

Dermatomyositis: a retrospective study

Mahroo Tajalli^a, MD, Terrence M. Vance^a, PHD, Burak Elkiran^b, MD, Anthony M. Reginato^b, MD, PHD, Abrar A. Qureshi^{a,c}, MD, MPH

^a: Department of Dermatology, Warren Alpert Medical School of Brown University, Providence, RI, USA

^b: Division of Rheumatology, Department of Medicine, Rhode Island Hospital, Warren Alpert Medical School of Brown University, Providence, RI, USA

^c: Department of Epidemiology, School of Public Health, Brown University, Providence, RI, USA

Background

Dermatomyositis (DM) is an idiopathic inflammatory dermatomyopathy that is characterized by varying degrees of skin disease with or without muscle involvement. It can be categorized into two major subtypes: clinically amyopathic dermatomyositis (CADM) and classic dermatomyositis (CDM). CDM presents with typical skin findings, proximal muscle weakness, and evidence of myositis. CADM refers to patients with the characteristic skin findings of DM but the absence of clinically-evident muscular weakness (1). It has been demonstrated that DM is strongly associated with a wide range of cancers and the risk of malignancy is present in both genders and all age groups (2,3). In this study, we aimed to identify characteristics of patients with DM that were associated with cancer.

Methods

We used ICD-9 and ICD-10 codes to identify patients diagnosed with DM. To ensure the diagnosis of DM we reviewed patients' charts to confirm the diagnosis of DM. One patient was excluded because of a coding error. All data including demographic information, type of dermatomyositis (CADM or CDM), history of juvenile DM, cancer diagnosis, type and date of cancer diagnosis, medications, muscle enzymes (creatine phosphokinase (CPK) and aldolase), associated calcinosis cutis, panniculitis, esophageal and long involvement, autoantibody profile, body mass index (BMI) and smoking status (ever versus never) were obtained by reviewing patients' electronic medical records. We used descriptive statistics to present characteristics of DM patients. The association between a variety of factors and risk of cancer was examined using logistic regression (figure 5).

RESULTS

A total of 33 patients with DM were identified. Of these, 8 (24 %) were male and 25 (76 %) were female. The average age at diagnosis was 50 years, with a standard deviation (SD) of 17 years. The number of patients with CDM was 20 (61%), whereas 13 (39%) had CADM. The ethnicity of patients was White (70%), Hispanic (12%), Black (6%), Asian (3%), and unknown (9%). Pulmonary disease with restrictive pattern, esophageal involvement, calcinosis cutis and panniculitis were reported in 6 (18%), 7 (21%), 4 (12%) and 2 (6%) of patients, respectively. Muscle enzymes (CPK and/or aldolase) were high in 20 (61%), normal in 9 (27%) and unknown in 4 (12%) patients. Of note, among 13 patients with CADM, 4 patients had elevated levels of muscle enzymes. Positive antinuclear antibodies (ANA) were reported in 17 (52%) patients, with "speckled" the most common pattern. Only one patient was recorded as having juvenile DM.

Over one-third (n=12, 36 %) of patients had a history of cancer, including cancers of breast 4 (12%), prostate 2 (6%), T-cell lymphoma, gastric lymphoma, non-small cell lung cancer, cervix, squamous cell carcinoma (SCC) of the tonsil and invasive skin SCC, each one (3%). Among these cases of cancer, three cases occurred within 1 year, 5 cases within 5 years and 4 cases more than 5 years before/after the diagnosis of DM. In the last group a case of prostate cancer was diagnosed 12 years before the diagnosis of DM, however, the cancer recurred as a metastatic disease 2 months after the diagnosis of DM. Average age at diagnosis for patients with cancer was 57 years (SD: 12) and for patients without cancer was 45 years (SD: 19). We found no evidence of an association between gender, type of DM, enzyme level, pulmonary involvement, obesity or positive ANA and cancer, however, history of current or former smoking was strongly associated with increased risk of cancer (OR: 7.50, 95% CI: 1.49-37.66).

Medications used by patients for DM in order of frequency (n, %) include prednisone (21, 64%), hydroxy chloroquine (20, 61%), methotrexate (14, 42%), azathioprine (9, 27%), intravenous immunoglobulin (6, 18%), mycophenolate mofetil (3, 9%), rituximab (3, 9%), and thalidomide (1, 3%).

DISCUSSION

Our study results show a higher prevalence of malignancies among DM patients as compared to the general population, which is consistent with previous studies (2,3). Most cases of cancer (8 out of 12) occurred within 5 years of the diagnosis of DM. In our study, the average age at diagnosis of DM was higher in patients with cancer compared to those without cancer. It means risk of malignancy is more with greater age at diagnosis, which has also been demonstrated in other studies (3,5). With regard to gender, we found no association between gender and cancer which is in contrast to previous studies that reported higher risk of malignancy in males compared to females (3,4). This difference of cancer risk with respect to gender has been previously attributed to increased cancer risk among male individuals in the general population, possibly due to higher prevalence of male smokers. In our study, smoking (ever versus never) was strongly associated with cancer. This can be explained by the fact that smoking is a well-known risk factor for different types of malignancies and might have a synergistic effect in patients with DM, who already have a higher risk of cancer. We found no association between pulmonary involvement and cancer, which is consistent with the study by Fardet et al (5). However, Chen et al. showed interstitial lung disease was associated with a diminished risk of malignancy (4). Limitations of our study include small sample size and retrospective study design.

Conclusion

DM patients are at an increased risk of developing malignancy and history of smoking was significantly associated with an increase in this risk. DM patients should be strongly advised to quit smoking.

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RESULTS

Figure 1. Type of Dermatomyositis (A), Sex (B), and Race/ethnicity of cases (C)

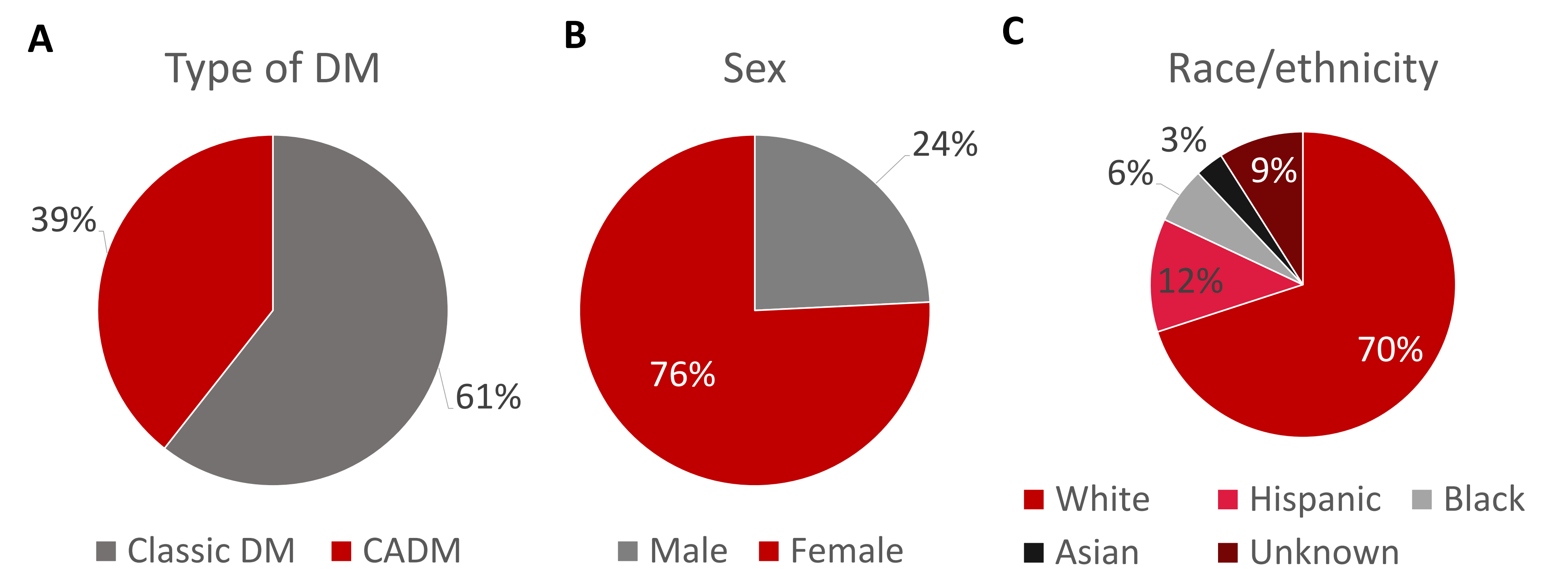


Figure 2. Type and frequency of medications used by cases

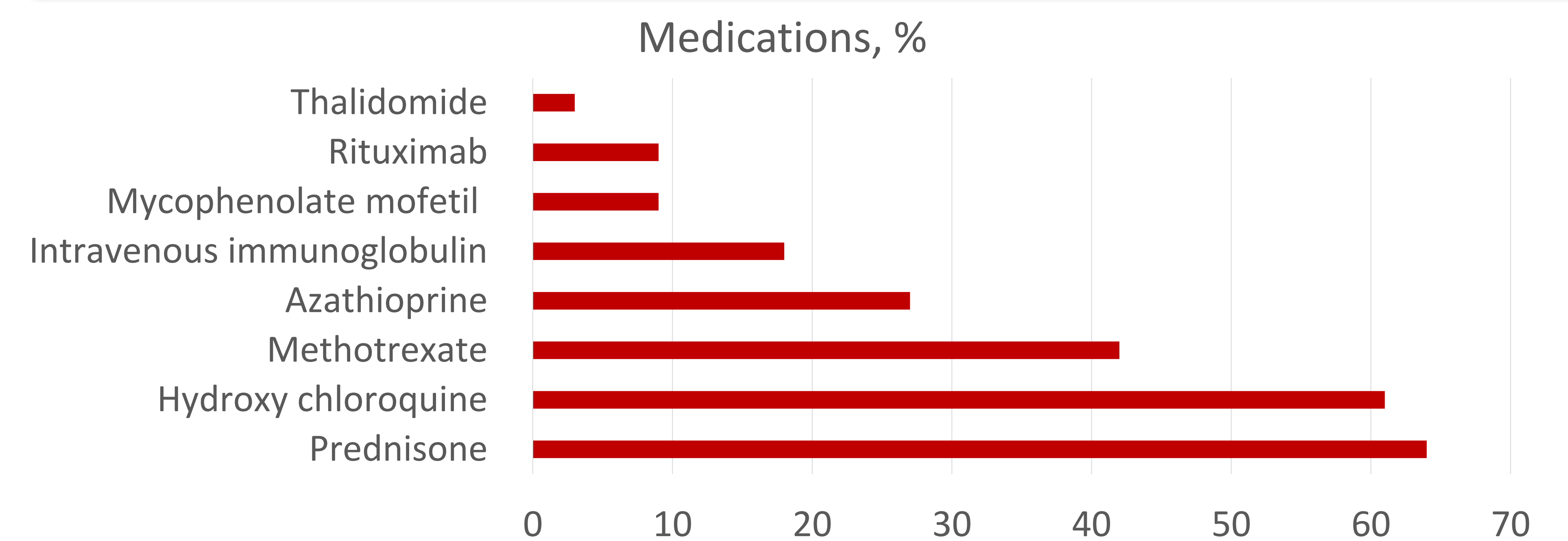


Figure 3. Prevalence of pulmonary disease, esophageal involvement, calcinosis cutis, and panniculitis among cases

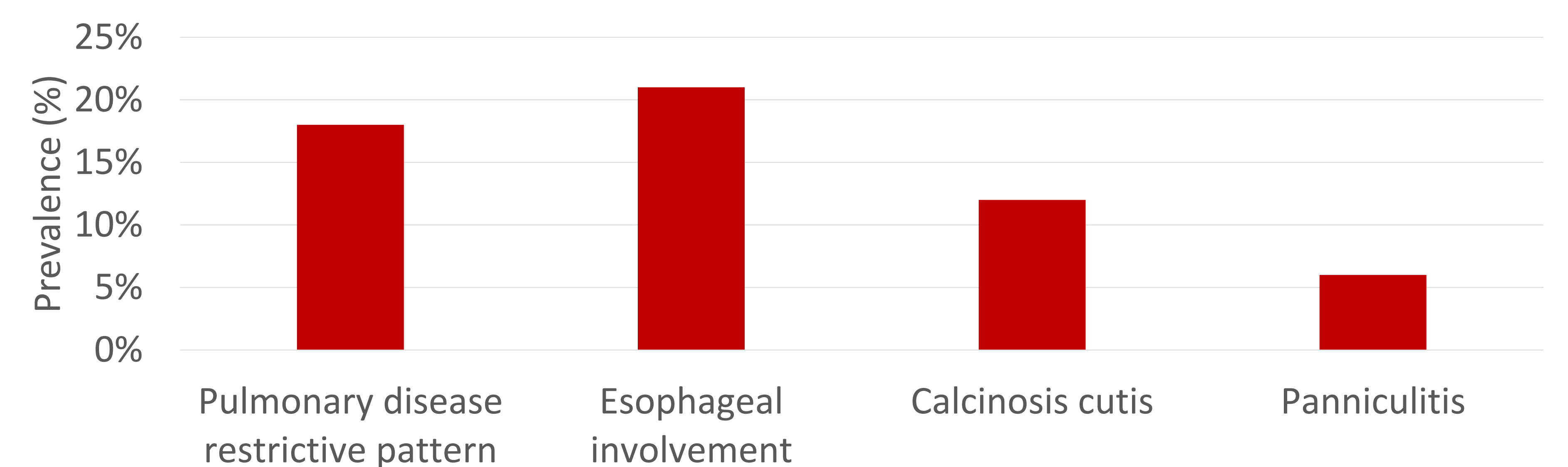


Figure 4. Age at DM diagnosis

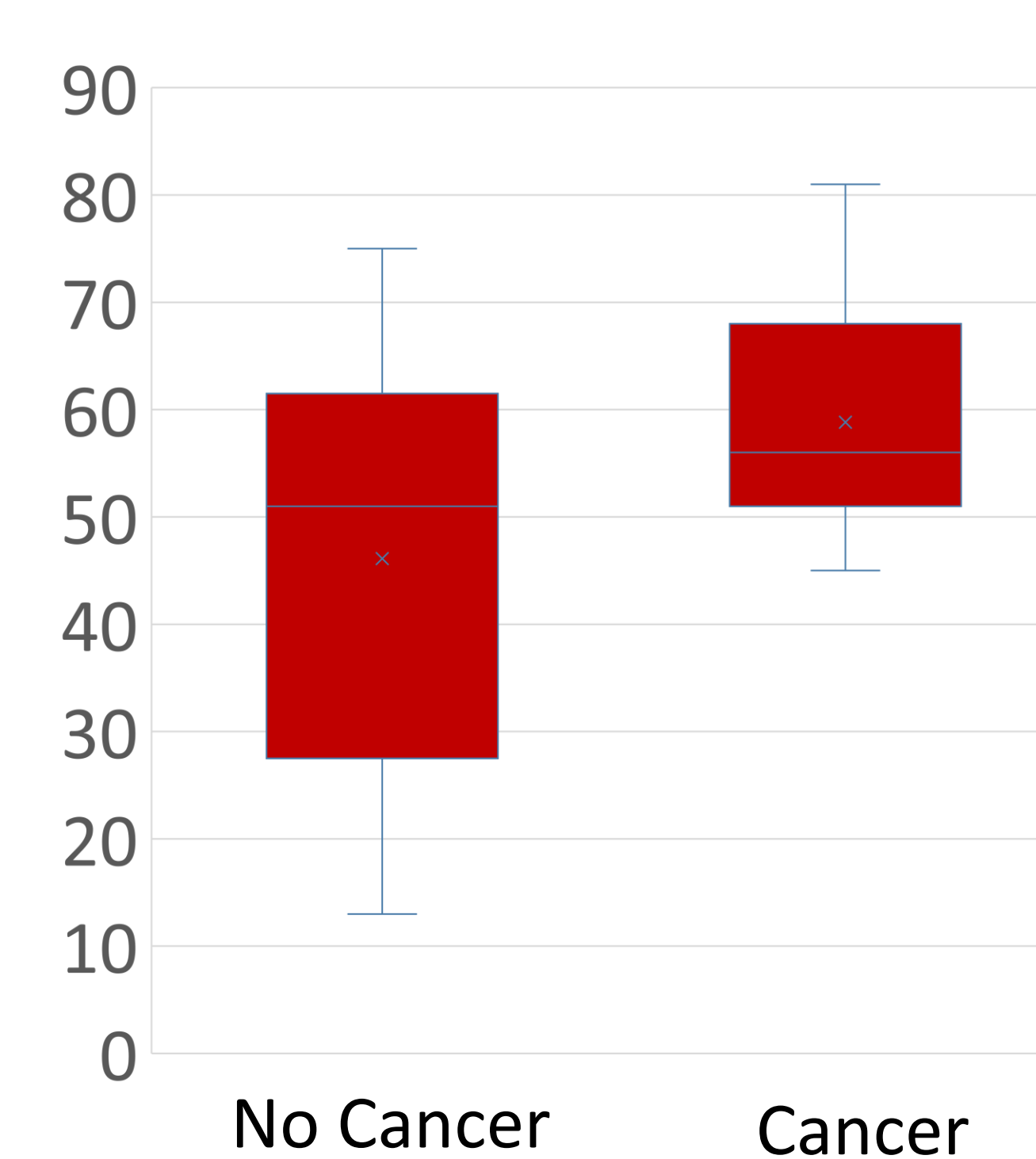
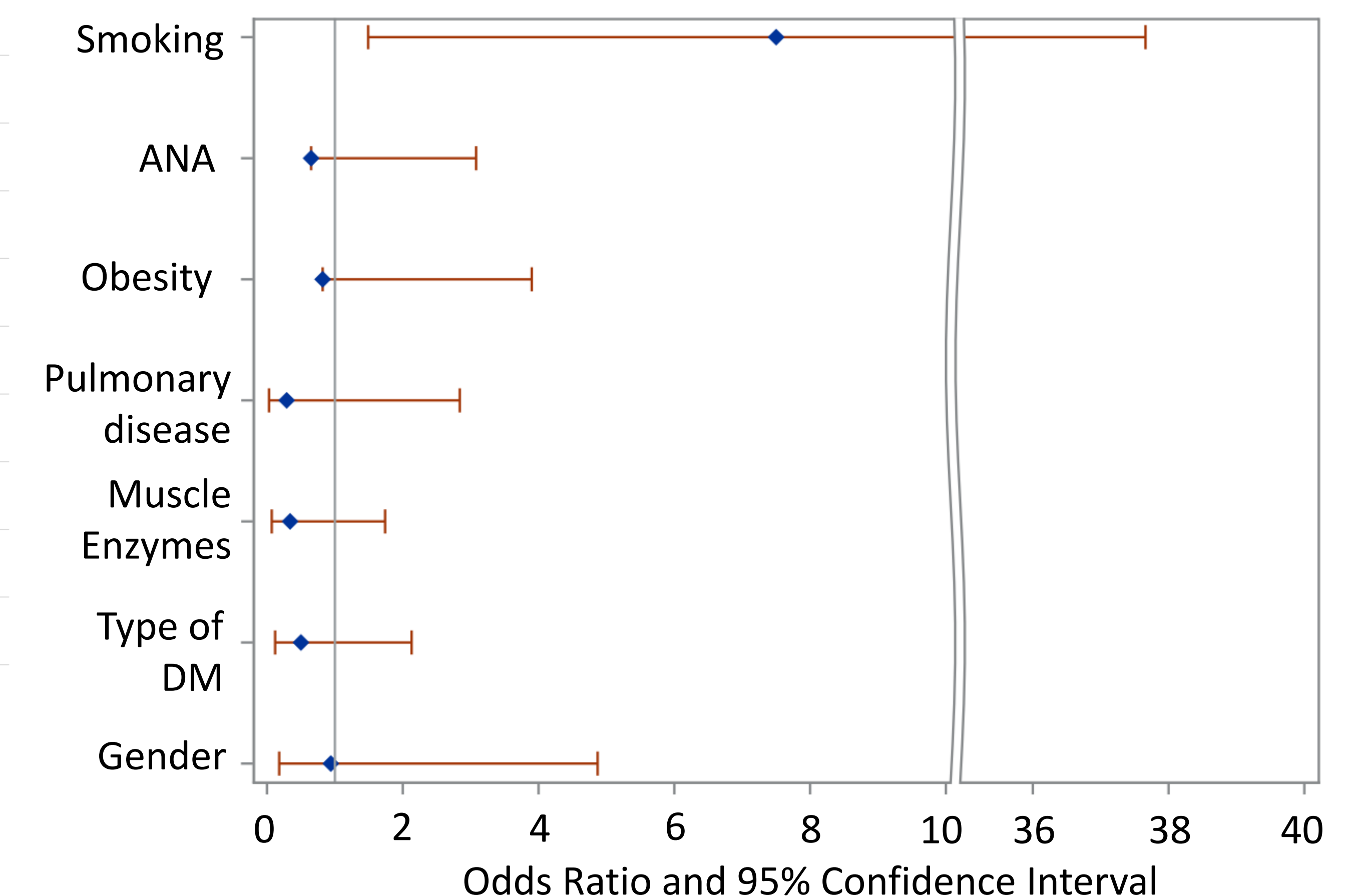


Figure 5. Risk Factors associated with cancer among cases



Disclosure: Authors have no relationships to disclose.

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Affiliations:

a: Department of Dermatology, Warren Alpert Medical School of Brown University, Providence, RI, USA

b: Division of Rheumatology, Department of Medicine, Rhode Island Hospital, Warren Alpert Medical School of Brown University, Providence, RI, USA

c: Department of Epidemiology, School of Public Health, Brown University, Providence, RI, USA

Background: The association between dermatomyositis (DM) and cancer has been established (1,2). In this study, we aimed to identify characteristics of patients with DM that were associated with cancer.

Methods: We used ICD-9 and ICD-10 codes to identify patients diagnosed with DM. All data was obtained by reviewing patients' electronic medical records.

Results: A total of 33 patients with DM were identified. Of these, 8 (24 %) were male and 25 (76 %) were female. The average age at diagnosis was 50 years, with a standard deviation (SD) of 17 years. The number of patients with skin and muscle involvement (classic DM) was 20 (61%), whereas 13 (39%) had clinically amyopathic DM (CADM). The ethnicity of patients was White (70 %), Hispanic (12 %), Black (6 %), Asian (3%), and unknown (9 %). Pulmonary disease with restrictive pattern, esophageal involvement, calcinosis cutis and panniculitis were reported in 6 (18 %), 7 (21 %), 4 (12 %) and 2 (6 %) of patients, respectively. Muscle enzymes (creatine phosphokinase and/or aldolase) were high in 20 (61 %), normal in 9 (27 %) and unknown in 4 (12 %) patients. Of note, among 13 patients with CADM, 4 patients had elevated levels of muscle enzymes. Positive antinuclear antibodies (ANA) were reported in 17 (52 %) patients, with "speckled" the most common pattern.

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immunoglobulin (6, 18 %), mycophenolate mofetil (3, 9 %), rituximab (3, 9 %), and thalidomide (1, 3 %).

Conclusion: Among all characteristics examined, only smoking was strongly associated with an increased risk of cancer in DM patients.

Limitations: small sample size.

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Authors have no relationships to disclose.