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CASE REPORT

SLE/ Polymyositis Overlap Syndrome

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ABSTRACT

Inflammatory myopathies (IM) is a rare inflammatory muscle disorder, which can be broadly divided into 5 subgroups. The accurate diagnosis of subtype of IM can be challenging due to a diverse presentation of the disease. On the other hand, skeletal muscle complication is common in patients with systemic lupus erythematosus (SLE) in the form of myalgia or myopathy. Inflammatory myopathy is a rare association of SLE and the diagnosis and treatment can be quite challenging. A 43-year-old lady with underlying systemic lupus erythematosus (SLE), presented with subacute onset progressively worsening muscle weakness involving upper limbs and lower limbs. Neurological examination showed findings consistent with proximal myopathy, with proximal power of 3/5 and distal power of 4/5. She has elevated creatinine kinase, ALT and AST level. Her myositis-specific autoantibodies were positive for anti-Ku antibodies. Her electromyography showed evidence of active myopathy of the upper and lower limb. Here, we would like to report a case of polymyositis in a patient with SLE.

INTRODUCTION

Inflammatory myopathies (IM) is a heterogeneous systemic muscle disorder that can affect multiple organs apart from muscle and often enough, it leads to significant morbidity to the patients[1]. Traditionally, IM was classified into 2 entities, namely dermatomyositis (DM) and polymyositis (PM)[2].

With the improvement in understanding of muscle pathology and introduction of myositis-specific antibodies (MSA), the understanding of IM has largely evolved[3]. At present, IM can be classified into 5 subgroups, namely, (1) polymyositis, (2) dermatomyositis, (3) immune-mediated necrotizing myopathy, (4) sporadic inclusion-body myositis and (5) overlap myositis (including antisynthetase syndrome)[4]. On the other hand, overlap syndrome is defined by the presence of two or more connective tissue diseases, primarily systemic lupus erythematosus (SLE), systemic sclerosis and IM[5].

CASE PRESENTATION

A 43-year-old lady with underlying systemic lupus erythematosus (SLE), presented with subacute onset progressively worsening muscle weakness involving upper limbs and lower limbs for 2 months duration. She also noticed difficulties in climbing stairs and stand from sitting position. Otherwise, she did not have dysphagia, dysphonia, difficulty in breathing or Raynaud's phenomenon. There was no numbness, headache or back pain. She also did not complain of urinary or bladder incontinence. There were no skin rashes, no symptoms of hyperthyroid or hypothyroidism. She does not consume alcohol or other supplements.

On examination, her blood pressure was 113/69 mmHg, pulse rate was 107 bpm, and the temperature was 37°C. There were no rashes, no oral ulcers and her nails were normal. She had marked alopecia with multiple areas of scarring. Neurological examination showed findings consistent with proximal myopathy, with proximal power of 3/5 and distal power of 4/5. Her reflexes and sensory examinations were normal. There was no fatigability and her cranial nerve were normal. Respiratory and cardiovascular examinations were normal.

She was then admitted for further investigation. Her full blood count showed normochromic normocytic anaemia with haemoglobin of 10.9 g/dl. Her renal function test was normal. She has elevated creatinine kinase level of 5976 U/L, ALT of 127 U/L, AST of 268 U/L (Table 1).

Table 1 Investigations of patient

	Day 1 of treatment	Day 5 of treatment	Day 19 of treatment	Day 30 of treatment	Day 48 of treatment	Day 86 of treatment	Day 99 of treatment	Normal value	Unit
Hb	10.9	11.4	12.4	11.6	13.4		13.3	13 – 18	g/dl
TWBC	5.4	7.9	11.7	9.83	11.8		13.1	4.0 – 10.0	10^9/L
Platelet	280	390	252	368	367		313	150 – 400	10^9/L
ESR	81		80						
Sodium	138		138	142	139	141	138	135 – 148	mmol/L
Potassium	4		4.3	3.7	4.6	4.4	3.9	3.5 – 5.1	mmol/L
Urea	3.5		3	2.5	4.7	1.9	3.3	2.8 – 7.8	µmol/L
Creatinine	41		21	23	29	25	29	61 – 110	mmol/L
Tbili	8.1	4.7	12.61	9.8	11.24	12.6	13.2	0 – 17	µmol/L
ALT	127	129	125	110	93	40	34	0 – 31	U/L
ALP	60	55	50	44	70	50	48	35 – 104	U/L
Alb	35	35	39	37	42	41	41	34 – 48	g/L
Globulin	52	49	34	35	38	31	32	20 – 35	g/L
AST	268	93	160	149	95	61	43	10 – 40	U/L
CK	5,976	1,146	2,740	3,345	1,559	1,185	717	22 – 198	U/L
LDH			712	646	536	359	327	140 – 280	U/L
SLE workup				Extractable nuclear antigen (ENA)			Myositis Specific antibodies (MSA)		
ANA	Positive 1:320			U1RNP	Negative		Mi-2	Negative	
C3	45	90 – 180 mg/dL		Anti Sm	Negative		PM-Scl100	Negative	
C4	9	10 – 40 mg/dL		SS-A	Negative		PM-Scl75	Negative	
Anti-DsDNA	Positive 1:40	<1:10		SS-B	Negative		SRP	Negative	
				Scl-70	Negative		PL-7	Negative	
T4	15.3	12.2 – 22.4		Jo-1	Negative		PL-12	Negative	
TSH	1.59	0.35 – 4.55					EJ	Negative	
							OJ	Negative	
Tumour markers:		Normal range					Ro52	Negative	
Alpha Feto protein	1.8	<10 ng/ml					Ku	Positive	
CEA	0.8	<2.5 ng/ml							
CA125	9.1	0 – 35 units/ml							
CA15-3	26.6	<30 U/ml							
CA19-9	13.6	0 – 37 U/ml							

Her extractable nuclear antigen (ENA) was negative, myositis-specific autoantibodies were positive for anti-Ku antibodies. Her electromyography showed evidence of active myopathy of the upper and lower limb (Table 2).

Table 2 Electromyography of the patient

Side	Muscle	Nerve	Root	Ins Act	Fibs	PSW	Fasc	Myotonia	Myokimia	CDR	Amp	Dur	Poly	Recrt	Int Pat	Interpretation
Right	Biceps	Musculo-cutaneous	C5-6	Incr	2+	2+	Nml	Nml	Nml	Nml	Decr	Decr	2+	Reduce	Nml	Proximal Myopathy
Right	Ext Dig Brevis	Dp Dr Fibular	L5,S1	Incr	2+	2+	Nml	Nml	Nml	Nml	Decr	Decr	2+	Reduce	Nml	Proximal Myopathy
Right	Deltoid	Axillary	C5-6	Incr	3+	2+	Nml	Nml	Nml	Nml	Decr	Decr	2+	Rapid	Nml	Proximal Myopathy
Right	Vastus Lateralis	Femoral	L2-4	Incr	3+	3+	Nml	Nml	Nml	Nml	Decr	Decr	2+	Reduce	Nml	Proximal Myopathy
Right	1st Dorsal Interosseous	Ulcer	C8-T1	Incr	Nml	Nml	Nml	Nml	Nml	Nml	Nml	Nml	0	Reduce	Nml	Myopathy

Abbreviation:
CRD: Complex Rep Disch
Amp: Amplitude
Dur: Duration
Fasc: Fasciculation potentials
Fib: Fibrillation potentials
Ins Act: Insertion activity
Recrt: Recruitment
PSW: Positive sharp waves

She was then diagnosed to have SLE/polymyositis overlap syndrome and treated with intravenous methylprednisolone 500 mg once daily for 3 days followed by oral prednisolone 1 mg/kg. She was discharged after a week in the hospital. One month after corticosteroids, her muscle power markedly improved. She was able to ambulate without help. Azathioprine was added as steroid-sparing agent and prednisolone was subsequently tapered down. She was keeping well since then. Her malignancy screening was also negative with negative whole-body computed tomography scan. Gynaecological and otorhinolaryngology evaluation also excluded malignancy.

DISCUSSION

SLE is an autoimmune disease that affects virtually any organs in the body. It affects females more commonly than males. Skeletal muscle complication is common in SLE patients, which is seen in the form of myalgia, muscle weakness and atrophy[6]. Myalgia is the most skeletal muscle manifestation, affecting 40 – 80% of patients[7]. On the other hand, muscle weakness can be caused by myopathy, neuropathy or central nervous system complications. Myopathy in SLE can be caused by a range of pathologies, such as endocrine, inflammatory, paraneoplastic or infectious etiologies[8]. Other differential diagnoses that we need to consider in SLE patient with muscle weakness include myasthenia gravis, lupus myositis or drug-induced neuromyotoxicity[9]. Hydroxychloroquine and corticosteroids are established treatment of SLE and muscle weakness are a well known side effect of the drugs. It is important to diagnose the underlying cause of muscle weakness as the treatment is different.

Inflammatory myopathies (IM) is a group of acquired myopathy characterized by muscle inflammation and motor weakness of varying severity[2]. Its incidence is estimated to be around 4.27 – 7.89 cases/100,000 population per year[10]. IM is broadly classified

into 5, namely polymyositis, dermatomyositis, immune-mediated necrotising myopathy, sporadic inclusion body myositis and overlap myositis[4]. Clinically, a patient can present with acute or subacute onset muscle weakness of different pattern, often accompanied by raised creatinine kinase (CK)[1]. In severe cases, respiratory and oesophageal muscles can be affected[11]. The diagnosis of IM requires careful clinical evaluation paired with serological markers, neurophysiological testing and muscle biopsy. In history taking, we need to take a relevant family history, myopathic drugs, alcohol and features of endocrinopathy[12]. Magnetic resonance imaging (MRI) of muscle can help in certain cases.

In this patient, she has underlying SLE, currently presented with subacute onset of proximal muscle weakness. There were no extraskeletal manifestations. Clinical examination was consistent with proximal muscle weakness. At this point of time, the possible differential diagnosis to consider for her weakness includes myopathy, pure motor peripheral neuropathy or transverse myelitis. Absence of sensory involvement excluded transverse myelitis. Elevation of creatinine kinase confirms the weakness is likely due to myopathy. Possible causes of myopathy include polymyositis, dermatomyositis, overlap myositis, myasthenia gravis or drug-induced neuromyotoxicity. We think that she most likely has polymyositis as she does not have any extraskeletal findings to suggest dermatomyositis. There was no fatiguability to suggest myasthenia gravis. There were also no features to suggest possible antisynthetase syndrome. Her anti-Ku antibodies positive, which can be seen in SLE with polymyositis. Electromyography was also consistent with the myopathic pattern. Muscle biopsy was not done as service was not available.

Inflammatory myopathy is rare in SLE patient with only 4 – 16% patients affected[13]. Both dermatomyositis (DM) and polymyositis (PM) can be associated with SLE with DM more commonly seen in SLE. PM commonly affects those age from infancy to late adulthood, with 40 – 60 years old more common[14]. Patients usually present with subacute onset proximal muscle weakness of both upper and lower limbs[13]. In the later stage of the disease, trunk muscles, pharyngeal muscles and respiratory muscle can be involved. Extramuscular features such as Gottron papules, Raynauds phenomenon are usually absent[1]. A laboratory test will show raised CK, aspartate aminotransferase and alanine transaminase. Antibodies such as extractable nuclear antigen (ENA), myositis specific autoantibodies (MSA) are helpful serological markers[12]. Characteristics electromyography include spontaneous fibrillation at rest or with needle insertion, spontaneous high-frequency discharges and positive sharp waves[15]. Importantly, we also need to screen the patient for malignancy[16].

The optimal treatment of IM remains a challenge due to its low prevalence and wide clinical heterogeneity. In general, immunosuppressive therapy with a corticosteroid and steroid-sparing agents remains the mainstay of treatment. Majority of patients respond well to immunosuppressive therapy. Induction of steroid with 1 mg/kg or pulse parenteral steroid can provide rapid response[17]. The steroid should be continued for 4 – 12 weeks and reduction should be considered based on the improvement of power strength and CK level[1]. Steroid sparing agent such as azathioprine or methotrexate should be started based on patient clinical profile[10]. Intravenous immunoglobulin can be given in those with the severe disease while rituximab and cyclophosphamide are options for refractory disease[1].

CONCLUSION

In conclusion, inflammatory myopathy is a rare inflammatory disorder with diverse and heterogeneous clinical presentation.

Diagnosis of IM requires thorough clinical evaluation, coupled with a laboratory test, electromyography and muscle biopsy. In approaching SLE patients with muscle weakness, IM needs to be excluded. Immunosuppressant therapy remains the mainstay of treatment for IM.

CONFLICT OF INTEREST

The authors declare that they have no competing interests in publishing this article.

CONSENTS

Written informed consent was obtained from the patient to publish this case. A copy of the written consent is available for review by the Chief Editor.

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