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Characteristics of patients with juvenile dermatomyositis from 2001-2021 at a tertiary care center

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Abstract

Background: Juvenile dermatomyositis (JDM) is the most common inflammatory myopathy in the pediatric population and can represent a medical emergency. However, many features of JDM remain poorly understood, disease presentation is highly variable, and predictors of disease course have yet to be identified.

Methods: This retrospective chart review included 47 JDM patients seen at a tertiary care center over a 20-year period. Characteristics such as demographics, clinical signs and symptoms, antibody positivity, dermatopathology features, and treatments were recorded.

Results: All patients had evidence of cutaneous involvement, whereas 88.4% experienced muscle weakness. Constitutional symptoms and dysphagia were commonly present. The most frequent cutaneous findings were Gottron papules, heliotrope rash, and nailfold changes. Anti-TIF1γ was the most prevalent myositis-specific autoantibody. Management involved systemic corticosteroids in nearly all cases. Strikingly, the dermatology department was only involved in the care of four in every ten (19/47) patients.

Conclusions: Prompt recognition of the strikingly reproducible skin findings present in JDM can improve disease outcomes in this population. This study highlights the need for increased education of such pathognomonic findings as well as more multidisciplinary care. In particular, a dermatologist should be involved in the care of patients presenting with muscle weakness and skin changes.

Keywords: autoantibody, autoimmune, JDM, juvenile dermatomyositis, myopathy, retrospective study, vasculopathy

Introduction

Juvenile dermatomyositis (JDM) is the most common inflammatory myopathy in the pediatric population [1]. Although the exact cause of JDM is unknown, it is suspected that a genetic predilection may be triggered by environmental factors, leading to an immune-mediated inflammatory cascade and subsequent systemic vasculopathy [2,3]. This ultimately causes capillary destruction and surrounding tissue necrosis in most of an affected patient's tissues [4,5]. Characteristic clinical features are related to tissue inflammation and include proximal muscle weakness and dermatologic changes such as Gottron papules, heliotrope rash, nailfold changes, and dystrophic calcification. Additional findings can include nonspecific systemic findings such as fatigue, fever, or arthralgias. The most feared complication is vasculopathy, which can result in bowel perforation and even death. Unlike adult dermatomyositis (DM), JDM is not commonly associated with internal malignancy or interstitial lung disease. Autoantibodies such as anti-NXP2 and anti-TIF1γ are commonly seen in JDM and can help predict disease phenotype, but testing remains variable [6,7]. The disease course of JDM can be monophasic, polyphasic, or chronic and predictors of disease course have yet to be identified [8]. Reports

of clinical and dermatopathological findings in JDM populations are rare in the literature. Juvenile dermatomyositis is a multifaceted disease; characterization of this diverse patient population at University of California, Davis (UC Davis), [9], a tertiary care center in Sacramento, California, can help define the presentation of JDM and inform patient care by clinicians and researchers.

Methods

This was a retrospective chart review study that assessed characteristics of all patients with a diagnosis of JDM who were seen at UC Davis between 2001-2021. This study was approved by the UC Davis Institutional Review Board. The electronic medical record was searched for the International Classification of Diseases 9th Revision (ICD 9) code 710.3 (dermatomyositis) and ICD 10 codes M33.0 (juvenile dermatomyositis) and M33.10 (dermatomyositis) between November 1, 2001, and November 30, 2021. A total of 354 charts resulted and 307 were excluded based on diagnosis after age 18 or due to physician-documented rule-out of JDM. Patients of any age were included if they received a diagnosis of JDM prior to the age of 18 years. A total of 47 individuals were included in statistical analyses which consisted of descriptive statistics and Fisher's exact test. Because this is a small, exploratory study, a P value of 0.1 was considered statistically significant and an indication of a promising direction for future research. Statistical analyses were conducted using SAS® software version 9.4 (SAS Institute Inc., Cary, NC) for Windows®.

Results

Demographics

The mean age at diagnosis in our study cohort was 7.1 years (SD 4.4), consistent with the national average [1]. Two-thirds were female (74.5%, 35/47) and 46.8% (22/47) were White, followed by 36.2% Unknown, 29.8% (14/47) Hispanic or Latino, 17% (8/47) African American or Black, and 4.3% (2/47) Asian (**Table 1**). Note that some patients identified with multiple races/ethnicities. Our cohort traveled a mean of 104.3 miles (range 2-2289) from their home zip code to receive care at UC Davis Medical Center, highlighting the wide geographic region cared for at UC Davis.

Table 1. *Demographics of patients with juvenile dermatomyositis at University of California Davis from 2001-2021.*

Demographic	Value (%) N=47
Age at diagnosis (years)	Mean 7.1
Legal sex:	
Female	35 (74.5)
Male	12 (25.5)
Race/ethnicity:	
African American or Black	8 (17.0)
Asian	2 (4.3)
Hispanic or Latino	14 (29.8)
White	22 (46.8)
Unknown	17 (36.2)

Care setting

Of the patients with care setting reported, it was more common for our cohort to have been diagnosed in the outpatient setting (42.6%, 20/47) than inpatient (25.5%, 12/47), though this frequency was much lower than the 85.3% of adult dermatomyositis patients diagnosed in the outpatient setting at UC Davis. Strikingly, less than half of patients had a dermatologist involved in their JDM care (40.4%, 19/47). Like the adult DM population at UC Davis, the top two specialties that diagnosed dermatomyositis in our cohort were Rheumatology/Allergy (48.3%, 14/29) and Dermatology (31.0%, 9/29), followed by Pediatrics (20.7%, 6/29), [10]. The remaining 18 patients did not have sufficient records available regarding which specialty made their JDM diagnosis.

Past medical/family history

A small number of patients (8.5%, 4/47) had concomitant autoimmune disease such as type one diabetes or overlap syndrome with systemic sclerosis. However, given the young age and limited length of follow-up in most, this likely does not represent our cohort's potential to develop additional autoimmune disease later in life. One in every five had a family history of autoimmune disease, which was the same frequency seen in the adult DM population at UC Davis (21.3% and 21.8%, respectively), [10].

Cutaneous findings

Cutaneous findings in JDM are often strikingly reproducible, such as the pathognomonic Gottron papules, Gottron sign, and heliotrope rash [1]. Accordingly, in the 40 patients with physical examination findings on record, the most common cutaneous findings were Gottron papules (70%, 28/40), heliotrope rash (55%, 22/40), and nailfold changes (55%, 22/40), followed by Gottron sign (42.5%, 17/40) and malar rash (40%, 16/40), (**Table 2**). All 40 of these patients had some evidence of cutaneous involvement. Nailfold changes such as capillary abnormalities, erythema, and ragged cuticles are considered characteristic findings of JDM, along with shawl sign, V-neck sign, and Holster sign, which were seen in 7.5% (3/40), 5% (2/40), and 2.5% (1/40) of our cohort, respectively. Given the lack of dermatologist involvement, it is likely that these specific dermatologic signs were underreported. Notably, dystrophic calcinosis occurred in 20.9% (9/40) of patients, which is consistent with the literature but significantly higher than the 1.9% of affected adult DM patients at UC Davis [1,10]. More rare and nonspecific dermatologic examination findings included the following: 12.5% with oral mucosal involvement (e.g., ulceration), extensor surface rash, and erythema, each. There were 7.5% with pruritus. 5% with inverse Gottron papules. and 2.5% with Holster sign, mechanic's hands, flagellate erythema, livedo reticularis, sclerodactyly, and edema, each. Records were insufficient to assess for prevalence of lipodystrophy or contractures in our cohort.

Evidence of myositis

Juvenile dermatomyositis can be classified as myopathic or amyopathic. Clinically amyopathic juvenile dermatomyositis (CAJDM) is characterized

Table 2. *Cutaneous findings noted on examination at the time of clinical presentation.*

Cutaneous finding	Morphologic features	Number (%) N=40
Gottron papules	Violaceous lichenoid papules affecting extensor joints, often on the hands	28 (70)
Heliotrope rash	Patchy reddish-purple rash on or around the eyelids, often accompanied by edema	22 (55)
Nailfold changes	Presence of any of the following on nailfold capillaroscopy: capillary dropout, branching and dilatation, areas of hemorrhage, decrease in number of vessels per millimeter	22 (55)
Gottron sign	Symmetric confluent macular violaceous hue affecting olecranon processes, patellae, and/or medial malleoli	17 (42.5)
Malar rash	Erythematous flat or raised rash affecting the bridge of the nose and cheeks, often notably sparing the nasolabial folds	16 (40)
Calcinosis cutis	Palpable nodule(s) formed by calcium deposition in the skin	9 (20.9)
Oral mucosal involvement	Ulceration of oral mucosa	5 (12.5)
Nonspecific extensor surface rash	Nonspecific rash affecting extensor extremities which cannot be better described by another pathognomonic finding	5 (12.5)
Nonspecific erythema	Nonspecific areas, red in color, which cannot be better described by another pathognomonic finding	5 (12.5)
Pruritus	Itching	3 (7.5)
Shawl sign	Violaceous erythema or poikiloderma of the upper back, often extending onto the lateral upper arms	3 (7.5)
V-neck sign	Violaceous erythema or poikiloderma on the anterior chest	2 (5)
Inverse Gottron papules	Violaceous lichenoid papules affecting the palmar hands	2 (5)
Holster sign	Papules or plaques involving the lateral hips	1 (2.5)
Mechanic's hands	Fingers and hands with rough, irregular cracking of the skin	1 (2.5)
Flagellate erythema	Pigmented or erythematous linear streaks	1 (2.5)
Livedo reticularis	Netlike pattern of reddish-blue skin discoloration	1 (2.5)
Sclerodactyly	Skin tightening which causes thin and shiny fingers	1 (2.5)
Edema	Nonspecific area of swelling due to excess fluid collection	1 (2.5)

by pathognomonic cutaneous findings in the absence of both clinical weakness and evidence of myopathy in laboratory and imaging data [11]. Our cohort presented with weakness in 88.4% of cases where clinical records were sufficient (38/43). Concordantly, 83% (39/47) had indications of muscle destruction by elevated muscle enzymes, muscle biopsy, electromyography (EMG), and/or muscle MRI consistent with JDM. This suggests that 11.6% (5/43) to 19.1% (9/47) of our overall cohort had clinically amyopathic disease, likely identified by pathognomonic cutaneous findings. The manifestations associated with clinically amyopathic disease in adult DM patients are rare in children. Although some CAJDM patients may have disease that progresses to classical JDM, the outcome of CAJDM appears to be good [6].

Myopathy can be assessed by invasive or non-invasive methods. Invasive testing includes EMG and muscle biopsy, whereas non-invasive methods include MRI and laboratory evaluation of creatinine kinase (CK), aldolase, aspartate aminotransferase (AST), alanine transaminase (ALT), and lactate dehydrogenase (LDH). As expected, in our juvenile cohort, invasive methods were utilized less frequently than non-invasive tests, with 19.2% (9/47) receiving muscle biopsy and only 2.1% (1/47) receiving EMG in contrast to 26.3% (41/156) of adult DM patients undergoing EMG at UC Davis [10]. Six in ten patients (59.6%, 28/47) received a muscle MRI for JDM diagnostic purposes, with 82% (23/28) of these revealing muscle disease involvement via evidence of edema.

Laboratory testing in our cohort revealed myopathy based on elevated muscle enzymes in 78% of those tested (32/41). Specifically, LDH was most commonly tested, with elevated levels averaging 397.3 (30/32). In addition, elevated AST averaged 121.9 (25/32); elevated aldolase levels averaged 24 (22/32); elevated ALT averaged 114.2 (20/32); and elevated CK averaged 6188.7 (19/32). Although the adult DM population at UC Davis demonstrated an association between low complement C3 or C4 levels with clinical/laboratory evidence of muscle destruction, complement levels were very rarely tested in the JDM population [10]. Instead, lymphopenia and von Willebrand Factor (vWF) antigen, which have been suggested as indicators for disease activity [12], were present in 42.9% (18/42) and 31% (13/42) of those tested, respectively.

The highly variable diagnostic modalities utilized likely reflects the fact that there is no gold standard for diagnosing JDM. Although muscle biopsy and EMG are invasive, they offer specific information about the inflammatory response. Given the invasive nature of a muscle biopsy, it is striking that our juvenile cohort underwent more muscle biopsies than skin biopsies (19.2% and 12.8%, respectively). Muscle MRI is both noninvasive and highly sensitive for identifying muscle inflammation, but it cannot identify the cause of inflammation which may also be evident in some dystrophies. Of note, nailfold capillaroscopy, which was not utilized in our cohort, is highly sensitive for diagnosing JDM and can be particularly helpful for differentiating JDM from muscular dystrophies and other myopathies [1].

Antibody positivity

One in every four of those tested had positive antinuclear antibodies (ANA), (43.9%, 18/41). The most common myositis-specific autoantibodies (MSA) in those who underwent testing were anti-TIF1γ (12.8%, 5/39), which has been identified as the most prevalent MSA in the overall JDM population as well [6]. Anti-MJ/NXP-2 was the next most prevalent MSA in our cohort (7.7%, 3/39). These findings could have related to the association of high ultraviolet (UV) light index with higher prevalence of anti-TIF1γ and anti-MJ/NXP-2 positivity, given that all but one patient in our cohort lived in California [6]. Our population had 5.1% positivity with anti-Mi2, anti-MDA5, and anti-Scl100, each (2/39). Only one case of positivity was found for each of the following antibodies: anti-Scl70, anti-GAD-65, anti-DNaseβ, anti-SSA52, and rheumatoid factor (2.6%, 1/39). Note that larger studies have found that at least one MSA can be identified in around 60% of JDM patients [13]. Additionally, MSAs have recently been studied by various groups in the literature in association with certain clinic features and treatment responsiveness. Although MSA phenotypes in JDM are considered to be similar to adult DM patients with the same MSAs [6], we were unable to clearly identify disease

associations with specific antibodies in our cohort due to small sample size and a lack of cohesive testing given the twenty-year period that was studied.

Dermatopathology findings

Only 6 (12.8%) of our 47 patients received a skin biopsy for JDM diagnostic purposes. Two biopsies were performed at hospitals not affiliated with UC Davis and did not have available pathology reports in our system. The four biopsies performed at UC Davis were re-examined and characterized by a dermatopathologist (MK), (**Table 3**). As expected, interface dermatitis with vacuolar changes was reported in all, as well as vascular dilatation and periadnexal inflammation. In 75% (3/4) of skin biopsies, extravasated erythrocytes, pigment incontinence, and superficial dermal vessel wall thickening were noted. Half the specimens had increased dermal mucin, as well as nuclear dust which has not previously been described. Orthokeratosis was most commonly described (75%, 3/4), followed by 50% with hyperkeratosis and a basket-weave pattern; one exhibited parakeratosis. Each of the following features were identified in 25% (1/4) of specimens: basement membrane thickening, epidermal atrophy, spongiosis, and fibrosis in the superficial dermis. As expected, the inflammatory infiltrate was lymphocytic and limited to the superficial dermis in all four specimens. We observed extension of inflammation into the mid-dermis in one case, as well as sparse neutrophils (25%, 1/4). There is a dearth of data on dermatopathological findings in JDM and clinicopathologic correlation in the literature, with an emphasis on muscle biopsy pathology instead [14]. Of note, Wolstencroft et al. recently published a study on clinicopathologic correlation in the adult DM population [15]. Our most frequently observed findings were consistent with theirs, except our cohort had a higher incidence of periadnexal inflammation and lower incidence of increased dermal mucin. However, due to our small sample size, it is difficult to draw any significant conclusions from these differences.

Systemic findings

Systemic complications were relatively uncommon in our cohort, with the most commonly involved organ system outside of the skin and skeletal muscle being the gastrointestinal tract (44.7%, 21/47), (**Table 4**). Recall that, as discussed above, 20.9% of our cohort had dystrophic calcinosis as well. Pulmonary involvement was present in the form of interstitial lung disease in 8.5% (4/47). Although analysis of the UC Davis adult DM population revealed a significant association between African American/Black race and pulmonary disease involvement, we were unable to demonstrate this association in our cohort (P value >0.99), [10]. In terms of vascular disease involvement, one patient (2.1%) had a deep vein thrombosis at the age of 19 years. As expected, there were no patients in our cohort with malignancy, cardiac involvement, or death. However, most patients had a limited length of follow up available in our records. Of note, at the time of presentation, a majority of patients had constitutional symptoms such as fever, malaise, and arthralgias (61.7%, 29/47), and 21.3% (10/47) had dysphagia.

Table 3. *Histopathology findings in lesional skin from patients with juvenile dermatomyositis.*

Histopathology finding	Number (%) N=4
Perivascular inflammation	4 (100)
Basal vacuolization	4 (100)
Dyskeratotic keratinocytes	4 (100)
Vascular dilatation	4 (100)
Periadnexal inflammation	4 (100)
Extravasated erythrocytes	3 (75)
Pigment incontinence	3 (75)
Superficial dermal vessel wall thickening	3 (75)
Dermal mucin	2 (50)
Nuclear dust	2 (50)
Basement membrane thickening	1 (25)
Epidermal atrophy	1 (25)
Spongiosis	1 (25)
Fibrosis in the superficial dermis	1 (25)
Inflammatory infiltrate:	
Superficial dermis involvement	4 (100)
Mid-dermis involvement	1 (25)
Lymphocytes	4 (100)
Neutrophils	1 (25)
Scale:	
Orthokeratosis	3 (75)
Hyperkeratosis	2 (50)
Basket-weave pattern	2 (50)
Parakeratosis	1 (25)

Table 4. *Systemic findings in patients with juvenile dermatomyositis.*

Systemic finding	Number (%) N=47
At time of presentation:	
Constitutional symptoms	29 (61.7)
Dysphagia	10 (21.3)
Complications:	
Gastrointestinal disease	21 (44.7)
Calcinosis	9 (20.9)
Interstitial lung disease	4 (8.5)
Vascular involvement	1 (2.1)
Cardiac involvement	
Malignancy	

Treatment

Limited data are available from randomized controlled trials to guide management decisions in JDM. In general, treatment goals focus on controlling active disease, normalizing physical function, and preventing long-term damage [16]. The mainstay of therapy at the time of diagnosis consisted of high-dose systemic corticosteroids and methotrexate [17]. Records regarding treatment were sufficient in 44 of our 47 patients. As expected, the most common treatment utilized throughout the disease course of patients in our cohort was systemic corticosteroids (87.2%, 41/44), followed by methotrexate (84.1%, 37/44) and hydroxychloroquine (70.5%, 31/44), (**Table 5**). Over half received intravenous immune globulin, (56.8%, 25/44), one-third received mycophenolate (34.1%, 15/44), and one in every five received a calcineurin inhibitor (tacrolimus or cyclosporine), (22.7%, 10/44). Strikingly, topical corticosteroids were utilized in only 7 patients (15.9%, 7/44) despite the presence of cutaneous involvement in 100% of those with physical examination records. Treatments that were less frequently utilized included cyclophosphamide (9.1%, 4/44), topical tacrolimus (6.8%, 3/44), etanercept (4.6%, 2/44), and rituximab (4.6%, 2/44). Note that most patients received multiple treatment modalities throughout their disease course. We found that 93.8% of those with myositis based on muscle enzymes were treated with methotrexate compared to 66.7% of those without myositis, but this association did not reach statistical significance (P value 0.06) and this effect size is in the opposite direction of expected.

Table 5. *Treatments utilized throughout the disease course of patients with juvenile dermatomyositis.*

Treatment	Number (%) N=44
Systemic steroids	41 (87.2)
Methotrexate	37 (84.1)
Hydroxychloroquine	31 (70.5)
Intravenous immunoglobulin	25 (56.8)
Mycophenolate	15 (34.1)
Calcineurin inhibitor	10 (22.7)
Topical steroids	7 (15.9)
Cyclophosphamide	4 (9.1)
Topical tacrolimus	3 (6.8)
Etanercept	2 (4.6)
Rituximab	2 (4.6)

Discussion

Juvenile dermatomyositis is an autoimmune inflammatory myopathy that presents variably, but often with symmetric proximal muscle weakness and pathognomonic cutaneous findings. Although JDM is rare, with an incidence of 3.2 per million children per year, the potential vasculopathic nature of disease makes it a medical emergency [1,18].

This study characterizes the variable clinical, laboratory/imaging, autoantibody, and dermatopathological findings in 47 JDM patients seen at UC Davis from 2001-2021. All patients with sufficient records had evidence of cutaneous involvement, whereas 88.4% presented with weakness. It was also common for our cohort to present with constitutional symptoms and dysphagia. Our most frequent cutaneous findings were Gottron papules, heliotrope rash, and nailfold changes. Dermatopathology findings present in all examined specimens included interface dermatitis with vacuolar changes, perivascular inflammation, vascular dilatation, periadnexal inflammation, and lymphocytic infiltrate in the superficial dermis. Although myositis-specific autoantibodies weren't consistently tested, anti-TIF1γ was most prevalent. Disease sequelae included gastrointestinal involvement and dystrophic calcinosis. Muscle MRI was frequently utilized for diagnosis and management involved systemic corticosteroids in nearly all cases.

One of our most striking findings was the involvement of a dermatologist in the care of only

four in every ten (19/47) JDM patients throughout their disease course. Given this low frequency, many findings such as specific dermatologic examination signs and dermatopathology features were limited in our cohort. Additionally, skin biopsies were utilized less frequently than muscle biopsies in the diagnosis of this juvenile cohort and topical corticosteroids were underutilized whereas multiple immunosuppressants with potentially severe side effects were employed.

The clinical course of JDM is variable and little is known about prognostic indicators. However, time elapsed to treatment has been shown to affect disease outcomes, morbidity, and mortality, making it critical for clinicians to diagnose and treat in a timely manner [1,8,17,19]. Given that JDM patients present with strikingly reproducible skin findings, recognition of pathognomonic dermatologic signs can be a key to prompt recognition of JDM and thus improved outcomes for this patient population. We propose increased education of such pathognomonic findings and more multidisciplinary care, particularly involvement of a dermatologist.

Conclusion

Juvenile dermatomyositis is a medical emergency, but its variable presentation can make it difficult to diagnose. Additional, larger-scale studies are required to further characterize this multifaceted disease, including its variable presenting features, pathophysiology, clinicopathologic correlation, disease course predictors, and treatment. In the meantime, studies such as ours highlight the need for more multidisciplinary care, particularly with increased involvement of dermatologists, to improve disease outcomes in this patient population.

Potential conflicts of interest

The authors declare no conflicts of interest.

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