

Case Report

Overlap Syndrome- Systemic Lupus Erythematosus Plus Dermatomyositis- A Rare Case Study

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ABSTRACT:

The incidence of myositis in patients with systemic lupus erythematosus (SLE) is low among different series. Here we attempt to presented the case report of a 35 year old patient with the chief complaint of darkening of skin of face and body since two years.

Key words: Myositis, Overlap Syndrome, Systemic Lupus Erythematosus, Dermatomyositis

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INTRODUCTION

The term overlap syndrome includes a large group of conditions characterized by the coexistence of signs, symptoms and immunological features of 2 or more connective tissue diseases and occurring simultaneously in the same patient.¹⁻³ Myositis (polymyositis PM or dermatomyositis DM) identifies a group of patients in whom the muscular weakness is the principle clinical feature often associated with muscle pain, tenderness and wasting, or other form of connective tissue diseases; the muscle biopsy generally demonstrates areas of muscle fiber necrosis accompanied by interstitial and/or perivascular cellular infiltrates.⁴⁻⁶

Hence; in the present paper; we have presented the case report of a 35 year old patient, who reported to the department of medicine with the chief complaint of darkening of skin of face and body since two years.

CASE REPORT

A 35 year old male patient reported to the department of medicine opd with the chief complaint of darkening of skin of face and body since two years. Patient also gave history

of presence of lesion on the elbow and shoulders. Clinical symptoms reported by the patient included joint and bone pain, difficulty in standing up from sitting position. Patient gave positive history of drinking alcohol and smoking. Clinical presentation of the lesions included well defined hyper-pigmented atrophic plaques with hypo-pigmented areas in the center. Oral lesions included well defined erosions. Laboratory investigations were carried out. ELISA reports show positive serum antinuclear antibody values indicating the presence of systemic rheumatic diseases. Patient was also found to be positive for anti-ds-DNA on testing with indirect immunofluorescence, indicating presence of Systemic Lupus Erythematosus (SLE), Serum Aldolase was also positive. Ultrasonography of abdomen showed presence of splenomegaly. A decrease in the values of WBC, hemoglobin and platelet count was seen. Based on the clinical profile and investigation reports of the patient, a confirmed diagnosis of overlap syndrome-SLE plus dermatomyositis was given.



Figure 1: Reduced muscle bulk and foot drop (due to dermatomyositis)



Figure 2: Loss of tongue papillae



Figure 3: Multiple rash



Figure 4: Malar rash



Figure 5: Shawl sign

DISCUSSION

The existence of patients with signs and laboratory tests results suggestive of a systemic autoimmune disease but fulfilling more than one classification criteria for well-defined connective tissue disease is a more and more frequent situation in clinical practice and define an overlap syndrome. In the literature, and because of the diversity of clinical symptoms, reliable data concerning the prevalence of overlap syndromes are not available. However, those patients appear to occur less frequent than patients with SLE, more frequent than patients with systemic sclerosis (SSc) or idiopathic inflammatory myositis (IIM). In a longitudinal study of 100 patients with IIM, Troyanov et al. found a frequency of 24% of myositis associated with connective tissue disease according to the original classification. When using its novel classification of myositis, the author found that 60% of their patients were classified as having overlap myositis, and that Systemic sclerosis was the most common disease associated with IIM. In the literature, SLE associated with myositis occurs in 4–16% of cases. In contrast to myalgia which can affect nearly half of patients with SLE. Myositis can occur before, after SLE, or sporadically both diseases can be present simultaneously.⁷⁻¹⁰

A photosensitive rash often is the initial manifestation; it may precede muscle disease by more than a year. The severities of the rash and muscle disease may be parallel or follow disparate courses. Gottron papules (seen in 60% to 80% of patients) and the heliotrope rash (seen in fewer than 50% of patients) are pathognomonic for DM; many other cutaneous lesions are characteristic but not as specific. Lower extremity weakness often manifests first, with

difficulty in climbing stairs or rising from a chair or toilet. Upper extremity weakness leads to difficulty in combing or washing hair.¹¹

In the primary autoimmune myositis, the ANA were less commonly positive and the presence of a strongly positive ANA may lead to a more diligent search for an associated autoimmune rheumatic disease.³ However, patients can have a high frequency of specifically associated autoantibodies to nuclear and cytoplasmic antigens, termed myositis specific antibodies (MSA). MSA are found almost in patients with DM/PM and associated overlap syndromes. Autoantibodies detected in myositis associated overlap syndromes also include anti-U1 RNP, anti-Ro/SSA, anti-La/SSB, and anti-Sm. In general, the picture of overlap syndromes is complex and heterogeneous. The presence of specific autoantibody profiles is certainly a useful tool in the diagnosis evaluation of such patients.¹²

The following disorders are considered to be "undifferentiated" rheumatic diseases and/or "overlap syndromes".

- A. Mixed connective tissue disease
 - Lupus-scleroderma-polymyositis-rheumatoid arthritis
- B. Undifferentiated systemic rheumatic disease
- C. Non-classic systemic rheumatic disease
- D. Overlap syndromes
 - Rheumatoid arthritis-lupus
 - Scleroderma-polymyositis/dermatomyositis
 - Scleroderma-lupus
 - Scleroderma-rheumatoid arthritis
 - Polymyositis overlaps
 - Juvenile idiopathic arthritis-lupus
 - Sjögren's syndrome overlaps
- E. Others
 - Undifferentiated polyarthritis syndrome
 - Undifferentiated spondyloarthritis¹³

Hematologic abnormalities are a common finding in MCTD. Anemia is found in 75% of patients, and the usual profile is most consistent with the anemia of chronic inflammation. A positive Coombs test is seen in about 60% of patients, but an overt hemolytic anemia is uncommon. As in SLE, a leukopenia affecting mainly the lymphocyte series is seen in about 75% of patients and tends to correlate with disease activity. Less common associations have been thrombocytopenia thrombotic thrombocytopenia purpura and red cell aplasia. Hypocomplementemia has been described in several studies⁶³; it is not as prevalent as in classic SLE and has not been correlated with any particular clinical situation. Positive tests for RF have been found in about 50% of patients.^{11, 12} Although the responsibility of autoantibodies themselves in the pathogenesis of idiopathic inflammatory myopathies is still uncertain, these may represent an increased risk of characteristic clinical associations.

CONCLUSION

Overlap syndromes happen regularly in patients with rheumatic pathologies. Therefore, whenever a patient does not fit into one specific pathologic group, broad work up including autoimmune and histopathological surveys should be started and then tried to determine if they fit into any of the overlap syndromes.

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