International Journal of Advances in Medicine
Srujith CH et al. Int J Adv Med. 2019 Apr;6(2):236-239
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: http://dx.doi.org/10.18203/2349-3933.ijam20190543

Original Research Article

A study on demographic pattern of idiopathic inflammatory myopathies in a tertiary care hospital

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Received: 03 February 2019
Accepted: 13 February 2019

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ABSTRACT

Background: Idiopathic inflammatory myopathies (IIMs) are a group of chronic systemic autoimmune diseases characterized by proximal muscle weakness and elevated muscle enzymes. Aim and Objective was to analyze the demographic profile of patients with idiopathic inflammatory myopathies (IIM).
Methods: This was a cross sectional observational study conducted over a period of two years (2016-2018). After obtaining institutional ethical committee clearance, informed consent from patients. 16 patients who fulfilled the criteria were included in the study. The demographic and the clinical data were analysed.
Results: The mean age was 47.3±11.2 years. The study showed female predominance. ANA was positive in 11(68.7%) patients. Among the 16 patients, 5 (31.25%) had polymyositis and 11 (68.7%) had dermatomyositis. The median enzymes levels were creatinine kinase 1134 U/L, lactic dehydrogenase 477U/L, ALT (alanine aminotransferase) 154 IU/L, AST (aspartate aminotransferase) 236IU/L. Raynaud's phenomenon was seen in 37.5%. In our study, 31.25% had hypothyroidism and 6.25% had diabetic mellitus. On follow up 37.5% developed interstitial lung disease (ILD) and 18.75% were found to have malignancy.
Conclusions: Steroids and immunomodulators are the mainstay of treatment in patients with idiopathic inflammatory myositis. All our patients improved with steroids. It is important to evaluate these patients during early stages and follow up to prevent complications.

Keywords: Dermatomyositis, Inclusion body myositis, IIM, Myositis, Proximal weakness, Polymyositis

INTRODUCTION

Idiopathic inflammatory myopathies (IIMs) are a group of chronic systemic autoimmune diseases characterized by proximal muscle weakness and elevated muscle enzymes.[1]

Autoimmune inflammatory myopathies include dermatomyositis (DM), polymyositis (PM), myositis associated with anti-synthetase syndrome (ASS), immune-mediated necrotizing myopathy (IMNM), and inclusion body myositis (IBM). In 1975, Bohan and Peter proposed the diagnostic criteria for DM and PM using clinical, pathological and laboratory features which remain to be the gold standard in clinical practice.[2-4]

Both adaptive and innate immune pathways are implicated in the development of IIM. Although there are many similarities, PM and DM demonstrate distinct immuno histopathological phenotypes, suggesting that the underlying pathogenesis may not be the same. The immune-mediated necrotizing myopathies have very little inflammatory infiltrate on biopsy, necrosis is the predominant finding. Muscle biopsy of dermatomyositis

shows perifascicular and interfascicular inflammation. Polymyositis is associated with endomysial depositions of inflammatory cells

Clinical features include proximal muscle weakness, arthritis, dysphagia, dysphonia and Raynaud's phenomenon. Dermatomyositis is associated with skin manifestations like heliotrope rash, Gottron's papule, shawl sign, V sign and holsters sign. Dermatomyositis is commonly associated with malignancy compared to polymyositis. Common malignancies associated are nasopharyngeal carcinoma, ovarian, breast and gastrointestinal malignancy.

The main aim of treatment is to suppress the inflammation, improve muscle power and prevent chronic damage to muscles and extra muscular organs. Oral steroids and disease modifying agents like methotrexate, azathioprine and cyclophosphamide are the most commonly used drugs. This study aims to report a systematic review of case series in order to characterize the demographic profile, clinical data in patients with IIMs.

METHODS

The study was done after getting institutional ethical committee (IEC) approval and informed consent from the patients. It is a cross sectional observational and descriptive study. 16 patients who were attending the medicine and rheumatology department in a tertiary care hospital were selected and followed up for a period of 2 years (2016-2018). The above patients fulfilled the Bohan and peter diagnostic criteria for IIM which includes a) Symmetric proximal muscle weakness determined by physical examination, b) Elevation of serum skeletal muscle enzymes, including creatine kinase, aldolase, serum glutamate oxaloacetate and pyruvate transaminases, lactate dehydrogenase, c) The electromyographic triad of short, small, polyphasic motor unit potentials; fibrillations, positive sharp waves, and insertional irritability; and bizarre, high-frequency repetitive discharges, d) Muscle biopsy abnormalities of degeneration, regeneration, necrosis, phagocytosis, and an interstitial mononuclear infiltrate and e) Typical skin rash of DM, including a heliotrope rash and Gottron's sign/papules.

The exclusion criteria were patients who were previously diagnosed with myopthies (steroid myopathy), overlap syndrome, malignancy, or underwent post-transplantation/post radiotherapy immuno-suppression, or with HIV/AIDS. The patients were assessed and evaluated with basic hematological, biochemistry and immunological workup. They underwent creatine phosphokinase (CPK), lactate dehydrogenase (LDH), Ultrasound abdomen, high-resolution computed tomography (HRCT) chest, magnetic resonance imaging (MRI) and muscle biopsy. The results were analysed by using SPSS 16 software.

RESULTS

Our study had 16 patients who fulfilled the criteria with IIM. The study showed female predominance. 87.5% were females and 12.5% were males (Figure 1). The mean age of the study population was 47.3±11.2 years. In the study population majority of patients were between 40-60 years (62.5%). 18.75% of patients were more than >60 years and 6.25% of patients were less than <30 years (Figure 2).

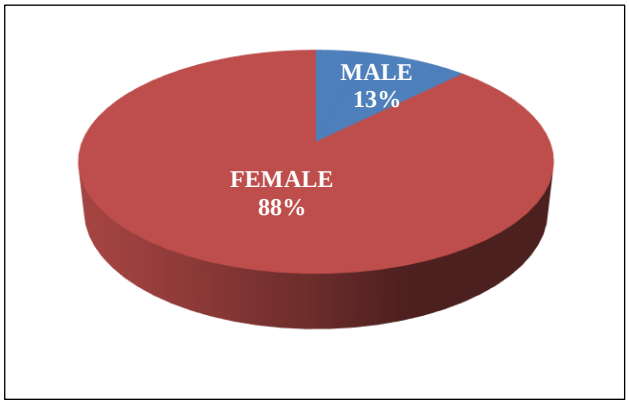

Figure 1: Sex distribution.

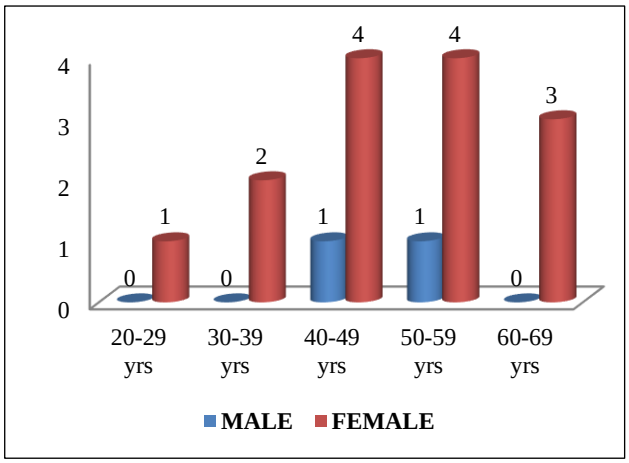

Figure 2: Age distribution.

Among the group 11 (68.7%) cases had of dermatomyositis, 5 (31.25%) had polymyositis. Patients diagnosed with dermatomyositis showed heliotrope rash with associated periorbital edema, Gottron's papules and shawl sign. On physical examination proximal muscle weakness was noted in all patients, while distal muscle weakness and respiratory muscle weakness was noticed in 12.5% respectively. Other presenting complains included dysphagia (6.25%), dysphonia (6.25%), and arthritis (68%) (Figure 3).

Around 68.7% of our patients were ANA positive, nine patients showed speckled pattern, one patient had cytoplasmic and other patient had nucleolar pattern (Figure 4). The Anti bodies associated were Mi 2 in 4

patients, U1RNP in 3 patients, Ku in 1 patient, SSA in 1 patient and PM Scl in one patient.

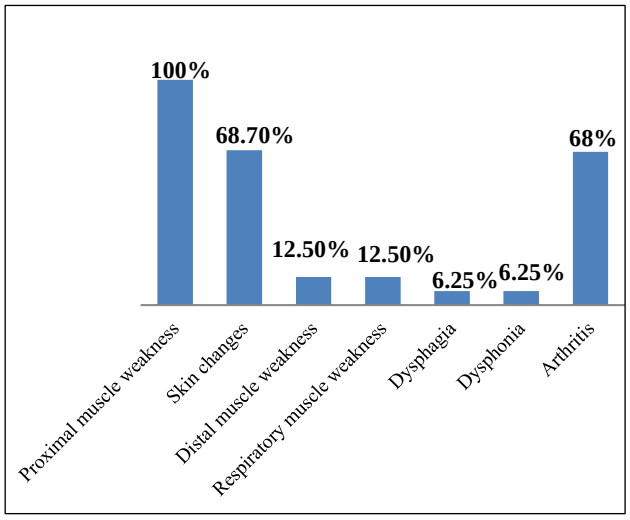

Figure 3: Clinical features.

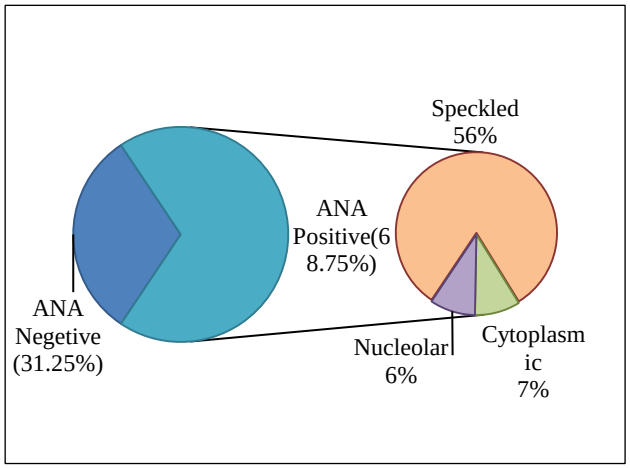

Figure 4: Results of ANA in IIM.

ESR was markedly raised mean 75mm/hr (26-150mm/hr). Muscle and liver enzymes were raised in all patients. Creatinine kinase median was 1134 IU/L, lactic dehydrogenase median was 477 U/L, ALT (alanine aminotransferase) was 154 IU/L (19-233 IU/L), AST (aspartate aminotransferase) was 236IU/L (25-725 IU/L). Some of the patients also presented with co-morbidities, 5 out of 16 patients had hypothyroidism (31.25%) and 1 patient had diabetic mellitus (6.25%).

Six (6) patients developed interstitial lung disease (ILD) (37.5%) and 3 patients were diagnosed with malignancy (18.75%) on follow up (Figure 5). The malignancies were ovarian cancer, breast carcinoma and papillary thyroid carcinoma.

All patients started on oral prednisone 1mg/kg were treated with disease modifying drugs. 50% patients on methotrexate (mean 17.5mg/week) ,18.75% patients were on azathioprine (2.5mg/kg), cyclophosphamide (20mg/kg for 6months) and 12.5% patients treated with rituximab 1000mg IV infusion 0, 14days. All patients were started on steroids for remission control.

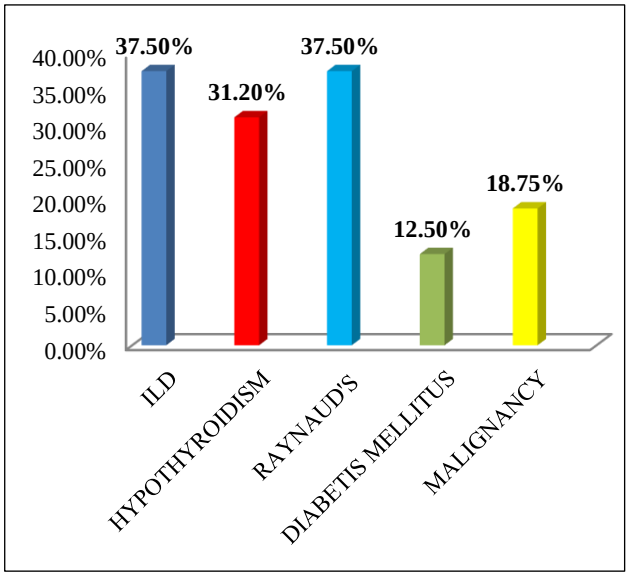

Figure 5: Associated conditions with IIM.

DISCUSSION

In our study, out of 16 patients 5 were diagnosed to have polymyositis and 11 had dermatomyositis. 87.5 % were female patients, implying a female preponderance (M:F = 1:7). The mean age of onset in years was 47.3. These findings similar to the study done by Ramesh KN et al, who showed that the mean age at presentation was 36.5 years (range, 16-68 years) with 13% of the patients being older than 50 years.[5]

The classical presenting symptom in all the patients was proximal muscle weakness, while distal muscle weakness was seen in 12.5% along with respiratory muscle weakness in 12.5%. Other presenting complains included dysphagia was noted in 6.25% and dysphonia in 6.25%. Arthritis was a relatively common symptom noticed, about 68 % presented with this feature. this correlates with the studies done by Malik et al, and Muggi N et al.[6,7]

Raynaud's phenomenon was seen to affect 37.5% of patients, it was predominantly noted in DM than PM, this data concurs with study done by Clark et al.[8] Other significant parameters noted in our study were elevated muscle enzymes in 85.1% patients, positive electromyogram in 60%.[9] Hypothyroidism noted in 37.5% of patients by Gamsky et al.[9] In this study describe the coexistent autoimmune thyroiditis and dermatomyositis, showing clearly that these diseases can coexist.

Interstitial lung disease (ILD) is found in more than a third of patients (37.5%), the majority of cases of ILD

occurred in patients between the ages of 40 to 60 years. The study done by Douglas WW et al also showed 18.6% patients with ILD.[10] Anti-nuclear antibody was positive in 68.75%. 68.7% of our patients were ANA positive. In our study group, nine patients showed speckled pattern, one patient cytoplasmic and one patient nucleolar pattern. Anti-bodies associated are Mi 2 in 4 patients, U1RNP in 3 patients, Ku in 1 patient, SSA in 1 patient and PM Scl in one patient. All patients were started on oral prednisone 1mg/kg and treated with disease modifying drugs. 50% patients on methotrexate (mean 17.5mg/week), 18.75% patients were on azathioprine (2.5mg/kg), cyclophosphamide (20mg/kg for 6months) and 12.5% patients treated with rituximab 1000mg IV infusion 0, 14days. All patients were subjected to periodic screening and follow up for the period of 2years. During follow up 3 developed malignancies (18.75%). The malignancies were ovarian cancer, breast carcinoma and papillary thyroid carcinoma. A study done by Tembe AG et al showed that the most common malignancy was that of the breast.[11]

When PM and DM where compared, it was seen in the study that patients with DM were at more risk for malignancy. Death occurs due to the malignancy rather than the myositis, which responds to treatment. Buchbinder et al have reported that 74% of malignancies detected during initial screening or during the follow up after the diagnosis of myositis.[12] The overall risk of cancer is greatest in the first 3 years after the diagnosis of myositis. Multiple studies done in south Asia showed a positive correlation with IIM and the increased risk of malignancy associated with it.[13-15]

CONCLUSION

Steroids and immunomodulators are the mainstay of treatment in patients with idiopathic inflammatory myositis. It is important to evaluate these patients during early stages and follow up for associated conditions like interstitial lung disease, hypothyroidism, Raynaud's and malignancy as seen in our study.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Srujith CH, Mohanasundaram K, Jagadeesan M, Halleys Kumar E, Kannan R, Sivasubramanian K, et al. A study on demographic pattern of idiopathic inflammatory myopathies in a tertiary care hospital. Int J Adv Med 2019;6:236-9.