per minute, and oxygen saturation of 95% on room air. Chest examination revealed bilateral coarse inspiratory crepitations. Neurological examination showed reduced muscle power (2/2/1/1 in the right upper limb, left upper limb, right lower limb, and left lower limb), absent deep tendon reflexes, and reactive pupils to light.

2.3. Investigations

Labs revealed cerebrospinal fluid (CSF) analysis with albuminocytologic dissociation (elevated protein: 198.9 mg/dL; leukocyte count: 40 cells/µL). Serological tests were positive for rheumatoid factor (70.3 IU/mL), antinuclear antibodies (ANA) with a 3+ titer by immunofluorescence assay, anti-SSA (Ro) (3+), anti-SSB (La) (3+), and anti-Ro52 (3+), indicating an autoimmune etiology. Chest X-ray (**Figure 2**) demonstrated bilateral middle and lower zone haziness, cardiomegaly, and pleural effusion. Previous MRI (14/11/2023) revealed abnormal enhancement of the lumbar nerve roots without any evidence of spinal cord involvement or bony abnormalities, findings consistent with a diagnosis of Guillain-Barré syndrome (GBS). A CECT thorax (04/02/2024) (**Figure 3**) showed bilateral lower lobe consolidation, ground-glass opacities, and cardiomegaly, suggesting cardiogenic pulmonary edema. Additional findings included hypoproteinemia, metabolic acidosis, pleural effusion, and ascites (transudative).

2.4. Management

The patient was diagnosed with Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) secondary to primary Sjögren's syndrome, based on the 2016 ACR-EULAR Classification Criteria for primary Sjögren's syndrome (

Table 1). (4) The presence of autoimmune markers supported this diagnosis; a lip biopsy was done to meet the diagnostic criteria according to the 2016 ACR-EULAR (**Figure 4**), revealing focal lymphocytic infiltration and the clinical presentation of ascending paralysis with chronic features.[4]

Table 1: Application of 2016 ACR-EULAR Classification criteria for primary sjögren's syndrome in the present case (3)

2016 ACR-EULAR Classification Criteria for Primary Sjögren's Syndrome*		
Item	**Weight / Score**	**Score in our patient**
Labial salivary gland with focal lymphocytic sialadenitis and focus score ≥ 1.3	3	3
Anti-SSA (Ro) +	3	3
Ocular staining score ≥ 5 (or van Bijsterfeld score ≥ 4) on at least one eye	1	0
Schirmer ≤ 5 mm/5min on at least one eye	1	0
Unstimulated whole saliva flow rate ≤ 0.1 ml/min	1	0
Total score	9	6

***Note:** A total score of ≥4 supports the classification of primary Sjögren's syndrome in the absence of exclusion criteria. Inclusion criteria include ocular and/or oral dryness, or clinical suspicion of Sjögren's syndrome based on the EULAR Sjögren's Syndrome Disease Activity Index (ESSDAI). Exclusion criteria include a history of head and neck radiation, active hepatitis C virus (HCV) infection, HIV/AIDS, sarcoidosis, amyloidosis, graft-versus-host disease, or IgG4-related disease.

The patient was initiated on intravenous dexamethasone (a potent immunosuppressive agent), antibiotics to address potential infections, diuretics for fluid management (due to pleural effusion and ascites), and physiotherapy to manage the progressive weakness. Close monitoring was maintained for any respiratory or cardiovascular deterioration.

The patient showed significant clinical improvement after treatment, resolving fever, weakness, and respiratory symptoms. She was gradually weaned off intravenous steroids and discharged on tapering oral steroids (Prednisolone) with continued physiotherapy for rehabilitation. The patient was stable at discharge with substantial motor strength and improvement in respiratory function. A summary of the patient's clinical timeline, key investigations, and interventions is illustrated in **Figure 5**.

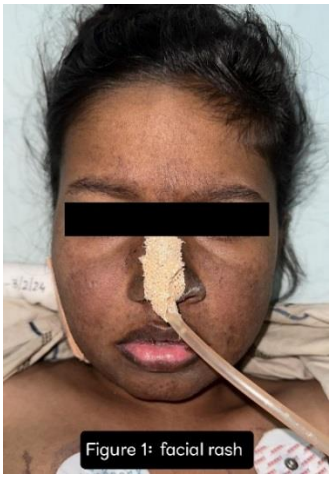

Figure 1: Facial rash observed on presentation, characterized by erythematous lesions over the cheeks, noted in association with systemic symptoms and autoimmune activity.

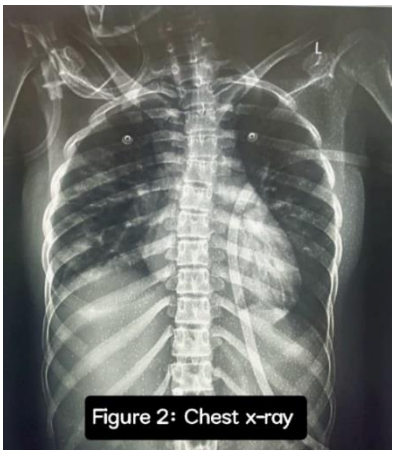

Figure 2: Chest X-ray displaying perihilar shadows accompanied by cardiomegaly.

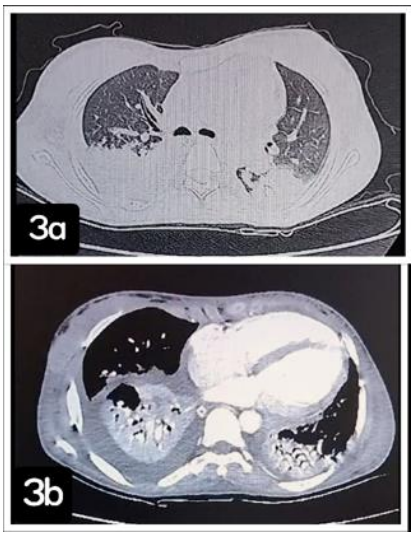

Figure 3: a: and **b:** CT thorax indicative of bilateral pleural effusion, consolidation, GGOs, and cardiomegaly suggestive of pulmonary edema.

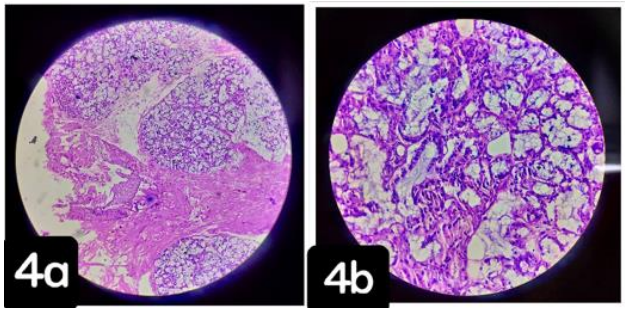

Figure 4: **a,b:** Histology of Lip biopsy showing focal lymphocytic infiltrations with salivary gland tissue

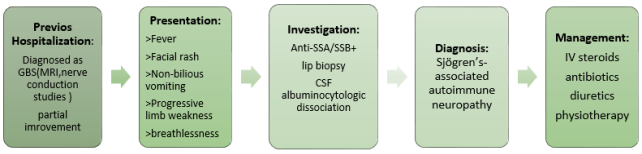

Figure 5: Case summary of patient presentation, diagnosis and management

3. Discussion

Sjögren's syndrome, while primarily affecting adults, has been increasingly recognized in pediatric populations, albeit with atypical features.[5] Classic sicca symptoms are often absent in children, contributing to delayed or missed diagnoses. In such cases, extra-glandular manifestations—including neurological symptoms—may serve as the first clinical clue.[6] Our patient presented with progressive ascending motor weakness, initially resembling GBS and later evolving into a CIDP-like picture. This underscores the need to consider autoimmune etiologies, including Sjögren's syndrome, in the differential diagnosis of pediatric neuropathies that deviate from classical post-infectious GBS trajectories.

Though the 2016 ACR-EULAR Classification Criteria were developed for adults, they provide a structured framework applicable in selected pediatric cases. Our patient fulfilled 6 points—3 each from anti-SSA (Ro) positivity and lip biopsy findings—meeting the ≥4 threshold required for classification. While ophthalmologic and salivary testing was non-contributory, the absence of exclusion criteria and the presence of extra-glandular neurologic manifestations support the diagnosis.[4]

This case underscores the atypical nature of pediatric-onset Sjögren's syndrome and illustrates that GBS-like or CIDP-like neuropathies may serve as the initial presentation of systemic autoimmune diseases such as primary Sjögren's syndrome. Unlike adults, children with SS often do not fulfill the full 2016 classification criteria and may lack classic sicca symptoms. Younger children typically present with recurrent or persistent parotitis, while older children, as in our case, may manifest systemic or neurological features. The

pathogenesis of such neuropathies is presumed to involve immune-mediated peripheral nerve injury. While sensory-predominant neuropathies have been described more frequently in Sjögren's syndrome, isolated motor involvement—especially in pediatric patients—is rare. These age-specific variations and atypical neurological presentations highlight the need for heightened clinical suspicion and careful, context-driven application of adult diagnostic criteria in children.[7]

Differential diagnoses including systemic lupus erythematosus (SLE), mixed connective tissue disease (MCTD), and vasculitis were considered. ANA positivity raised suspicion, but the absence of anti-dsDNA, anti-Sm, or U1-RNP antibodies, normal complements, and lack of systemic features characteristic of SLE or MCTD made these less likely. There were no clinical or laboratory signs of systemic vasculitis.

Glucocorticoids are the most established treatment for autoimmune neuropathies linked to Sjögren's syndrome. Our patient responded well to parenteral dexamethasone followed by tapering oral prednisolone, along with supportive therapy, achieving marked improvement in motor function.[8,9] In addition to glucocorticoids, various immunomodulatory agents—such as azathioprine, intravenous immunoglobulins (IVIG), mycophenolate mofetil, rituximab, and even intensified therapies like plasmapheresis and cyclophosphamide—have shown favorable responses in patients with Sjögren's-associated motor neuropathies.[10]

Although long-term data in pediatric Sjögren's-associated motor neuropathy are scarce, early diagnosis and timely immunosuppressive therapy can significantly improve outcomes.[8,10] Continued follow-up is essential to monitor for recurrence or systemic progression, with multidisciplinary coordination between neurology, rheumatology, and pediatrics.

4. Conclusion

Early recognition of Sjögren's syndrome is essential, as neurological symptoms, including isolated limb weakness, can precede the classic sicca manifestations. Paediatric patients presenting with unexplained limb weakness, even in the absence of typical glandular symptoms, should be considered for evaluation of Sjögren's syndrome. A comprehensive autoimmune workup, including serological testing and biopsy, facilitates early diagnosis and intervention. Timely initiation of immunomodulatory treatment may prevent disease progression and improve neurological outcomes, underscoring the importance of a multidisciplinary approach in managing atypical autoimmune presentations. Delayed diagnosis can lead to irreversible neurological damage, highlighting the need for greater awareness of the broad clinical spectrum of pediatric Sjögren's syndrome. Further documentation of atypical pediatric presentations may help refine diagnostic criteria and improve early recognition of autoimmune neuropathies.

5. Declaration of Patient Consent

Patient's consent is not required as the patient's identity is not disclosed or compromised.

6. Source of Funding

None.

7. Conflicts of Interest

There are no conflicts of interest.

References

1. Aytekin E, Coşkun H, Pekin Doğan Y, Dede BT, Burnaz Ö, Emre U. Rheumatic Diseases Presenting with Guillain-Barré Syndrome: Sjögren's Syndrome and Systemic Lupus Erythematosus. *Istanbul Med J*. 2020;21(1):9–11.
2. Ketabforoush AHME, Khoshsirat NA, Maghoul A, Nirouei M, Dolatshahi E. Acute polyneuropathy as the main manifestation of primary Sjogren's syndrome: A case report. *Clin Case Rep*. 2022;10(5):e05828.
3. Shiboski CH, Shiboski SC, Seror R, Criswell LA, Labetoulle M, Lietman TM, et al. American College of Rheumatology/European League Against Rheumatism Classification Criteria for Primary Sjögren's Syndrome: A Consensus and Data-Driven Methodology Involving Three International Patient Cohorts. *Arthritis Rheumatol*. 2017;69(1):35–45.
4. Liao R, Yang HT, Li H, Liu LX, Li K, Li JJ, et al. Recent Advances of Salivary Gland Biopsy in Sjögren's Syndrome. *Front Med*. 2022;8:792593.
5. Baik SJ, Han TH, Jung SY, Bang JI, Chae KY. A Juvenile Case of Primary Sjögren Syndrome Presenting as Limb Weakness. *Ann Child Neurol*. 2022;30(1):38-41.
6. Tanaka K, Nakayasu H, Suto Y, Takahashi S, Konishi Y, Nishimura H, et al. Acute Motor-dominant Polyneuropathy as Guillain-Barré Syndrome and Multiple Mononeuropathies in a Patient with Sjögren's Syndrome. *Intern Med*. 2016;55(18):2717–22.
7. Basiaga ML, Stern SM, Mehta JJ, Edens C, Randell RL, Pomorska A, et al. Childhood Sjögren syndrome: features of an international cohort and application of the 2016 ACR/EULAR classification criteria. *Rheumatology (Oxford)*. 2020;60(7):3144.
8. Liampas A, Parperis K, Erotocritou MF, Nteveros A, Papadopoulou M, Moschovos C, et al. Primary Sjögren syndrome-related peripheral neuropathy: A systematic review and meta-analysis. *Eur J Neurol [Internet]*. 2022;30(1):255–65.
9. Priori R, Mastromanno L, Izzo R. What about glucocorticoids in primary Sjögren's syndrome?. *Clin Exp Rheumatol.* 2021;38(4):S237–44.
10. Seeliger T, Prenzler NK, Gingele S, Seeliger B, Körner S, Thiele T, et al. Neuro-Sjögren: Peripheral neuropathy with limb weakness in Sjögren's syndrome. *Front Immunol [Internet]*. 2019;10(7):446252.

Cite this article Shaikh MA, Mishra GP, Gour SM, Munje R. Primary sjögren's syndrome in an adolescent: A rare presentation with ascending motor neuropathy mimicking CIDP. *IP Indian J Immunol Respir Med*. 2025;10(1):33-36.