

Anti-Ro52 Positive Idiopathic Inflammatory Myopathy Presenting as Dropped Head Syndrome

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Abstract: *Idiopathic inflammatory myopathies are acquired autoimmune muscle disorders in which serological profiling is central to diagnosis and prognosis. Anti-Ro52 (TRIM21) is the commonest myositis-associated antibody, but is usually detected along with a myositis-specific antibody and is strongly linked to interstitial lung disease. Isolated anti-Ro52 positivity, without any myositis-specific antibody or pulmonary involvement, is uncommon. We report a 40-year-old woman with ten months of insidious, painless, progressive proximal weakness of all four limbs and severe neck extensor weakness producing a dropped head. Examination showed symmetrical proximal weakness of grade 1 to 3 on the Medical Research Council scale, proximal wasting, hypotonia, preserved reflexes and normal sensation. Serum creatine kinase and aldolase were repeatedly normal, and nerve conduction and repetitive nerve stimulation studies unremarkable. Antinuclear antibody was 3+ speckled, and an extended myositis immunoblot was positive only for anti-Ro52 at 3+. Whole-body magnetic resonance imaging showed patchy T2 and STIR hyperintensity with symmetrical fatty infiltration of paraspinal, shoulder girdle, iliopsoas, gluteal and thigh muscles, suggestive of inflammatory myopathy. Chest computed tomography, pulmonary function testing, echocardiography and positron emission tomography revealed no interstitial lung disease or occult malignancy. She improved on corticosteroids and mycophenolate mofetil. Inflammatory myopathy should not be excluded on the basis of a normal creatine kinase alone.*

Keywords: Anti-Ro52 antibody; Idiopathic inflammatory myopathy; Dropped head syndrome; Normal creatine kinase; Muscle magnetic resonance imaging.

1. Introduction

Idiopathic inflammatory myopathies (IIM) are acquired, immune-mediated muscle diseases that classically present with symmetrical proximal weakness and may also involve the skin, joints, lungs, heart and gastrointestinal tract. Beyond polymyositis, dermatomyositis and inclusion body myositis, immune-mediated necrotising myopathy and the antisynthetase syndrome are now recognised subgroups (Lundberg et al., 2017). Autoantibodies in IIM are grouped into myositis-specific antibodies, which define distinct phenotypes, and myositis-associated antibodies, which also occur in other connective tissue diseases. Anti-Ro52, directed against tripartite motif-containing protein 21, is the commonest myositis-associated antibody and is reported in nearly half of all patients with IIM (Pan et al., 2026). It usually accompanies anti-Jo-1, anti-MDA5 or other synthetase antibodies and predicts interstitial lung disease (ILD) and, in some subgroups, a poorer outcome (McHugh, 2023). Isolated anti-Ro52 positivity with normal muscle enzymes and without lung involvement is far less familiar. We report such a patient who presented with a dropped head.

2. Case Report

A 40-year-old right-handed woman presented with difficulty in using all four limbs and in holding her neck upright for ten months. She first noticed that she could not keep her neck erect for a prolonged period and needed intermittent support. Over the preceding three months this worsened, so that her

chin fell on to her chest while sitting or standing and she could not lift her head off the pillow.

Eight months before presentation she developed insidious, slowly progressive difficulty in getting up from a squat, climbing stairs, rolling over in bed and rising from a chair. Within four months the upper limbs were involved, with difficulty in combing her hair, lifting objects on to overhead shelves and raising a water bottle to her mouth, although her grip remained good. By ten months she had difficulty in bringing food to her mouth. There was no pain, diurnal or exertional variation, fatigability, cramps, twitching or dark urine after exertion, and no dysphagia, nasal regurgitation, diplopia, ptosis, breathlessness or sensory, bladder or bowel disturbance. She reported loss of weight and an itchy rash over the back and arms. There was no drug or toxin exposure, statin intake, malignancy, thyroid disease, joint pain, sicca symptoms, antecedent fever or similar illness in the family. Physiotherapy and homeopathic treatment had not helped.

On examination she was conscious, oriented and afebrile. Blood pressure was 130/70 mmHg, pulse 98/minute, respiratory rate 16/minute, saturation 97% on room air and body mass index 19.2 kg/m². There was no pallor, lymphadenopathy, thyromegaly, parotid enlargement or neurocutaneous marker. Hyperkeratotic follicular lesions over the arms were attributed by a dermatologist to xerosis; there were no Gottron papules or heliotrope rash. Higher mental functions and cranial nerves were normal except for weakness of the left sternocleidomastoid. There was generalised proximal wasting with hypotonia of all four

limbs, and neck extensors were weaker than flexors (Table 1). Reflexes were preserved and symmetrical, plantars flexor, and sensory and cerebellar examinations normal. Gait could not be tested. Systemic examination was unremarkable.

The clinical picture was thus a subacute, gradually progressive, pure motor illness with proximal weakness and wasting of all four limbs and marked neck extensor weakness, localising to muscle, neuromuscular junction or anterior horn cell. Idiopathic inflammatory myopathy, myasthenia gravis, congenital myasthenic syndrome, endocrine and metabolic myopathy, limb girdle muscular dystrophy and adult-onset spinal muscular atrophy were considered.

Table 1: Muscle power at presentation (Medical Research Council grading)

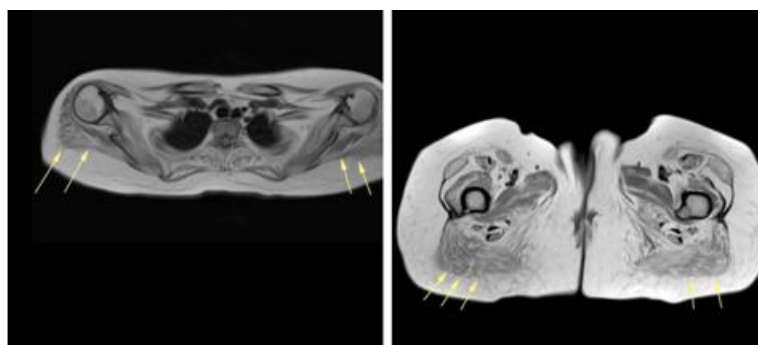
Region	Movement	Right	Left
Neck	Extension/flexion	1/5	1/5
Shoulder	Flexion/extension/abduction/adduction	2/5	2/5
Shoulder	Internal/external rotation	1/5	1/5
Elbow	Flexion/extension	3/5	3/5
Wrist	Dorsiflexion	4-/5	4-/5
Wrist	Palmar flexion	4+/5	4+/5
Hip	Flexion/extension/adduction	1/5	1/5
Hip	Abduction	2/5	2/5
Knee	Flexion	3/5	3/5
Knee	Extension	4-/5	3/5
Ankle	Dorsiflexion	4+/5	4-/5
Ankle	Plantar flexion	5/5	5/5

Investigations are summarised in Table 2. Serum creatine kinase, measured on two occasions, and serum aldolase were normal. Antinuclear antibody was 3+ in a speckled pattern, and an extended myositis immunoblot was strongly positive (3+) for anti-Ro52 alone, all myositis-specific and other myositis-associated antibodies being negative. Nerve conduction studies and repetitive nerve stimulation were normal and acetylcholine receptor antibody was negative. Whole-body magnetic resonance imaging showed patchy heterogeneous T2 and STIR hyperintensity with diffuse symmetrical fatty infiltration of the bilateral paraspinal,

shoulder girdle, iliopsoas, gluteal and thigh muscles, reported as suggestive of inflammatory myopathy (Figure 1). Chest computed tomography, pulmonary function testing, echocardiography and positron emission tomography were unremarkable. Muscle biopsy was deferred as the patient declined the procedure.

Table 2: Summary of investigations

Investigation	Result
Haemogram	Hypochromic microcytic anaemia; ESR 34 mm/hr
Serum creatine kinase ($\times 2$), aldolase	Normal
Renal, liver, thyroid profile, TPO antibody, electrolytes	Normal
CRP, VDRL, HBsAg, anti-HCV	Negative
Antinuclear antibody (IIF)	3+, speckled pattern
Extended myositis and ANA immunoblot (Mi-2, TIF1 γ , MDA5, NXP2, SAE1, Ku, PM-Scl, Jo-1, SRP, PL-7, PL-12, EJ, OJ, cN-1A, HMGR, SS-A, SS-B, Scl-70, dsDNA)	All negative
Anti-Ro52	3+, strongly positive
Anti-acetylcholine receptor antibody	Negative
Nerve conduction study (all four limbs), repetitive nerve stimulation	Normal
Chest CT, pulmonary function test, echocardiography, ultrasound abdomen	Normal; no interstitial lung disease
Whole-body PET-CT	Fatty infiltration of paraspinal muscles; no occult malignancy
Whole-body MRI (muscle)	Patchy T2/STIR hyperintensity with symmetrical fatty infiltration of paraspinal, shoulder girdle, iliopsoas, gluteal and thigh muscles; suggestive of inflammatory myopathy



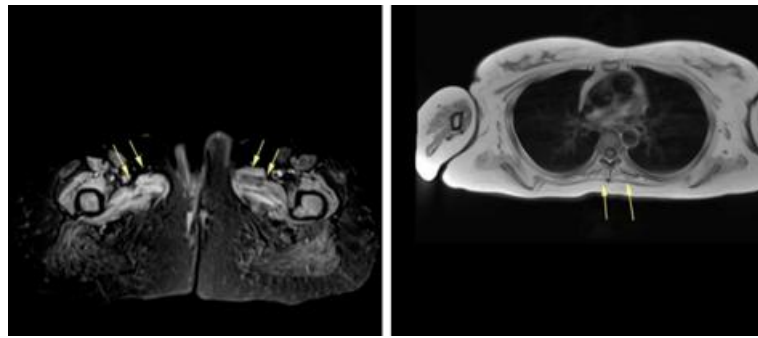


Figure 1: Axial T1-weighted and STIR whole-body magnetic resonance images showing patchy hyperintensity with symmetrical fatty infiltration (arrows) of the shoulder girdle, paraspinal, gluteal and thigh muscles

A diagnosis of anti-Ro52 associated idiopathic inflammatory myopathy was made. She was started on oral prednisolone and later switched to steroid sparing agent mycophenolate mofetil, along with calcium, vitamin D, gastric prophylaxis and supervised physiotherapy. At follow-up she showed clear improvement in neck holding and proximal power, and remains on regular clinical and pulmonary surveillance.

3. Discussion

Three features make this patient instructive. The first is the normal creatine kinase, often taken as evidence against active myositis. Normal or marginally raised enzymes may, however, be seen in chronic disease, when much of the muscle is already replaced by fat, or at the amyopathic end of the spectrum, and the EULAR/ACR criteria can still be satisfied without enzyme elevation (Lundberg et al., 2017). Here the diagnosis rested on the clinical pattern, serology and imaging rather than biochemistry.

The second is the value of muscle magnetic resonance imaging. Whole-body imaging demonstrated active myoedema with fatty replacement in exactly the muscle groups that were clinically weak, and would have guided biopsy had the patient consented. With normal enzymes and electrophysiology, imaging provided the only objective evidence of muscle inflammation.

The third is the antibody profile. Anti-Ro52 is the commonest myositis-associated antibody, found in about 46% of patients with IIM and most often in anti-MDA5 positive dermatomyositis and the antisynthetase syndrome (Pan et al., 2026). Its main clinical importance lies in its association with ILD. A meta-analysis of 59 studies found that anti-Ro52 positivity roughly trebled the odds of ILD in IIM and was also linked with rapidly progressive ILD (Nayebirad et al., 2023), and a hospital cohort reported an eight-fold rise in ILD risk with anti-Ro52 alone, increasing sharply when it coexisted with antisynthetase antibodies (Weng et al., 2024). Similar associations are described in connective tissue disease generally (Ferreira et al., 2012) and with objective sicca features (Meinecke et al., 2026). Our patient had neither sicca symptoms nor pulmonary abnormality. As anti-Ro52 is an associated rather than a specific antibody, it cannot by itself establish myositis; the diagnosis rested on a compatible syndrome, supportive imaging, exclusion of mimics and response to immunosuppression.

Dropped head as the dominant presenting feature also deserves emphasis. Neck extensor weakness occurs in inflammatory myopathy, myasthenia gravis, motor neuron disease and isolated neck extensor myopathy. Here the absence of fatigability, normal repetitive nerve stimulation and negative acetylcholine receptor antibody excluded a junctional disorder, while preserved reflexes without distal wasting or fasciculations argued against anterior horn cell disease. Absence of ILD at presentation is not permanently protective, and long-term surveillance with symptom review, pulmonary function testing and imaging when indicated remains essential (McHugh, 2023).

4. Conclusion

Anti-Ro52 associated inflammatory myopathy should be considered in subacute, progressive, painless proximal weakness with dropped head, even when the creatine kinase is normal. Muscle magnetic resonance imaging can reveal active myositis when enzymes and electrophysiology are unhelpful, and the myositis immunoblot may be positive only for anti-Ro52. Such patients respond well to corticosteroids with a steroid-sparing agent, but need sustained follow-up for ILD, which may appear later in the illness.

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Consent: Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

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