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Case Report

An uncommon cause of myalgia: A case report on systemic lupus erythematous myopathy

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ABSTRACT

Introduction: Myositis in systemic lupus erythematous may present in a wide range of clinical spectrum. It can be part of an overlap syndrome, or mixed connective tissue disease or a musculoskeletal manifestation of systemic lupus erythematous itself.
Case presentation: Here, we present a young girl with an underlying systemic lupus erythematous presented with the typical manifestation of severe proximal myopathy in the background of normal creatine kinase values. The diagnosis of systemic lupus erythematous myopathy was made after excluding other more common causes of myopathies which in itself is a very rare occurrence.
Discussions: A normal creatine kinase values does not exclude systemic lupus erythematous myositis, but make the diagnosis more challenging. However, there are other parameters or diagnostic tools which can be used to exclude a myositis.
Conclusion: This case elucidates the importance of history and physical examination in the face of some conflicting laboratory data.

1. Introduction

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease that is extremely heterogeneous in both its clinical and serological presentation [1]. Myositis is a rare presentation in SLE. Myositis is not a feature in the 2017 revised criteria according to American College of Rheumatology [2]. Myositis may have significant overlapping features of other conditions of similar aetiology, including mixed connective tissue disease and Sjögren syndrome or part of SLE musculoskeletal manifestation itself. Myopathies are well known to affect both the adult population and the paediatrics age group. The prevalence of SLE myositis in paediatrics may even be higher than the adults SLE patients. Additionally, the presenting features in children are often more severe than in adults, and these children have been shown to be twice as likely to require higher dose corticosteroids [3,4]. Primarily SLE patients may have muscle tenderness, myalgia secondary to inflammatory myositis, fibromyalgia or drug related myopathy. After excluding these myopathies, a SLE myositis should be considered as part of the lupus manifestation. Lupus myositis is relatively rare and further more with a normal creatine kinase. Other markers which may assist in diagnosis of myositis are thorough clinical evaluation alongside lab investigations which are serum Lactate dehydrogenase, aspartate transaminase, aldolase, myositis-specific antibodies, myositis-associated antibodies, proximal muscle magnetic resonance imaging, electromyography and biopsy. This case report describes a case of SLE who presents with proximal myopathy and a normal creatine kinase level. Myopathies associated with SLE may present with other associated conditions like rapidly progressive interstitial lung disease and bulbar involvement where thorough assessment for these conditions are crucial as it has prognostic significance. The possibility of myositis may have been missed due to the normal creatine kinase level and this may have caused the patient to deteriorate overtime.

A 14-year-old female, a known case of systemic lupus erythematous (SLE) presented to the emergency department with a complain of generalized body weakness for two weeks duration, which have progressively worsened and requires assistance with her activity of daily living. There were also multiple cutaneous vasculitis lesions on forearm, palms, legs and sole of the both foots. Otherwise, there was no other

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symptoms suggestive of SLE flare. She denied any symptoms of hypothyroidism, no family history of hereditary myopathies, and did not consume any traditional medications. She was diagnosed with SLE three months prior to presentation. Her initial clinical presentation was alopecia and mucocutaneous rash, and further supported with the immunological findings of positive anti-nuclear antibody, anti-ribosomal P antibody, anti U1-ribonucleoprotein antibody and anti-Ro antibody. She was on low dose oral steroid and hydroxychloroquine which she only managed to take for one month before she defaulted treatment due to the recent coronavirus pandemic.

On examination, there were multiple vasculitis lesions on the palm, sole, forearm and legs. There was mild muscle weakness over the proximal upper and lower limbs with the power grade about 3 over 5. There was no evidence of fatigability, no muscle wasting, no muscle tenderness and the reflexes and sensations were normal. The Manual Muscle Testing and a Subset of Eight Muscles (MMT8) score on admission was 98/150. Her initial laboratory investigations were taken and showed microcytic hypochromic anaemia without evidence of haemolysis in peripheral blood film and lymphopenia. Otherwise, the total white blood cell and platelet counts were normal. There was low C3 and C4 level, and slightly raised erythrocyte sedimentation rate. The thyroid, renal and liver function was normal. The myositis specific autoantibodies, myositis-associated autoantibodies and infective screening test was negative, and the creatine kinase and aspartate transaminase level were normal (as shown in Table 1). Electromyography and nerve conduction study were done which is consistent with irritative myopathy with active degeneration. Magnetic resonance imaging of the bilateral thigh showed oedema of the thigh muscles with adjacent subcutaneous oedema and mild knee joint effusion. She was started on intravenous (IV) hydrocortisone on day of admission. The muscle weakness and the rashes had only partial improvement after one week. Therefore, she was subjected to IV methylprednisolone 500mg once daily for three days. Her cutaneous lesions started to fade and her muscle power showed improvement with a MMT8 score of 121/150 after completion of the methylprednisolone. She was then switched to oral prednisolone tapering dose and started on oral azathioprine as the steroid sparing agent. She regained full muscle power and resolution of skin lesions two weeks after discharge.

Table 1
Initial blood investigations showed anaemia and lymphopenia. Otherwise, the total white blood cell and platelet counts were normal. There was low C3 and C4 level, and slightly raised erythrocyte sedimentation rate. The thyroid, renal and liver function was normal. The myositis specific and myositis-associated autoantibodies, and the viral infective screening test was negative. The creatine kinase and aspartate transaminase level were normal.

Blood parameters	Result	Normal Range
Haemoglobin	9.0 g/dL	12–18 g/dL
White blood cell	4.39 x 10^9/L	4.0–11.0 x 10^9/L
Lymphocytes	0.67 x 10^3/μL	1.0–3.0 x 10^3/μL
Platelet	218 x 10^9/L	150–400 x 10^9/L
Prothrombin Time	11.4 seconds	11–14.5 seconds
Albumin	28 g/L	35–50 g/L
Alkaline Phosphatase	68 U/L	50–150 U/L
Alanine Transaminase	57 U/L	5–35 U/L
Aspartate Transaminase	34 U/L	5–35 U/L
Total bilirubin	5.21 μmol/L	0–13 μmol/L
Creatinine	54.1 μmol/L	60–120 μmol/L
Sodium	142 mmol/L	135–150 mmol/L
Potassium	4.1 mmol/L	3.5–5.0 mmol/L
Urea	4.6 mmol/L	1.7–8.0 mmol/L
Corrected Calcium	2.2 mmol/L	2.15–2.55 mmol/L
Magnesium	0.81 mmol/L	0.66–1.07 mmol/L
Phosphate	0.98 mmol/L	0.75–1.50 mmol/L
Serum Thyroid Stimulating Hormone	3.55 iu/mL	0.4–4.0 iu/mL
Serum T4 level	14.97 pmol/L	11–22 pmol/L
Erythrocyte Sedimented Rate	30 mm/hr	0–20 mm/hr
C-reactive protein	0.7 mg/L	0–5 mg/L
C3 level	0.55	80–178 mg/dL
C4 level	0.09	12–42 mg/dL
Creatine Kinase	250 U/L	60–350 U/L
Myositis Specific antibodies	Negative	
Myositis-associated autoantibodies	Negative	
HbsAg/Anti-HCV/anti-HIV	Negative	

2. Discussion

The association between myositis and systemic lupus erythematous (SLE) had been explored before with the prevalence varies widely worldwide ranging from 3.4% to 16% of SLE patients [5,6]. Extensive muscle involvement in the form of severe weakness and wasting is not a common presentation in SLE associated myopathies [6]. The clinical presentation of purely SLE myositis resembles milder form of that which is described in other inflammatory myopathies. We have observed in this patient that her serial creatine kinase (CK) values were never elevated, which is a rare occurrence in SLE myositis. Creatinine kinase values appears to be a relatively sensitive and specific indicator of the degree of muscle fibre injury. However, the range varies significantly amongst patients, some in rare instances show normal values, and elevated by several hundred-fold in others. In this situation, the measurement of other serum muscle enzymes, including aldolase, aspartate transaminase, alanine transaminase, and lactate dehydrogenase, significantly assist in diagnosing myositis. Majority of cases with normal CK are seen especially in glucocorticoid induced myopathies, or in early polymyositis which has been excluded in the very beginning of this case. SLE and myositis may be complicated with presence of other overlap syndrome and may be difficult to categorize. Myositis and SLE may occur alone, together, or maybe seen in a context of more complex combined conditions [6].

In our patient, although there were no clinical features of mixed connective tissue disease, but the anti U1-ribonucleoprotein and anti-Ro autoantibodies were positive, which has been shown to be frequently associated with SLE myositis [3,6] The presence of this autoantibodies can help predict the muscle involvement and probable features of other overlap syndromes in future [7].

Although most cases of myositis are dependent on muscle biopsy [7], it is an invasive and a painful procedure which make is not routinely done in younger patients. Apart from that, our centre does not offer the services for muscle biopsy and usually we send our patients to another state for the procedure, which was not an ideal situation during the pandemic time of COVID-19. Therefore, we based our diagnosis on electromyography, magnetic resonance imaging thigh and clinical and laboratory definitions of myositis. ultrastructural pathology in patients with SLE includes inflammations along with muscle atrophy, microtubular inclusions, a mononuclear cell infiltrate and fibre necrosis.

Our patient initially was on intravenous hydrocortisone which did not respond too well in terms of regaining muscle power, subsequently we had to resort to intravenous methylprednisolone, which showed almost immediate improvement. This was similar to the case reported by Record JL et al. (2011) [3] which uses a higher dose of steroids to show any improvement. After two weeks on oral steroid, our patient regained full muscle power. Currently, she is well under regular rheumatology unit clinic follow up.

3. Conclusion

In conclusion, we illustrate a case of systemic lupus erythematous myopathy with an atypical biochemical feature that is normal creatine kinase values in a patient with proximal myopathy and in the absence of inflammatory myositis features, which may be more common than previously thought. Thus, a high clinical suspicion is important to identify and diagnose this condition particularly those with normal serum muscle enzymes.

Ethical approval

This case report does not need any ethical approval, but written informed consent from the patient and her guardian was taken to write and publish this case report.

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Authors contribution

Shahleni Paramasivam – drafting the article, gathering the team of authorship Malehah Mohd Noh – supervisor and final approval of the version Mya Sanda Khaing – data collection and interpretation of data Izdihar Marwani Dahlan - conception and design, or analysis and interpretation of data Alvin Oliver Payus –revising the article critically for important intellectual content, and final editing.

Research registration

Not available.

Guarantor

The Guarantor is the one or more people who accept full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

Guarantor

Alvin Oliver Payus.

Consent

The patient and her guardians have given their informed and written consent to the writing of this case report.

Provenance and peer review

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Declaration of competing interest

None declared.

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Appendix A. Supplementary data

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