Research article

Describing inflammatory muscle disease in Kenya: A single tertiary centre experience in Kenya

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Abstract

Background: Inflammatory muscle diseases are a rare group of connective tissue diseases. There is a paucity of documented literature on indigenous Africans in sub-Saharan Africa. We present herein the clinical patterns of inflammatory muscle diseases encountered at a rheumatology clinic, Nairobi, Kenya.

Objective: To describe the clinical spectrum of inflammatory myopathies at a tertiary rheumatology clinic in Nairobi. These included clinical, haematological and immunological characteristics of patients with Inflammatory myopathies.

Methods: Medical records of 10,998 patients presenting to the Nairobi Arthritis Clinic for various rheumatological conditions were reviewed. The records of 46 patients with muscle weakness with or without skin rash were selected and reviewed between January 2012 and December 2017 were retrospectively reviewed and reclassified as polymyositis (PM) and dermatomyositis (DM) based on the Bohan and Peter diagnostic criteria.

Results: Forty-six patients (F=36, M=9) were diagnosed with polymyositis and dermatomyositis. Twenty-five had possible dermatomyositis, eighteen had possible polymyositis with another three who had an overlap of polymyositis with other diseases. There were 3 patients with juvenile dermatomyositis. Majority of the patients were referred of which 14 had an alternative diagnosis to myositis. The mean age for PM was 36.36 years and for DM 41.13 years. The creatinine kinase mean was 2845.4 (697-7063)u/l. Serology for ANA tested positive in 8 patients (PM=4, DM=4). The most common symptoms of DM patients included Gottron papules (12), heliotropes rash (15) and shawl sign (5). Myositis antibody screening was not performed in any of the patients.

Conclusion: Inflammatory myopathies are still rare in Kenya. The clinical spectrum is largely similar to what is known in written literature. From referral notes and diagnosis of the primary physician, there is a paucity of information about these diseases. None of the patients had myositis antibody panel due to either unavailability or high cost of doing the tests. More effort should be on increasing awareness of diagnosis and management of these diseases.

Key words: Inflammatory muscle disease, Polymyositis, Dermatomyositis, Nairobi, Kenya

Introduction

Inflammatory myopathy is a larger term used to describe muscle diseases that represent dermatomyositis (DM) and polymyositis (PM). These group of diseases share features of muscle inflammation and proximal muscle weakness. The differentiating feature is that DM has characteristic skin manifestations[1]. The clinical and serological profile of DM and PM may vary from populations through the immune pathology within the muscle tissue is constant and distinct[2]. There are a wide variety of diagnostic criteria used with criteria of Bohan and Peter the most commonly used [3,4]. These criteria have a number of limitations with its inability to distinguish other forms of myopathy thus can misclassify IBM patients as PM. These limitations together with recent advances such as myositis-specific autoantibodies, that are associated with distinct clinical phenotypes has led to the development of new criteria for the diagnosis and treatment of myopathies by the EULAR/ACR groups[5]. As the disease is thought to be rare in Kenya and Africa, it is poorly understood as compared to other connective tissue diseases thus diagnosis can be a challenge. This is compounded by the limited number of rheumatologists and the high costs involved in making the diagnosis and treatment. This leads to incorrect diagnosis and delays in starting the proper treatment resulting in increased morbidity and mortality. There are few reports on inflammatory myopathies in sub-Saharan Africa. In this study, we describe the spectrum of inflammatory myopathies based on Bohan and Peter's criteria seen at a rheumatology clinic in Nairobi.

Materials and methods

This was a retrospective study carried out in the Nairobi Arthritis Clinic. The study site is situated in Nairobi, the capital city of Kenya and serves as a tertiary referral center. It not only serves the two million inhabitants of Nairobi but also patients from all over Kenya and the greater East and Central African region. Following ethical approval, we reviewed the case records of 10,998 patients attending the Nairobi Arthritis Clinic and those with a diagnosis of Inflammatory Myopathy (IM) based on Bohan and Peter's criteria attending the Nairobi Arthritis Clinic between January 2012 and December 2017 and had been on follow up for at least 6 months were recruited into the study. Clinical, haematological, immunological and other relevant findings from the history were obtained from the available records. Patients were classified into two as either polymyositis or dermatomyositis. Patients with conditions that may mimic IM such as endocrine disorders, adverse effects of medication, metabolic myopathies and muscular dystrophies were excluded from the study.

Data were collected from medical records using a questionnaire including demographic data, the subtype of the myopathy, referring diagnosis, gender, age at diagnosis, association with malignancy or connective tissue diseases, clinical features recorded at the time of the diagnosis and during follow up, and other systemic involvement and the pharmacological agent used. Laboratory tests comprised biochemical tests creatine phosphokinase levels (reference levels of 22 to 198 U/L), positivity of Anti-Nuclear Antibodies (ANA) and myositis antibody panel at diagnosis were documented. For each patient, the electromyography (EMG) and muscle biopsy results were recorded at the time diagnosis was made. Using Bohan's and Peter criteria for muscle biopsy, they were recorded as positive if the results had evidence of necrosis of myofibers, phagocytosis, regeneration with basophils, large vesicular sarcolemmal nuclei, and prominent nucleoli, atrophy in a perifascicular distribution, variation in fiber size and an inflammatory exudate, often perivascular was recorded as positive as per Bohan's and Peter criteria. Review of anti-rheumatic pharmacologic treatments used during the study period included corticosteroids (intra-articular/ systemic), methotrexate (MTX), azathioprine (AZA), mycophenolate (MMF) and biologic agents was done.

Results

A total of 46/10,998 patients were identified, 21 patients had PM, 21 had DM, 4 had JDM and 4 had associated diseases (systemic lupus erythematosus, HIV and mixed connective tissue disease). There were 37 females and 9 males giving a female to male ratio of 4.1:1. The ages ranged from 3 to 60 years with a mean age of 37.45 years as shown in Table 1. There were three cases of juvenile dermatomyositis. The male to female ratio was 1:2. The most common clinical presentation was proximal myopathy (100%) of patients followed by arthritis (78.2%), dysphagia (65.2%), Gottron's papules (26.1%), heliotrope's sign (21.7%) and V-shawl sign (10.8%). Two patients had interstitial lung disease. One of the patients had a malignancy as shown in Figure 1. She was 42 years diagnosed with DM who on further evaluation was found to have ductal cell carcinoma. There were two patients with HIV with polymyositis. One was a 32-year-old male on zidovudine/lamivudine/efavirenz combination and the second a 48-year-old female on atazanavir/ ritonavir/ raltegravir combination. A total of 24 out of 34 patients were referred by clinicians with alternative rheumatological diagnoses other than myositis. The referrals included rheumatoid arthritis (11), connective tissue disease (5), fibromyalgia (4), scleroderma (2) cervical spondylosis (2) and myositis (10). The EMG findings were consistent with PM in 5 patients and 4 were positive for DM. The muscle biopsy results were consistent with inflammatory myopathies in 6 patients. Important to note is that EMG and muscle biopsy was not done on all patients. Myositis antibody panel was not done on any of the patients in this series mainly due to financial constraints and some had been referred already on steroid therapy.

All the patients received steroids with normalization of CK in 43.47% and muscle power in 78.7%. Other treatments included mycophenolate mofetil 11(23.9%) patients, methotrexate and hydroxychloroquine combination 8 (17.34%) patients, two patients on

Table 1: Demographic data of inflammatory myopathy from select studies in Africa

	Genga EK	Adelowo OO	Diallo M	Toumi S	Khelifa E	Mebazaa A
PM	21	7	6	20	0	0
DM	25	7	15	50	13	130
M: F	1:4.11	1:13	0:0.6	1:2.5		3:3
Mean age (years)	37.45	35		40.7	32.85	49.5
Mean CPK(u/l)	2845.4	1134				
Malignancy	1		3	12.8	2	20

azathioprine (4.3%) and one patient on rituximab (2.1%). There were 9 patients lost to follow up during this period.

Figure 1: Symptoms of patients with DM

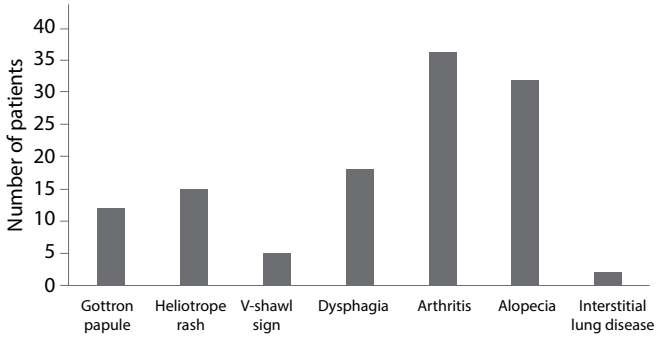
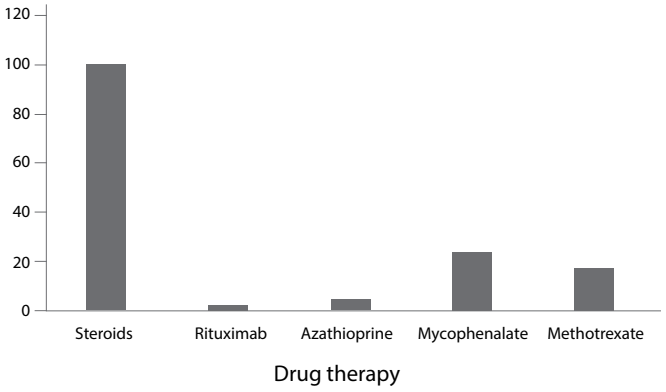


Figure 2: Steroid-sparing drugs used



Discussion

The study covered a period of 7 years and yielded 46 cases of inflammatory myopathies. This is the largest case series of inflammatory myopathies in East and Central Africa reporting on clinical and laboratory features as well as any association with malignancy and treatment modalities. This study was predominantly female with a male to female ratio of 1:4.1 with a mean age of diagnosis of 37.45 years (range from 3-60 years). This is similar to studies around the continent from Nigeria to Tunisia[6-9]. The mean age of onset is similar to other studies across the African continent[6-9]. There is a Tunisian study that differs with the above as the gender is equally balanced and had an older age of onset[10]. However, this looked exclusively at malignancy and dermatomyositis and may explain the difference. Our analysis shows a predominance of dermatomyositis which is in contrast to what is known in American and European literature[11-14]. There were low numbers of JDM but the data largely mirrored a South African study by Faller *et al*[15]. Generally, our results as compared to previously reported studies show clinical feature similarities. The most common clinical presentation was proximal myopathy (100%) of patients followed by arthritis (78.2%). This is common to what is known in literature both from Africa and around the world[8,16-18]. Involvement of other organs with myopathy is well known. The number of patients with extra-skeletal manifestations was low, take for example two in this case series had interstitial lung disease. This highlights the challenges of managing these rheumatic diseases in a poor resource set up where funds to do a comprehensive approach to these patients is limited. This is highlighted by the low numbers of muscle biopsies, EMG and myositis antibody tests. More effort should be done by the Government and the private sector to make these tests readily accessible by improving healthcare infrastructure and lowering costs to do these tests.

There has been an association established between inflammatory myopathies and malignancy[8,17]. A Tunisian study reported up to 90% of their cohort had myositis as a paraneoplastic symptom[8]. The most common malignancies in this cohort were breast cancer (35%) followed by nasopharynx (25%). They also reported having malignancy with a poor prognostic marker in dermatomyositis. The results of this study differ with what is known in literature where the order of malignancies is ovarian cancer, lung cancer, and pancreatic cancer[19,20]. Asian studies report a predominance of nasopharyngeal malignancies[20]. Our data had one malignancy. She was 42 years diagnosed with DM who on further evaluation was found to have ductal cell carcinoma. Studies have reported the association between HIV/AIDS and myopathic disease with some reporting prevalence as high as 25% of infections[22]. Asymptomatic elevations in CK, myalgias, and rhabdomyolysis are all possible complications of HIV. Contribution from drugs used to treat HIV for example zidovudine has been established. Studies have estimated zidovudine-induced myopathy between 8-17%[22]. Our study had two cases with HIV with one of the patients on the zidovudine-based regiment. A large number of our patients responded to corticosteroids which is in keeping with what is known in the literature[18].

Limitations and recommendations

Our study was limited by the small sample size and its retrospective design. This may have been due to the low numbers of inflammatory muscle disease generally. Due to financial constraints most patients were unable to do muscle biopsies and myositis antibodies. This may have made us miss out on further subclassifying our patients. A recommendation would be for longer follow up looking into prognostic and survival of these patients. As per the referral diagnosis sent by our colleagues, another recommendation would be to improve knowledge of myositis in Kenya and also making diagnostic tests more readily available and affordable. This will help improve the holistic approach including diagnosis, malignancy screening, and management of these patients.

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