

Progressive brachial diplegia unmasking flail arm syndrome, a regional variant of amyotrophic lateral sclerosis: a case report

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ABSTRACT

Background: Flail arm syndrome (FAS) is an uncommon variant of amyotrophic lateral sclerosis (ALS) characterized by progressive, predominantly lower motor neuron weakness confined mainly to the upper limbs. Because structural cervical disease is common in older adults, its diagnosis may be delayed or misattributed to compressive or peripheral nerve disorders.

Case Presentation: A 69-year-old man developed progressive asymmetric upper limb weakness over 2 years, evolving into bilateral brachial diplegia with preserved lower limb function. Neurological examination showed marked upper limb atrophy, fasciculations, and extensor plantar responses, without sensory deficits. MRI showed no significant cervical compression, while electromyography revealed multisite denervation, supporting the diagnosis of FAS as a phenotype of ALS. Multiple alternative diagnoses, including cervical myelopathy, multifocal motor neuropathy, and myopathy, were systematically excluded.

Conclusion: This case highlights the importance of recognizing FAS within the ALS spectrum. Electrophysiological evidence of multisite motor neuron involvement is crucial to avoid misdiagnosis and unnecessary interventions.

Keywords: Amyotrophic lateral sclerosis, motor neuron disease, flail arm syndrome, electromyography, neurophysiology.

Type of Article: CASE REPORT **Specialty:** Neurology

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Background

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disorder affecting upper and lower motor neurons, typically presenting with limb or bulbar weakness that spreads across anatomical regions over time [1]. In addition to the classical phenotype, regional variants have been described, including flail arm syndrome (FAS) and flail leg syndrome, which are characterized by a more restricted anatomical distribution at onset and a slower rate of progression [2,3].

FAS, also known as brachial amyotrophic diplegia or “man-in-the-barrel” syndrome, is characterized by progressive, predominantly lower motor neuron weakness affecting the proximal upper limbs, often evolving into symmetric bilateral arm paralysis with relative early sparing of lower limb and bulbar function [2,4]. Although survival is generally longer than in classical ALS, progression to generalized disease is frequent [3]. Despite its distinct phenotype, diagnosis is frequently delayed due to overlap with common conditions such as cervical spondylosis and immune-mediated neuropathies.

We report a case illustrating the diagnostic challenges of FAS and discuss the key clinical and electrophysiological features supporting its recognition as an ALS variant.

Case Description

A 69-year-old right-handed man with no significant medical history, presented to our neurology department with a 24-month history of progressive upper limb weakness and muscle wasting. The symptoms began with proximal weakness of the left upper limb, which gradually extended distally over the following months and subsequently involved the right upper limb 12 months after onset. Over the following year, the patient developed severe bilateral functional impairment of the upper limbs, leading to referral for neurological evaluation. During the early phase of the disease, symptoms had been attributed to degenerative cervical spine disease, but no meaningful improvement was observed with symptomatic treatment. The patient reported mild dysphagia but denied choking, dysarthria, or dyspnea. There were no sensory symptoms, sphincter disturbances, or cognitive or behavioral changes. Family history was negative for neuromuscular disease.



Figure 1. Clinical photographs of the patient demonstrating severe bilateral upper limb muscle wasting. (A) Anterior view showing marked deltoid and pectoral muscle atrophy. (B) Posterior view demonstrating pronounced wasting of the trapezius, supraspinatus, and scapular girdle muscles. (C) Lateral view illustrating proximal upper limb muscle atrophy.

Neurological examination revealed severe bilateral upper limb weakness affecting proximal and distal muscle groups (MRC grades 1–2 proximally and 2–3 distally), with marked atrophy of the scapular girdle muscles (Figure 1) and severe wasting of the intrinsic hand muscles, particularly the thenar eminences (Figure 2). Fasciculations were visible in deltoid and biceps muscles bilaterally. Mild neck extensor weakness was present (MRC grade 4). Lower limb strength and gait were normal. Deep tendon reflexes were absent in all limbs, while plantar responses were extensor bilaterally. No tongue atrophy or fasciculations were observed. Sensory examination and cranial nerve function were otherwise normal. The clinical course of the disease is summarized in Figure 3.

Laboratory tests, including metabolic panels, inflammatory markers, and creatine kinase levels, were performed to evaluate for metabolic, inflammatory, or myopathic etiologies and were all within normal limits, arguing against primary muscle disease or systemic inflammatory conditions. Cerebrospinal fluid (CSF) analysis was normal, making inflammatory or infectious polyradiculopathies unlikely. Brain MRI was unremarkable, making a central structural cause unlikely, while cervical



Figure 2. Palmar view of both hands of the patient showing severe thenar and intrinsic hand muscle wasting, more pronounced in the abductor pollicis brevis muscles bilaterally.

spine MRI demonstrated multilevel spondylotic degenerative changes from C3 to C7 without significant spinal canal stenosis, nerve root or spinal cord compression, or intrinsic spinal cord signal abnormalities. A CT scan of the chest, abdomen, and pelvis, along with paraneoplastic

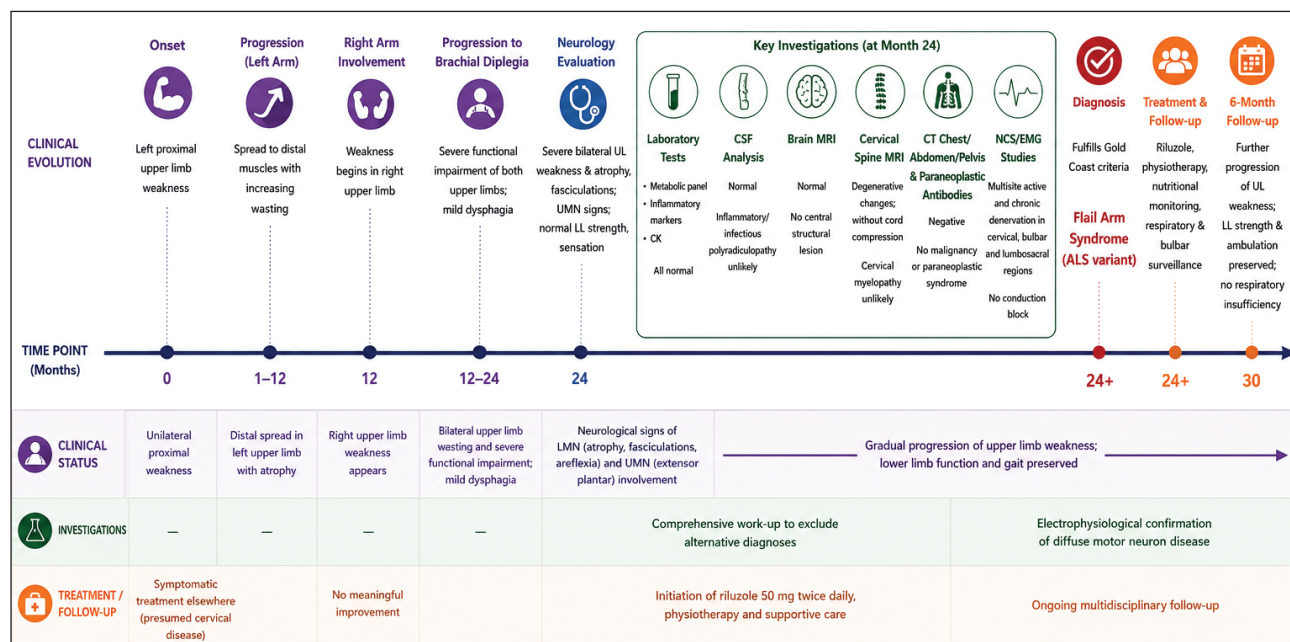


Figure 3. Clinical timeline of the patient.

Table 1. Motor nerve conduction studies demonstrating reduced CMAP amplitudes in the bilateral median and ulnar nerves, with preserved distal latencies, conduction velocities, and F-wave latencies. No definite conduction block was identified following proximal and distal stimulation across all tested segments.

Nerve/site	Latency (ms)	CMAP (mV)	MCV (m/s)	F-wave latency (ms)
Median L - Wrist/Elbow	3.27/7.75	3.5/3.1	49	31
Median R - Wrist/Elbow	3.23/8.31	4.1/3.8	59	32
Ulnar L - Wrist - Below elbow - Above elbow - Axilla	3.19	4.6 4.3 4.1 3.9	52	28
Ulnar R - Wrist - Below elbow - Above elbow - Axilla	3.08	5.2 5 4.8 4.7	50	30
Tibial R -Ankle/Popliteal fossa	3.54/11.88	8.9/6.8	48	47
Tibial L -Ankle/Popliteal fossa	2.81/11.41	9.1/6.9	49	49.5
Peroneal R -Ankle / Fibular head / Popliteal fossa	4.27/11.2/12.19	3.6/3.2/3.1	52	51
Peroneal L -Ankle / Fibular head / Popliteal fossa	3.69/10.42/11.35	4.1/3.7/3.6	54	49

CMAP, compound muscle action potential; L, left; MCV, motor conduction velocity; R, right.

antibody testing, was performed to evaluate for an underlying malignancy or paraneoplastic motor neuron syndrome, both were negative.

On nerve conduction studies, compound motor action potential (CMAP) amplitudes were reduced in the upper

limbs, while lower limb motor conduction studies were preserved. Sensory nerve conduction studies were normal, and no conduction block or demyelinating features were identified (Tables 1 and 2). Needle electromyography (EMG) examination demonstrated widespread neurogenic

Table 2. Sensory nerve conduction studies with preserved SNAP amplitudes and sensory conduction velocities in both upper and lower limbs.

Nerve	Distal latency (ms)	SNAP amplitude (μ V)	Conduction velocity (m/s)
Median (R)	2.5	30.6	53
Median (L)	2.9	25.1	51
Ulnar (R)	2.2	20.4	60
Ulnar (L)	1.98	18.9	56
Sural (R)	2.34	20.2	50
Sural (L)	2.50	19.5	49

L, left; SNAP, sensory nerve action potential; R, right.

Table 3. Needle EMG Examination of the patient revealing widespread active and chronic neurogenic changes, predominantly involving cervical myotomes, with additional abnormalities in bulbar, abdominal, and lumbosacral muscles.

Muscle	Spontaneous activity			Recruitment (MUPs)
	FIB	FASC	PSW	
Upper limbs				
First Dorsal Interosseous L	+1	+2	None	Reduced
First Dorsal Interosseous R	+1	+3	None	Reduced
Brachioradialis L	None	+1	+1	Intermediate
Brachioradialis R	None	+1	None	Intermediate
Extensor Digitorum Communis L	None	+1	+1	Intermediate
Deltoid R	+1	+3	+1	Reduced
Lower limbs				
Tibialis anterior R	None	None	None	Normal
Tibialis anterior L	None	+1	None	Intermediate
Gastrocnemius L	None	+1	None	Normal
Vastus lateralis R	+1	+1	None	Intermediate
Bulbar muscles				
Genioglossus R	+1	+2	None	Intermediate
Genioglossus L	None	+2	None	Intermediate
Mentalis R	None	+1	None	Normal
Orbicularis oculi L	None	None	None	Normal
Abdominal muscles				
Rectus abdominis	None	+1	None	Normal

Fib, fibrillation potentials; Fasc, fasciculation potentials; L, left; MUPs, motor unit potentials; PSW, positive sharp waves; R, right.

abnormalities with evidence of both active and chronic denervation, including fibrillation potentials and fasciculations, associated with reduced or intermediate recruitment in muscles innervated by cervical segments, as well as in selected bulbar and lumbosacral territories (Table 3). These findings were consistent with a diffuse motor neuron disease.

Several alternative diagnoses were systematically evaluated based on the available clinical, electrophysiological, laboratory, and imaging findings. Cervical spondylotic amyotrophy was considered unlikely based on the absence of significant spinal cord or nerve root compression on

MRI and by electrophysiological abnormalities extending beyond the cervical region. Multifocal motor neuropathy and chronic inflammatory demyelinating polyradiculoneuropathy were not supported because of the absence of conduction block or demyelinating features, normal CSF analysis, and the presence of upper motor neuron signs. Brachial plexopathy, Kennedy disease, and primary myopathies, including inclusion body myositis, were considered unlikely based on the clinical presentation, normal creatine kinase levels, neurogenic electromyographic findings, and lack of supportive clinical features. Likewise, a paraneoplastic motor neuron syndrome was considered

unlikely based on negative imaging and paraneoplastic antibody testing.

The combination of progressive bilateral upper limb weakness, with marked atrophy and fasciculations, preserved lower limb function, absence of sensory involvement, and electrophysiological evidence of multisite motor neuron degeneration supported the diagnosis of FAS, a regional variant of ALS. The diagnosis fulfilled the Gold Coast criteria by demonstrating progressive motor impairment following previously normal motor function, unequivocal lower motor neuron dysfunction established clinically (weakness, muscle atrophy, and fasciculations) and electrophysiologically (active and chronic denervation in multiple body regions), together with upper motor neuron dysfunction evidenced by bilateral extensor plantar responses, after appropriate evaluation of alternative diagnoses.

Supportive care was initiated, including physiotherapy and nutritional monitoring. The patient was started on riluzole 50 mg (twice daily), following discussion of potential benefits and limitations. The patient was followed for 6 months after diagnosis. During this period, upper limb weakness continued to progress, while lower limb strength and independent ambulation remained preserved, with no clinical spread to the lower extremities. Mild dysphagia remained stable, without the development of dysarthria or other significant bulbar dysfunction. Serial spirometry performed at diagnosis and at the 6-month follow-up remained stable, showing forced vital capacity at 78% of the predicted value, with no clinical or functional evidence of respiratory involvement.

Discussion

FAS is a rare but distinctive variant of ALS characterized by predominant involvement of the upper limbs, usually sparing the lower limbs and bulbar function [2]. Unlike classical ALS, upper motor neuron signs may be minimal or delayed, and disease progression is typically slower, with prolonged survival in many patients [3,5]. It accounts for approximately 5–8% of ALS cases and shows male predominance compared with other phenotypes [3,5–7].

FAS is characterized by progressive proximal upper limb weakness with relative sparing of bulbar and lower limb function during the early stages. Previous studies have shown a marked male predominance, slower disease progression, and longer survival compared with classical limb-onset ALS, supporting its recognition as a distinct regional ALS phenotype.

This case illustrates the prolonged diagnostic delay frequently encountered in FAS (24 months in our patient). The main contributing factor is the frequent coexistence of cervical degenerative changes, which may be incorrectly assumed to explain symptoms. However, key red flags include progressive bilateral weakness, lack of sensory involvement, and a poor correlation between imaging and clinical severity [2]. Differentiation from plexopathy,

myopathy or immune-mediated motor neuropathies, particularly multifocal motor neuropathy, is also essential, as these disorders may respond to immunotherapy. The absence of conduction block, lack of sensory involvement, progressive course, and diffuse denervation support ALS rather than multifocal motor neuropathy or other treatable conditions [2].

Although hyperreflexia is commonly associated with ALS, the patient exhibited absent deep tendon reflexes despite bilateral extensor plantar responses, indicating corticospinal tract involvement. This pattern is compatible with ALS, where severe lower motor neuron degeneration may abolish tendon reflexes despite concomitant upper motor neuron dysfunction. Such clinical dissociation has been described in lower motor neuron-predominant ALS phenotypes, including FAS, in which upper motor neuron signs may be subtle or partially masked by extensive lower motor neuron loss [8].

In the present case, EMG demonstrated widespread denervation beyond the clinically affected cervical region, including bulbar and lumbosacral muscles, providing decisive evidence of multisite motor neuron involvement. These findings are consistent with current diagnostic frameworks that emphasize combined clinical and electrophysiological evidence across multiple anatomical regions, particularly in regional ALS variants where subclinical involvement may precede clinical spread [1,9,10].

Although disease-modifying therapy remains limited, timely recognition of FAS enables appropriate prognostic counseling, anticipatory planning, and access to multidisciplinary care, which improves quality of life and may prolong survival [11]. It also prevents unnecessary immunological or surgical interventions that may carry risks without benefit. This case highlights the importance of maintaining a high index of suspicion for ALS variants in patients with progressive isolated upper limb weakness and underscores the pivotal role of electrophysiological evaluation in establishing the diagnosis.

Conclusion

FAS represents an uncommon but clinically significant variant of ALS that may mimic structural or peripheral neuromuscular disorders. Progressive bilateral upper limb weakness with muscle atrophy and fasciculations, combined with electrophysiological evidence of multisite denervation, should prompt consideration of this diagnosis even when imaging shows degenerative cervical changes. Accurate recognition of this phenotype can reduce diagnostic delay and improve patient management.

What's new?

This case illustrates the diagnostic challenges of FAS in a patient whose symptoms were initially attributed to cervical degenerative disease despite progressive bilateral brachial

weakness. It highlights the decisive role of electrophysiological studies in demonstrating multisite motor neuron involvement, excluding alternative diagnoses, and establishing FAS according to the recent ALS diagnostic criteria.

List of Abbreviations

ALS	amyotrophic lateral sclerosis
CMAP	compound muscle action potential
CSF	cerebrospinal fluid
CT	computed tomography
EMG	electromyography
FAS	flail arm syndrome
FASC	fasciculation potentials
FIB	fibrillation potentials
FVC	forced vital capacity
LMN	lower motor neuron
MCV	motor conduction velocity
MRC	Medical Research Council
MRI	magnetic resonance imaging
MUP	motor unit potential
NCS	nerve conduction studies
PSW	positive sharp waves
SNAP	sensory nerve action potential
UMN	upper motor neuron

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Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

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Ethical approval

Ethical approval was not required by our institution for the reporting of individual cases or case series.

Informed consent

Informed consent for publication of the case details and accompanying clinical photographs was obtained from the patient.

Guarantor

Yassine El Adraoui accepts full responsibility for the content of this case report.

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Summary of case

1	Patient (gender, age)	M, 69y
2	Final Diagnosis	Flail arm syndrome, amyotrophic lateral sclerosis
3	Symptoms	Progressive upper limb weakness and muscle wasting
4	Medications	Riluzole
5	Clinical Procedure	Electromyography
6	Specialty	Neurology