BJMHR
British Journal of Medical and Health Research
Journal home page: www.bjmhr.com

Importance of follow up in the diagnosis of Overlap Myositis – A short communication

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ABSTRACT

Overlap myositis are the group of conditions characterized by varying degree of muscle weakness in an underlying connective tissue disorder like Systemic lupus erythematosis, Systemic sclerosis and Rheumatoid arthritis . Overlap myositis presents subacutely and muscle weakness may not be present at the time of presentation of the underlying connective tissue disease. Many cases tend to have vague clinical features which makes the diagnosis more difficult. The myositis tends to be diagnosed during follow up of these diseases. Here we present three cases in which patients developed muscle weakness later than the initial presentation of the primary disease. Hence we emphasize the importance of regular follow up of the patients having connective tissue disorder and to specifically enquire about muscle pain and weakness. This will help in the early diagnosis of overlap myositis.

Keywords: Overlap myositis, Follow up, Systemic sclerosis, Muscle biopsy.

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25 November 2017, Accepted 08 December 2017

Please cite this article as: Kulkarni A *et al*., Importance of follow up in the diagnosis of Overlap Myositis – A short communication. British Journal of Medical and Health Research 2017.

INTRODUCTION

Myositis-overlap syndromes are characterized by group of clinical syndromes of which many are closely linked with specific autoantibodies. [1] Overlap myositis is a form of idiopathic inflammatory myopathy characterized by myositis with at least one clinical and/or autoantibody overlap feature. Prevalence and annual incidence of overlap myositis are not known. Estimates are difficult to determine because of low recognition levels . Patients with myositis and one clinical and/or autoantibody overlap feature are considered to have Overlap myositis. These disorders usually present subacutely with multisystem involvement. The pattern of organ involvement reflects the characteristic features of the well-defined rheumatic diseases occurring together with muscle involvement. For many patients, non-specific symptoms are often present for many months before a diagnosis is made. Diagnostic suspicion should be raised when patients present with muscle pain and weakness with simultaneous feature of other rheumatological disease. Apart from mixed connective tissue disease (MCTD) , these are best considered in the context of specific clinical features, auto-antibody profiles, and immunogenetics. In many instances it has been found that the features of myositis may not be present at the time of presentation but may develop consequently. In such instances the continuous follow up of such patients helps in the recognition of overlap myositis. The most common scleroderma overlap syndromes are mixed connective tissue disease (MCTD), scleromyositis and synthetase syndrome. [2] Here we present three such cases where the features of myositis appeared much later than the primary disease.

CLINICAL SUMMARY

Case 1

A 49 year old male patient presented with history of thickening and tightening of skin around his fingers, hand, upper limb, chest and face since 1 year. He also gave history of regurgitation on lying supine since 5 months. On examination, there was tightening of skin over the upper limbs, chest, back and face. There were a few lesions of calcinosis over knuckles. Hemoglobin - 16.2 g/dL; Total Leukocyte Count - 7420 cells/mm^3.Differential Leukocyte Count was normal; ESR - 13mm/hr; Renal function tests - Normal; Anti Nuclear Antibody by immunoflourescence - Positive ++ (nucleolar pattern); Anti- Histone, Anti Centromere, Anti-topoisomerase antibodies were negative; Skin biopsy - Dermis shows thick homogenous collagen bundles and perivascular and periadnexal lymphoplasmacytic cells (features consistent with systemic sclerosis) HRCT chest- Normal; Pulmonary Function tests - reveal moderate restrictive lung disease;. . Based on the 2015 scoring system by American College of Rheumatology/European League Against Rheumatism criteria for the classification of systemic sclerosis, this patient has a score of 11 (score above 9 is diagnostic

of systemic sclerosis). After 3 months, he started developing weakness of neck muscles. He has difficulty holding his head up right. He also had mild weakness of all 4 limbs. There was weakness of neck muscles and power in upper limbs and lower limbs was 4/5. Deep tendon reflexes were intact. Investigations revealed: Creatinine Kinase -1500 U/L which was high; Muscle biopsy (sternocleidomastoid) - showed peri fasical atrophy and diffuse infiltration of muscle fibres with perivascular infiltrates consistent with dermatomyositis; EMG- features suggestive of myopathic pattern. This is the case over overlap between Systemic sclerosis and dermatomyositis. In this case muscle weakness was not present at the time of presentation but developed later which was diagnosed during follow up.

Case 2

A 45yrs old female presented to hospital with chief complaints of low grade fever since 2 months , diffuse alopecia . There was diffuse thickening, edema and erythema of her skin. Total leukocyte count: 11240cells/cumm. Platelet count: 3.47 lakh/cumm, Serum creatinine: 0.6mg/dl, Na+: 139mEq/dl, k+: 4.5mEq/dl ESR- 90mm, Chest X-ray was normal, ANA-IF: 4+ positive(speckled pattern) ANA profile : positive for Anti ds-DNA , PM-Scl antibodies. Urine routine was normal. In view of above clinical features and auto-antibody profile , she was diagnosed as SLE . After 15 days, the patient developed muscle pain and weakness manifested as difficulty in combing hair, carrying food to mouth, climbing stairs and later progressed to involve distal muscles. Nervous system examination : Power in muscle of upper and lower limb was 3/5. Deep tendon reflexes: 2+. No sensory involvement. Investigations: Creatinine phosphokinase: 7973U/L, Serum Anti-Jo antibodies was positive. Electromyogram: suggestive of myopathic pattern. Muscle biopsy: there were areas of necrosis, regeneration and inflammatory cells infiltrated around muscle fibres predominantly CD8+ T

lymphocytes suggesting inflammatory myopathy. In this case patient had an overlap of systemic lupus erythematosus and polymyositis.

Case 3

A 40 yrs old female presented to hospital with complaints of fever since 1 month. On examination she had malar rash, diffuse aloepecia and multiple cervical lymphadenopathy. Biopsy of lymphnode was suggestive of Kikuchi Disease. ANA – IF: POSITIVE (NUCLEOLAR PATTERN). Anti ds DNA antibodies positive. She was diagnosed to have SLE based on ACR Criteria. .1 month later the patient presented with myalgias and weakness of all 4 limbs (Proximal>>Distal). On examination she had symmetric proximal muscle weakness. Serum CPK levels were 2500 and EMG showed Myopathic pattern of weakness. Muscle biopsy could not be done as patient did not give consent. In this case patient was

diagnosed to have SLE and Kikuchi's disease. However after 1 month she developed involvement of muscles.

DISCUSSION

Overlap myositis syndrome is a form of idiopathic inflammatory myopathy characterized by myositis with at least one clinical and/or autoantibody overlap feature. Estimates are difficult to determine because of low recognition levels of this form of disease. Overlap myositis is a clinically heterogeneous, poorly recognized subtype of inflammatory myopathy. Usual clinical overlap features include polyarthritis, Raynaud phenomenon, sclerodactyly, scleroderma , lung interstitial pneumonia, and/or clinical signs of systemic lupus erythematosus. Autoantibodies associated with overlap myositis /syndrome are Anti Pm-Scl , Anti Ku , Anti Synthetase antibodies [(7)].The distinction between classic Polymyositis and Dermatomyositis and an overlap has prognostic and therapeutic significance. The above cases satisfied the Bohan's and Peter's criteria for the diagnosis of myositis. In this case there is clinical overlap of scleroderma and myositis. Overlap myositis in males is very rare which was seen in our first case. Patients of SLE associated with myositis had more incidence of alopecia, oral ulcers, pulmonary disease. [(8)] . According to Bhansing et al pulmonary fibrosis and cardiac manifestations were higher in patients of overlap myositis compared with systemic sclerosis. [(3)]. According to James B Lilleker et al incidence of dysphagia was more common in patients of overlap myositis. [(4)] According to a study conducted by E C Bury et al 40-50% of patients of overlap myositis had Pm-Scl autoantibody. [(5)] According to Pia Moinzadeh et al patients of overlap myositis developed musculoskeletal involvement , muscle weakness and muscle atrophy.[(6)] Here we want to emphasize that in all the three cases feature of myositis were not present at the time of diagnosis of the primary disease but developed later.

CONCLUSION

Diagnosis of overlap myositis requires high index of suspicion. Clinical features may be subtle at the time of diagnosis . In many cases muscle weakness may not be present at the time of presentation of the primary connective tissue disease and may appear later. Hence, regular follow up of such cases is required for the diagnosis overlap myositis. During follow up detailed clinical examination, musculoskeletal and neurological examination should be done. And if necessary serum CPK levels, Electromyogram, Overlap antibody testing and muscle biopsy should be carried out to diagnose overlap myositis. Diagnosis of overlap myositis is important as treatment and prognosis differs when compared to pure myositis. However further studies are required to determine the exact treatment of overlap myositis.

REFERENCES

1. Loannou Y et al; Myositis overlap syndromes; Current opin Rheumatology 1999 November;11(6);468-74.
2. Jablonska S, Blaszcyk M et al ; Scleroderma overlap syndromes ; Adv Exp Med Biology 1999;45;85-92.
3. Bhansing et al. Arthritis Research & Therapy 2014, 16:R111
1. Scleroderma-polymyositis overlap syndrome versus idiopathic polymyositis and systemic sclerosis: a descriptive study on clinical features and myopathology
4. James B Lilleker et al The EuroMyositis registry: an international collaborative tool to facilitate myositis research; Annals of rheumatology 2017
5. E C Jury, D D'Cruz, W J W Morrow ; Autoantibodies and overlap syndromes in autoimmune rheumatic disease; Journal of clinical pathology; 2001 May ; 54 (5):340-7.
6. Pia Moinzadeh et all ; Disease progression in systemic sclerosis-overlap syndrome is significantly different from limited and diffuse cutaneous systemic sclerosis; Annals of Rheumatic diseases; volume74 , issue 4, Jan-2014.
7. Pari Basharat, Idiopathic inflammatory Myopathies: Association with Overlapmyositis and syndromes: Classification, clinical, characteristics and associated antibodies; European Medical Journal, Rheumatology2016:3(1); 128-135.
8. Dayal NA et al; SLE/myositis overlap: are the manifestations of SLE different in overlap disease? ; Lupus. 2002;11(5):293-8