ADDIS ABABA UNIVERSITY

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DEPARTMENT OF DERMATOVENEREOLOGY

Magnitude of dermatomyositis among dermatology patients at ALERT Hospital Addis Ababa, Ethiopia: A five year retrospective study

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Abbreviations/ Acronyms

AAU- Addis Ababa University

ALERT- All African Leprosy Rehabilitation Training Center

CADM- Clinically amyopathic dermatomyositis

CDM- Classical dermatomyositis

DM- Dermatomyositis

G.C-Gregorian calendar

EMG- Electromyography

IIM- Idiopathic inflammatory myopathies

JDM- Juvenile dermatomyositis

SPSS - Statistical Package for Social Science

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Abstract

Background

Dermatomyositis is an inflammatory multisystemic disease characterized by muscle weakness and a characteristic and pathognomonic cutaneous eruption. It also has increased risks of interstitial lung disease (ILD) and malignancies and a greatly increased risk of mortality compared to the general population.

Dermatomyositis is a rare connective tissue disease. Data regarding the incidence and prevalence of DM are scarce in our country.

Objective

The objectives of this study were to identify new and existing cases of dermatomyositis at ALERT Hospital from January 2017 through December 2022 G.C and to establish a hospital-based estimate of the magnitude of dermatomyositis.

To analyze the average age, sex distribution, and skin manifestation.

Method

A hospital-based retrospective cross-sectional study of dermatomyositis patients seen at ALERT Center from January 2017 to December 2022 G.C. was conducted.

A structured data extraction sheet was used to collect the data from medical chart records.

Data was analyzed using Statistical Package for the Social Sciences (SPSS) version 26, and frequency distributions, percentages, tables, and charts used to show results.

Result

27 dermatomyositis patients were found; of these, 25 (92.5%) were female and 2 were male. The mean age was 33.07 years. 22 patient charts evaluated for laboratory and biopsy results from these classical adult-type dermatomyositis was the commonest type of DM, which accounts for 68.2% (15), whereas clinically amyopathic DM 1 (4.5%), juvenile dermatomyositis 4 (18.2%), and 2 (9.1%) specific types were not mentioned. The most common clinical manifestations are symmetric erythema 21 (95.5%), helitrope sign 14 (63.6%), and skin hyperpigmentation on photodistributed areas 12 (54.5%).

Conclusion

The magnitude of dermatomyositis in this study is found to be low and predominantly classical adult type. The low finding could be attributed to patients with predominant muscle weakness visit rheumatology unit than dermatology department. Skin hyperpigmentation was a common finding in our study.

Background

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous findings that can affect both adults and children. This systemic disease mainly affects the skin and muscles, but it can also affect the joints, oesophagus, lungs, and, less frequently, the heart. (1,2).

DM is grouped under idopathic inflammatory myopathies along with polymyositis and inclusion body mysositis. It is an autoimmune connective tissue disease that presents with proximal extensor myopathy and typical skin lesions. Current available evidence points out that underlying pathogenesis involves a humoral immune process in which components are deposited in capillaries, leading to ischemia(3).

Dermatomyositis can affect any age group with a characteristic bimodal age distribution: adult and juvenile. Most cases of dermatomyositis in pediatrics present between the ages of 5 and 14 years of age, and in adults in the 5th and 6th decades of life(4).

A recent journal article by Bolender et al. (2022) indicated that the prevalence and incidence of DM are greater than previously reported, and associated mortality is increased for those with muscle involvement but not for clinically amyopathic DM (5).

A paper published by J. A. Tan et al. (2013) demonstrated that females are more affected by DM than males. They found an F:M ratio of 2.75 DM cases in their 30-year retrospective study (6).

The prevalence of dermatomyositis ranges from 8.7 to 28 per 100,000 amongst various populations. Dobloug C. et al. (2015) estimated the prevalence at 8.7/100,000 of the general population based on their 9-year tertiary hospital-based cohort study. A population-based 30-year retrospective cohort study done in 2019 estimated the prevalence of DM at 13 per 100,000 (3). Another population-based cohort study done in Canada estimated the prevalence at 28.6 cases per 100,000 adults. Rosa et al. (2013) found an incidence of DM of 10.22 per 100,000 people (3,7,8). Prevalence of DM also seems to be increasing among various studies done over time (9–11).

A recent study to estimate the epidemiology of IIMs in Africa reviewed 39 papers, which included 683 cases of IIM, and estimated the incidence rates of DM at 1.2 per 1000 000 person-years. The sex ratio varies among studies in which 50–100% of patients are female.(9).

Skin involvement in DM usually manifests with characteristic Gottron papules, the Gottron sign, sysmmetric erythema on photoexpsoed areas, a heliotrope sign, periungual

telangiectasias, poikiloderma, and dystrophic cuticles(12,13).

Although in DM proximal muscle weakness affects the majority of cases, amyopathic variants with no muscle involvement have been described in multiple literatures (14–16). The relationship between DM and internal malignancy, especially in the older age group, is described. Cancer risk was greatest in patients with DM who were > 50 years of age at the time of diagnosis (17–20).

Given that dermatomyositis (DM) is a multisystem disease, it also affects lung diseases and cardiac(1).

Objectives

General objective

To evaluate the Magnitude of dermatomyositis in ALERT hospital, Addis Ababa, Ethiopia, among patients seen at ALERT hospital, dermatology department, between 2017 and December 2022 G.C.

Specific objectives

- To identify the magnitude of dermatomyositis among dermatology patients seen over five years period
- To assess the associated demographic characteristics such as: age and sex distribution.
- To assess clinical features.

Statement of the problem

Dermatomyositis is a relatively rare autoimmune connective tissue disease that occurs throughout the world. Although the epidemiology of dermatomyositis has been extensively studied in America, Europe, and Asia, data regarding dermatomyositis in Africa is scarce. In particular, there is no available data regarding the epidemiology of dermatomyositis in our country.

To address this gap, it would be of help to carry out a study in our country and compare it with the available literature.

Significance of the study

This study helps assess the magnitude of dermatomyositis in our hospital by assessing the associated demographic characteristics such as age and sex distribution.

It compares the patterns seen in Ethiopia with those of other African literature and the patterns in other parts of the world looking for similarities and differences.

As there has been no study done in Ethiopia so far on dermatomyositis, this study will give an insight into how common this is in our country.

This will be helpful for dermatologists and other specialties to be informed about the burden of this disease in our country and this diagnosis.

The present study can also encourage other researchers to carry out further studies in the field utilizing it as a base.

Methods and Materials

Study area

The study was conducted at All African Leprosy Rehabilitation Training Center (ALERT). ALERT center is located in an area locally called Zenebework, in the Kolfe Keraniyo sub-city of Addis Ababa.

It was first established as a treatment center for leprosy, and it focuses on the rehabilitation of leprosy patients, training programs for leprosy personnel from around the world, and leprosy control. The hospital is the main dermatologic center in the country and functions as the referral dermatology institute in and around Addis Ababa.

Study Period

The study was done from June 2023 to September 2023 G.C.

Source population

All patients seen at ALERT hospital dermatology department.

Study population

All patients with clinical and histopathology diagnosis of dermatomyositis in ALERT hospital, dermatology department, from January 2017 and December 2022 G.C.

Study design

A five-year hospital based retrospective cross-sectional study was conducted to assess the magnitude of dermatomyositis during the period of January 2017 and December 2022 G.C in ALERT Hospital.

Eligibility criteria

Inclusion Criteria

- All patients with clinical and/or histopathology diagnosis of dermatomyositis cases seen during the mentioned period.

Exclusion criteria

- Incomplete charts

Study Variables

Dependent Variable

- Dermatomyositis

Independent Variables

- Sex
- Age
- Distribution

Sample size

All patient with a diagnosis of dermatomyositis seen at ALERT Center during January 2017 and December 2022 was included.

Operational definitions

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by classical skin rash (symmetric erythema involving the face, neck, upper chest, lateral parts of extremities, skin overlying bony prominences and joint, and pokilodermatous changes) with or without muscle weakness. Patients with pathognomonic skin rashes (heliotrope rash, Gottron's papules, and/or Gottron's sign) or symmetric muscle weakness with skin rash consistent with DM are considered to have probable DM in this study.

Data collection tools and procedures

After obtaining ethical clearance, data was collected from medical records of patients visiting ALERT Center by using a structured data extraction sheet.

Data processing and analysis

Data entry, coding, and cleaning were performed using Epi-Info version 7.0, and statistical analysis will be done using SPSS version 26.

Frequency distributions, percentages, tables and charts were used to show descriptive results. Finally, the study finding will be presented using diagrams, tables, and figures**.**

Data quality management

Trained data collectors were involved in data collection.

The data was checked for completeness, clarity, and consistency after being filled out each day.

Ethical considerations

Ethical clearance was obtained from the Institutional Review Board (IRB) of Addis Ababa University before starting the research.

Permission to review patient charts and ethical clearance was also obtained from the AHRI/AHRI Ethical Review Committee. Any revealing information about the patient's identity was not collected. The data collected was not disclosed and will remain confidential as it will only be passed between the investigators listed on this protocol.

Data dissemination and utilization

The finding of the study will be submitted to AAU, Department of Dermatovenereology. It will also be submitted to scientific journals for possible publication.

Research Result

Socio-demographic data

Out of a total of 51568 patients seen at the ALERT Centre dermatology department during the five-year period (from June 2017 to June 2022 G.C.), 27 had dermatomyositis, making the magnitude of DM 0.05%.

Euwer and Sontheimer's diagnostic criteria for ADM and Bohran's criteria for CDM were used (8–10). Sontheimers and et al. (1991) proposed that a diagnosis of ADM can be made if a patient has 1 or 2 pathognomonic cutaneous DM features Plus 1 or more classic skin signs and a compatible skin biopsy result. On the 2017 EULAR/ACR Classification criteria for IIM and their major subgroups, authors recommended patients with pathognomonic skin rashes (heliotrope rash, Gottron's papules, and/or Gottron's sign) be classified as DM patients (8). The latter criteria is also used for further classifications of DM as adult and juvenile based on age, classical adult and amyopathic DM, and JDM based on the presence and absence of clinical muscle weakness.

Bohan and Peter's diagnostic criteria were proposed in 1975, in which a possible DM is diagnosed when the patient has a skin rash plus one muscle criteria.

27 patients with a diagnosis of dermatomyositis seen in past years were identified, and from these 5 were excluded because they were uncharted, all 22 patients' charts were thoroughly studied for clinical history, physical examination findings, and laboratory studies. All patients (27) were evaluated for sex and age proportion but only 22 were included in other parts of the study questionnaires.

From 22 DM patients found classical adult-type dermatomyositis was the commonest type of DM which accounts for 68.2%(15), whereas clinically amyopathic DM 1(4.5%), juvenile dermatomyositis 4(18.2%), and 2(9.1%) specific type not mentioned.

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In this study patients' age ranged from 10 to 58 years and mean age was 33.07 years. Majority of patients were in their 3rd and 4th decades of life with 59.2% (16 out of the 27) frequencies. There were 4/27 cases who presented under the age of 18 years. The average age was 36.3 and 14.25 years for the adult and juvenile groups, respectively.

Table 1 Age distribution

Age group	Frequency	Percent
<18	4	14.8
18-29	8	29.6
30-39	9	33.3
40-49	2	7.4
>50	4	14.8
Total	27	100

Out of the 27 dermatomyositis cases identified in the five-year period, 25 (92.6%) were female and 7.4% (2) were male, giving a female-to-male ratio of 12.5:1. The mean age for females was 34. Males aged 19 and 24

13 (86.6%) of adult classical DM, 1 amyopathic, and all juvenile DM were female patients. 2 adult CDM were male.

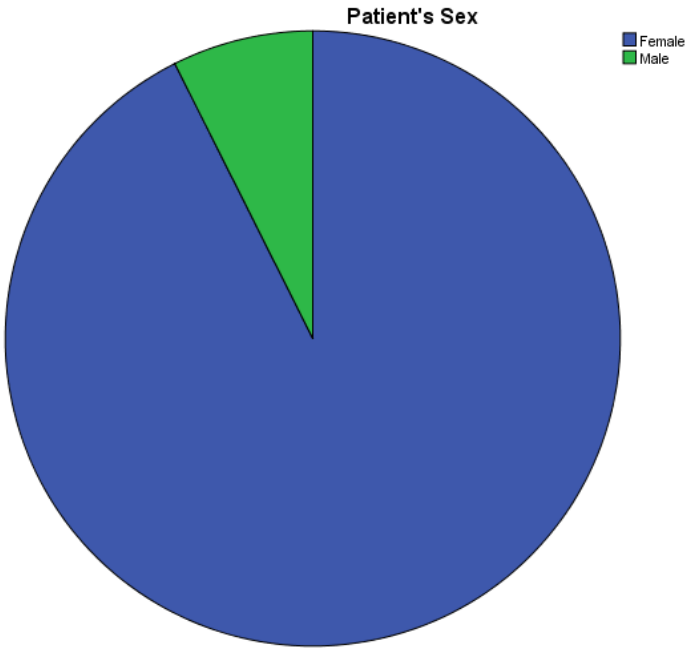

Figure 1 Sex distribution

Clinical Presentation

Duration of the disease at presentation ranged from 6 weeks to 8 years. Most cases presented with in 2 years of disease 20(90.9%), among them 10 (45.4%) presented within 6 months and

10(45.4%) within 6 months to 2 years. Only 2(9.1%) patients presented after 2 years of disease onset.

Table 2 Duration of symptoms

Duration at presentation	Frequency	Percent
<6 month	10	45.45
6 month- 2 years	10	45.45
>2years	2	9.1

77.3% (17) of patients had both skin lesions and muscle weakness, while 22.7%(5) of them had only skin lesions.

Table 3 CLINICAL PRESENTATIONS

CLINICAL PRESENTATION	Frequency	Percentage
skin lesions only	5	22.7
Both skin lesions and muscle weakness	17	77.3
Total	22	100.0

Symmetric erythema was the commonest skin lesion which was found in 95.5% (21) of the patient, helitrope sign was found in 14 (63.6%), skin hyperpigmentation in 12 (54.5%), Gottron papules 9(40.9%), Poikilodermatous change in 11 (50%) and telangiectasia in 7 (31.8%) were found.

Table 4 Clinical findings on skin examination

Skin lesions	Frequency	Percent
Symmetric erythema	21	95.5
Helitrope sign	14	63.6
Poikidodermatous change	11	50
Cutaneous telangiectasia	7	31.8
Skin hyperpigmentation	12	54.5%
Calcinosis	2	9.09
Gottron papules	9	40.9
Gottron sign	6	27.3

Shawl sign	5	22.8

In our study, we also looked into laboratory studies, which include muscle enzymes and EMG, age-dependent cancer screening tests, and skin biopsy.

22 patients had at least one muscle enzyme test, which included CK, LDH, or ALT. In 13 patients (59.68%), they were elevated, whereas in 9 cases they were normal.
The EMG test was determined only in 3 cases; 2 of them had results consistent with DM.
7 (including all amyopathic's DM) patients had skin histology consistent with cutaneous dermatomyositis.

Regarding cancer screening, all 22 patients had CBC and LFT. 15 (93.7%) adult female DM patients had abdominal-pelvic ultrasound, and 5 (22.7%) had chest x-rays. We did not find patient with evidence of underlying malignancy with limited investigation conducted.

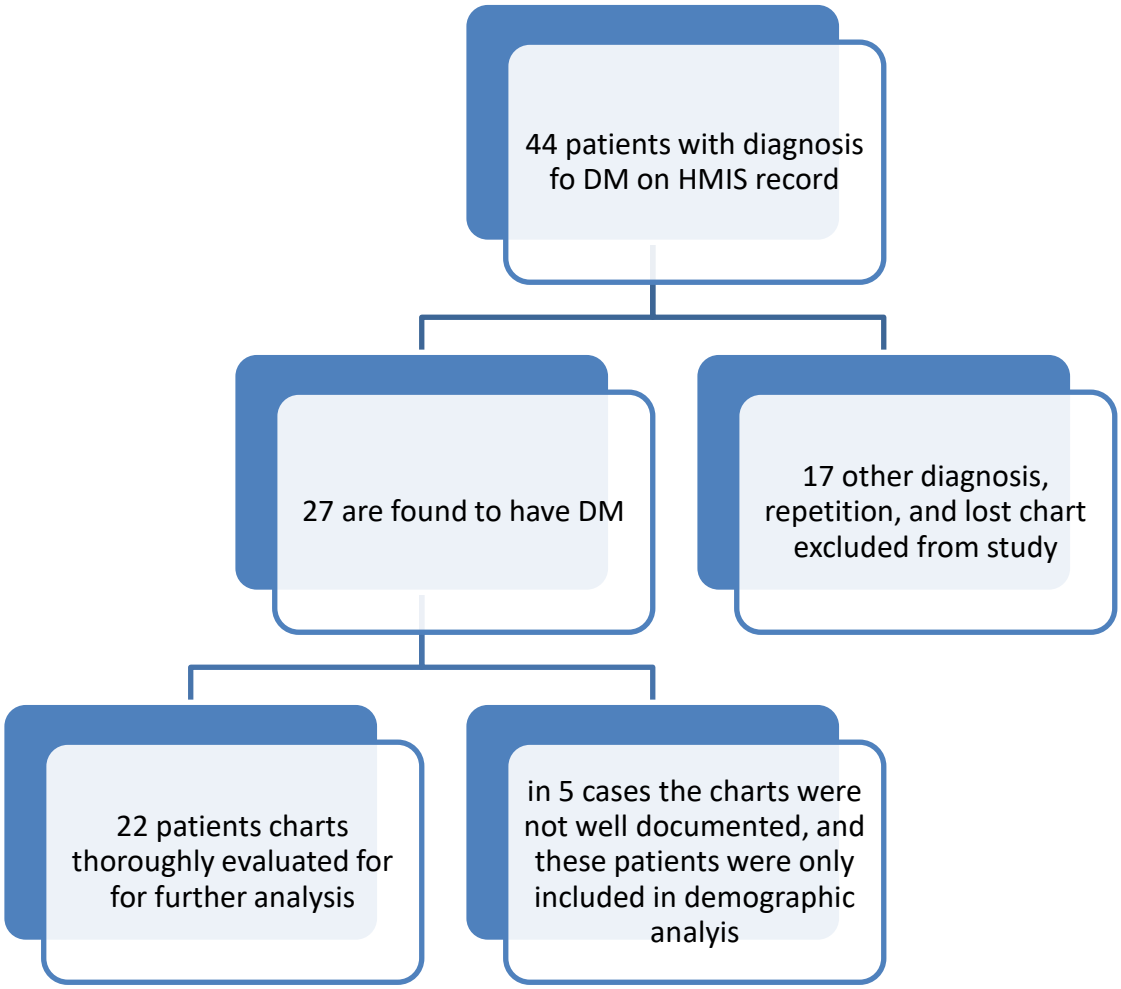

Figure 2 Flow chart

Discussion

Dermatomyositis (DM) is an idiopathic inflammatory myopathy (IIM). In addition to DM, IIM includes polymyositis (PM), inclusion body myositis (IBM), immune-mediated necrotizing myopathy (IMNM), and overlap myositis (OM). Dermatomyositis is the second-commonest IIM(21).

A systemic review by Alain Meyer et al. 2015 estimated overall IM incidence at 7.98/million/year and prevalence at 14.00 cases per 100,000, based on the results of 10 surveys in which prevalence ranged from 2.4 to 33.8 per 100,000.(11)

Our study is a five-year retrospective study that assesses the magnitude of Dermatomyositis at ALERT center.

In our study, dermatomyositis accounted for 0.05% of the total number of dermatology patients seen at ALERT Center during the five years.

In a prospective study conducted in 2014 by Aman S et al. to ascertain the pattern of skin diseases among patients attending a single tertiary care hospital, they found that 645 patients, or 0.90% of the patients, had connective tissue disorders, which included lichen sclerosis et atrophicus, systemic sclerosis, localised morphea, and systemic lupus erythaematosus, In another one-year prospective study done in single tertiary hospital in india dermatomyositis accounts for 0.008% dermatology visit (22,23)

In this study, the magnitude of DM was higher in females compared to males. This is similar to Mehmet et al, 2016, Bernatsky et al 2009, Osman et al,2023 and Keith J et al 2020 (8,24–26). Female:Male ratio is higher than He et al 2022, even though females are still more likely to affected, female: male ratio is lower which is 1.6 to 1(24).

Mean age at onset for adult-onset dermatomyositis is 36.3 years which is almost similar to similar retrospective study done on single tertiary hospital in the northern trace district of Turkey, Balci al,2016 (24). A retrospective study to determine patterns of IIMs in South Africa also showed similar findings(26). But studies from Europe and America suggest peak age of onset of DM is 40-60 years(3,27)

Among 22 patients charts evaluated for further information Symmetric erythema, Heliotrope sign, and skin hyperpigmentations are common findings found in 21(95.5%), 14(63.6%), and 12(54.5%) respectively.

Balci et al, 2016 In their study found a similar finding, among 23 patients 19(83%) had heliotrope signs and 20(90%) symmetric erythema(24).

Bendewald et al suggested association of DM with malignancy (28) but we did not find patients with evidence of malignancy in our study. This may be explained by the younger age of our patients compared to those studies, in mean age at onset was 57.4 years. Lauinger, J. et al. (2021) analyzed 63 DM patients retrospectively, and in their study, they found the mean age of patients with malignancy-associated DM was higher than that of those without malignancy (68.8 ± 11.6 years versus 52.4 ± 15.2 years), respectively. (27). Another possible explanation is cancer screening is not comprehensive enough.

In this study, we also tried to classify patients to specific DM, 15(55.6%) are typical adult type dermatomyositis, 1(3.7%) clinically amyopathic dermatomyositis, 4(14.8%) juvenile dermatomyositis, and in two patients specific type was not mentioned. This study is similar to the retrospective study done in Olmsted County, Minnesota, from 1976 through 2007 they found 29 DM with the following subclassifications: 23 patients with CDM (2 juvenile), 3 with ADM (1 juvenile), and 3 with CADM (28) . R.D. Sontheimer et al. 2002 also estimated prevalence of C-ADM to 10% among Dermatomyositis patients. But a 3-year retrospective study of 28 DM patients in a single dermatology center in Singapore found that 46% of their cases had C-ADM.

Conclusion

In this retrospective study to determine the magnitude of dermatomyositis seen at the ALERT Centre dermatology department over five years, we found 27 DM patients. which accounted for 0.05% of total visits, which suggests dermatomyositis is a rare entity. Our patients were found to be younger than what was suggested by much literature. The reason for this could be that this study includes both adult and juvenile DM. Female sex is majorly affected. Common skin findings were symmetric erythema, helitrope's sign, and skin hyperpigmentation, which are consistent with other studies except for skin hyperpigmentation.

Recommendations

We recommend to have a standard way of investigation and follow-up on underlying malignancies and muscle enzymes and improving the completeness of medical chart records for patients.

Future researchers may conduct a similar study in a different setting to discover new knowledge and add to the limited literature on dermatomyositis in our country. Lastly, heightened awareness is needed for the diagnosis of DM.

Limitations of study

Our study is a retrospective study and ALERT centre is a main dermatology service provider in the country we cannot generalise it to nationwide prevalence.

Since the method of documentation of charts by physicians varies and some patients' important clinical data were absent, in our study we found it difficult to include all probable cases of dermatomyositis.

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