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CHARACTERISTICS AND OUTCOME OF PATIENTS WITH DERMATOMYOSITIS IN CENTRAL SAUDI ARABIA, A RETROSPECTIVE EPIDEMIOLOGICAL STUDY

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Abstract:

Background: *Dermatomyositis (DM) is an autoimmune inflammatory condition that can cause inflammatory myositis along with characteristic skin manifestations.[1].It can be associated with significant morbidity including malignancies.*

Study objectives: *Our aim is to study the epidemiology, clinical features, investigations and treatments used in dermatomyositis patients at participating hospitals in central Saudi Arabia. We also aimed to study the prevalence of vitamin D deficiency among dermatomyositis patients.*

Methods: *We conducted our retrospective study at 3 hospitals in central Saudi Arabia using data extracted from electronic medical records (EMR). We included all dermatomyositis seen at follow up in participating hospitals (Prince Sultan Military Medical City, King Faisal Hospital and research center, King Fahad Hospital in Buraidah city) for the period between July 2017- July 2021. Only patients who met the diagnostic criteria of dermatomyositis based on the Bohan and Peter criteria were included. Data obtained from patient medical records included age, sex, investigations and treatment modalities. Statistical analysis was performed using SPSS version 25*

Results: *Of the 17 patients included in our study, 70% were females 30% were males with with mean age of 32.35 years. Three out of seventeen patients had juvenile dermatomyositis while the remaining 14 patients were diagnosed with classic adult dermatomyositis. Heliotrope rash was the most common skin feature and was found in 76 %. Gottron papules were seen in 65 %, poikloderma was seen in 70% and calcinosis was seen in 23% of patients. 70% of patients reported muscle weakness, 29.4% reported pulmonary symptoms and Gastrointestinal symptoms were seen in 35% of patients. 18% had weight loss, arthralgia was reported in 35% and 23.5% had dysphonia while 35% had dysphagia. Elevation in creatinine kinase (CK) was the most common laboratory abnormality and was seen in elevated in 65% of patients. ESR was elevated in 10/17 (59%), C- reactive protein (CRP) was elevated in 9/17 (52%) and ANA was positive in 6/17 (35%). Serum vitamin D was tested in 14 out of the 17 patients and 10/14 (71%) had low serum vitamin D. All patients received systemic steroids and methotrexate was used in 52% of patients. Mycophenolate mofetil (MMF) was used in 29% of patients and IV immunoglobulin (IVIG) was used in 52% of them. Rituximab was used in 41%, azathioprine in 11%, plasmapheresis in 5% and hydroxychloroquine (HCQ) was used in 17.5% of patients.*

Conclusion: *Clinical features and laboratory findings in our dermatomyositis patients are consistent with those findings reported in other studies from different countries with few minor differences as stated above. Based on our study findings, we suggest that all patients with dermatomyositis to be tested for vitamin D deficiency and treated accordingly given high prevalence of vitamin D deficiency among our study patients.*

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INTRODUCTION:

Dermatomyositis (DM) is an idiopathic autoimmune inflammatory condition that can cause inflammatory myositis along with characteristic skin manifestations.[1,2] It affects females more commonly than males. It can be diagnosed by using the Bohan and Peter criteria[1] , or by using 2017 EULAR/ACR diagnostic criteria which include proximal muscle weakness, raised serum levels of muscle enzymes, muscle biopsy showing disease and characteristic skin lesions.[1,6] Dermatomyositis is divided into two categories: without muscle involvement (amyopathic DM or DM without myositis) and with muscle involvement (adult DM associated or not with cancer , juvenile DM and overlap syndrome).[4]

Cutaneous manifestations of DM are grouped into pathognomonic (Gottron's papules), characteristic (shawl sign, heliotrope, periungual telangiectasias and mechanic hands) and compatible (poikiloderma atrophicans vasculare and cutaneous calcinosis).[5]

Dermatomyositis is a subset of Idiopathic inflammatory myopathies which are classified according to 2017 EULAR/ACR to the following subtypes: immune-mediated necrotizing myopathies (IMNM), polymyositis, inclusion body myositis, amyopathic dermatomyositis, dermatomyositis and juvenile dermatomyositis.[6]

Mortality rate from dermatomyositis is low when steroids and other immunosuppressants are used but there are still patients who suffer from the consequences of associated malignancies. A recent study in Saudi Arabia showed the prevalence of dermatomyositis associated malignancy to be 6.7%.[8] after introduction of systemic steroids.

Epidemiology and clinical outcome of dermatomyositis can vary from different parts of the world.[9,10] and we aim to specifically study dermatomyositis in Saudi Arabia and compare our data to published data from other countries. We also wanted to study the prevalence of vitamin D deficiency among dermatomyositis patients.

MATERIALS AND METHODS AND DATA ANALYSIS:

We conducted our retrospective study at 3 hospitals in central Saudi Arabia using data extracted from electronic medical records (EMR). We included all dermatomyositis seen at follow up in participating hospitals (Prince Sultan Military Medical City, King Faisal Hospital and research center, King Fahad Hospital in Buraidah city) for the period between July 2017- July 2021. Only patients who met the diagnostic criteria of dermatomyositis based on the Bohan and Peter criteria were included. Data obtained from patient medical records included age, sex, investigations and treatment modalities. Statistical analysis was performed using SPSS version 25

Prior to starting this study, ethical approval was obtained from Qassim Research Ethics Committee on July 10,2021 with IRB no. 200805 .

RESULTS:

17 patients were included in our study. 12 out of 17 were females (70%) and 5 were males (30%). Patients ranged in age from 7 to 58 years, with mean age of 32.35. Three out of seventeen patients had juvenile dermatomyositis while the remaining 14 patients were diagnosed with classic adult dermatomyositis.

With regards to skin manifestations, heliotrope rash which is defined as violaceous erythema on eyelids was seen in 13 out of 17 patients (76 %). Gottron papules defined as erythematous papules over MCP and DIP joints were seen in 11 out of 17 patients (65 %). Poikloderma which is a combination of skin atrophy, telangiectasia, hyperpigmentation and hypopigmentation was seen in 12 out of 17 patients (70%). Mouth ulcers were seen in 5 out of 17 patients (29%). Calcinosis was seen in 4 out of 17 patients (23%) with 3 out of these 4 patients classified as having juvenile dermatomyositis.

Table 1 Characteristic dermatomyositis skin lesions in our study patients.

Skin lesion	Number of patients affected (%)
Heliotrope rash	13/17 (76%)
Gottron papules	11/17 (65%)
Poikiloderma	12/17 (70%)
Mouth ulcers	5/17 (29%)
Calcinosis	4/17 (23%)

We reviewed the systemic symptoms associated with dermatomyositis in our patients and we found that 3/17 (18%) had weight loss and 12/17 (70%) reported weakness. Lymphadenopathy was seen in one patient only while shortness of breath was present in 5/17 (29.4%). Gastrointestinal symptoms were seen in 6/17 (35%) and arthralgia was present in 6/17 (35%). 4 out of 17 patients (23.5%) had dysphonia while 6/17 (35%) had dysphagia.

Table 2: Systemic symptoms of dermatomyositis in our study patients.

Systemic symptom	Number of patients affected (%)
Weight loss	3/17 (18%)
Muscle weakness	15/17 (70%)
Lymphadenopathy	1/17 (5%)
Shortness of breath	5/17 (29.4%).
GI symptoms	6/17 (35%)
Arthralgia	6/17 (35%)
Dysphonia	4/17 (23.5%)
Dysphagia	6/17 (35%)

Laboratory investigations were reviewed for all study patients, and we found that Creatinine Kinase (CK) was elevated in 11/17 (65%). Aldolase was tested in only 6 patients out of the 17 patients included in the study and aldolase was elevated in 4 out of those 6 patients. Aspartate aminotransferase (AST) was elevated in 4/17 (23.5%) and alanine aminotransferase (ALT) was elevated in 3/17 (18%). ESR was elevated in 10/17 (59%), C- reactive protein (CRP) was elevated in 9/17 (52%) and ANA was positive in 6/17 (35%). Anti Jo antibody was tested in 5 out of the 17 patients and none of them had positive anti jo antibody. Anti dsDNA antibody was tested in 10 patients and only one patient had a positive result. Serum vitamin D was tested in 14 out of the 17 patients and 10/14 (71%) had low serum vitamin D.

Table 3: Laboratory investigations of dermatomyositis in our study patients.

Investigation	Normal	Abnormal
Creatinine Kinase (CK)	6/17	Elevated in : 11/17
Aldolase (tested in 6 out of 17 patients only)	Normal level in 2 out of 6	Elevated in 4 out of 6 patient
AST	13/17	Elevated in 4/17
ALT	14/17	Elevated in 3/17
ESR	7/17	Elevated in 10/17
CRP	8/17	Elevated in 9/17
ANA	6/17	Elevated in 11/17
Anti Jo (tested in 5 out of 17 patients only)	Normal level in 5 out of 5 patients tested	Elevated in 0/5
dsDNA	9 out of 10 patients tested	Elevated in 1/10
Vitamin D	4 out of 14 patient tested	Low in 10/14

Table 4: Miscellaneous tests of dermatomyositis in our study patients.

Investigation	Number of patients
MRI	3/17
Muscle biopsy	12/17
EMG	1/17

With regards to treatment modalities in our study patients, systemic steroids were used in 17/17 (100%) and methotrexate was used 9/17 (52%). IV immunoglobulin (IVIG) was used in 9/17 (52%). Mycophenolate mofetil (MMF) was used in 5/17 (29%) and Rituximab was used in 7/17 (41%). Azathioprine was used in 2/17(11%), plasmapheresis was used in 1/17 (5%) and hydroxychloroquine (HCQ) was used in 3/17 (17.5%).

Table 5: Treatment modalities in dermatomyositis in our study patients .

Treatment :	Number of patients treated (%)
Systemic steroids	17/17 (100%)
Methotrexate	9/17 (52%)
IVIG	9/17 (52%).
MMF	5/17 (29%)
Rituximab	7/17 (41%).
Azathioprine	2/17 (11%)
Plasmapheresis	1/17 (5%)
Hydroxychloroquine (HCQ)	3/17 (17.5%).

DISCUSSION:

In this study, we reviewed the clinical features, investigations and treatment modalities in dermatomyositis patients in central Saudi Arabia. With regards to skin manifestations, heliotrope rash was the most common feature which is consistent with findings in other studies[10],[11],[12]. Poikloderma was seen in 70% of our study patients which is higher than the prevalence of poikloderma in Jordan reported by Mustafa et al.[10]. The prevalence of Gottron's papules was 65% of our patients which is consistent with a prevalence of 69% reported by Duncan AG et al [13] but more than a prevalence of 52% reported by Mustafa et al.[10]. Calcinosis is more frequently seen in juvenile dermatomyositis and our findings were consistent with that. Calcinosis was found in 4/17(23%) of our study patients with 3 of these patients having juvenile dermatomyositis and one with classic dermatomyositis compared to a prevalence of 38% of patients with juvenile dermatomyositis reported by Baurt K et al .[14]

Khadir Mustafa et al studied 30 patients with dermatomyositis and reported the prevalence of systemic symptoms in these patients 100% of them having muscle weakness, 40% with dysphagia, 26% with shortness of breath, 20% with arthralgia and that was somewhat similar to our findings.[10]

ET Koh studied 75 patients with dermatomyositis and reported the prevalence of laboratory abnormalities seen in this subset of patients with raised CK in 89% of patients, positive ANA in 47%, raised aldolase in 95%, raised ESR in 49.7%.[15] By comparing our findings to ET Koh's study, we found a lower prevalence of CK elevation of 65%, lower prevalence of aldolase elevation in tested patients. In our study, elevation in ESR and ANA was found in 59% and 65% of patients respectively compared to an elevated ESR in 49.7% and a positive ANA in 47% of ET Koh's study patients [15].

Ciudad-Blanco et al. studied 20 patients with dermatomyositis in Spain and reviewed treatments that were used for those patients and reported the use of systemic steroids in 100% of patients, methotrexate in 70%, azathioprine in 20% and hydroxychloroquine in 25% of patients and none of them received IVIG or rituximab.[16] Our patients received the above medications in a quite similar percentages reported by Ciudad-Blanco et al but in addition to that, 52% of our patients received IV immunoglobulin (IVIG) was used in 9/17 (52%) and Rituximab was used in 41% of them.

The prevalence of vitamin D deficiency in our study patients was 71% which is quite significant and this confirms findings from a previous study by Azali P et al who studied 52 patients with dermatomyositis and found that 65% have vitamin D deficiency compared to only 21% of 290 healthy controls. [17] and they concluded that vitamin D deficiency could be a risk factor for developing dermatomyositis given high prevalence of vitamin D deficiency among dermatomyositis compared to healthy individuals.

Our study limitations include: small sample size, the retrospective nature of this study and the fact that it was done in only 3 hospitals in Saudi Arabia.

In conclusion, clinical features and laboratory findings in our dermatomyositis patients are consistent with those findings reported in other studies from different countries with few minor differences as stated above. Based on our study findings, we suggest that all patients with dermatomyositis to be tested for vitamin D deficiency and treated accordingly given high prevalence of vitamin D deficiency among our study patients.

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Disclosure of conflict of interest:
The authors declare that there is no conflict of interest regarding the publication of this paper

Description of supplementary data:
All the data used to support the findings of this study are included within the article.

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