RESEARCH PAPER

Juvenile Dermatomyositis with Psoriasis: An Uncommon Pediatric Rheumatic Association

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ABSTRACT

INTRODUCTION: Autoimmune diseases are known to co-occur in genetically susceptible individuals [1,2]. The co-occurrence of juvenile dermatomyositis (JDM) and psoriasis is extremely rare, with only a few cases reported in the literature [3,4]. Management of this dual pathology presents unique therapeutic challenges, as conventional psoriasis treatments such as phototherapy and tumor necrosis factor-alpha inhibitors can paradoxically exacerbate clinical manifestations of JDM [5,6].

CASE REPORT: We report the case of an 18-year-old female presenting with features of both JDM and psoriasis. She was initially diagnosed with JDM at 8 years of age and received treatment at another hospital. Ten years later, she presented to our institution with progressive muscle weakness (Manual Muscle Testing score 64/80), calcinosis universalis, right knee contracture, and erythematous scaly plaques distributed over the scalp, neck, back, and abdomen. Laboratory investigations revealed positive anti-NXP2 antibodies. Histology from the plaques was suggestive of psoriasis vulgaris. She was treated with high-dose corticosteroids, methotrexate, and rituximab, following which her psoriatic lesions resolved, muscle strength improved significantly, and progression of calcinosis was arrested.

CONCLUSION: The coexistence of JDM and psoriasis represents a rare and complex clinical scenario. Selection of immunosuppressive agents that effectively treat both conditions without exacerbating either disease is crucial for optimal patient outcomes.

KEYWORDS: juvenile dermatomyositis, psoriasis, rituximab, calcinosis universalis, anti-NXP2 antibody.

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BACKGROUND

Autoimmune diseases frequently co-occur in genetically susceptible individuals, reflecting shared genetic etiologies and overlapping immune pathways [1,2]. Psoriasis, a chronic systemic inflammatory condition, has been reported to coexist with various autoimmune diseases, including systemic lupus erythematosus (prevalence 0.69-4.9%), systemic sclerosis, Sjögren's syndrome, adult-onset Still's disease, Behçet's disease, sarcoidosis, and dermatomyositis [7-9].

The prevalence of psoriasis in children is approximately 0.3% in North India and 1.4% in South India [10,11]. Notably, 20-35% of patients with psoriasis experience disease onset before 20 years of age [10]. HLA-Cw6 association with a familial incidence of 4.5-9.8% has been documented in children with psoriasis in India [12]. In a study conducted in North India, 37.8% of patients with juvenile dermatomyositis exhibited cutaneous

activity, with more than half developing complications such as calcinosis and lipodystrophy [13]. While a family history of psoriasis is present in 13% of cases of juvenile dermatomyositis, the actual co-occurrence of JDM and psoriasis is exceedingly rare, with only scattered case reports documented in the literature [3,4,14]. Recent population-based studies have demonstrated an increased risk of dermatomyositis in patients with psoriasis (hazard ratio: 2.41, 95% CI: 2.01-2.89), further supporting the epidemiological association between these two conditions [5,7,15]. We report a case of juvenile dermatomyositis with severe cutaneous complications co-occurring with psoriasis vulgaris.

CASE PRESENTATION

An 8-year-old girl, resident of West Bengal, India, initially presented to another hospital with symmetrical proximal weakness of all four limbs and cutaneous lesions over the trunk. She received intravenous methylprednisolone pulse therapy, which resulted in improvement of muscle weakness. However, she was subsequently lost to follow-up. At 18 years of age, she presented to our institution with recurrent symmetrical proximal muscle weakness involving all four limbs. Manual Muscle Testing (MMT-8) revealed a score of 64/80, indicating moderate muscle weakness. Physical examination demonstrated extensive dermatological manifestations, including calcinosis universalis over the right side of the neck (Figure 1a), right flank, lower abdomen, both thighs, right popliteal fossa with associated right knee contracture and lipodystrophy over the right cheek; multiple erythematous scaly plaques over the scalp(Figure 1a), abdomen (Figure 1b), and back (Figure 1d); multiple hypopigmented macules scattered over the back demonstrating Koebner's phenomenon over the lower back (Figure 1d); and cutaneous ulcers with crusting over both elbows and legs.

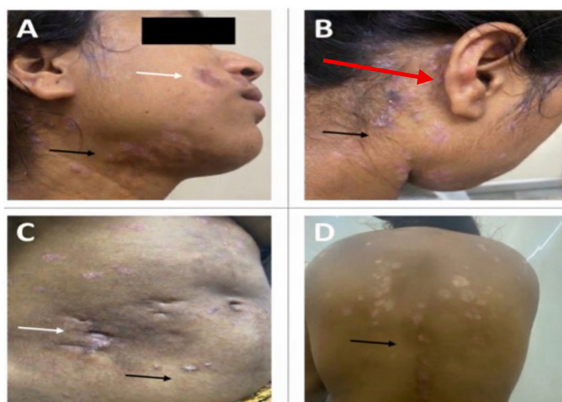

Figure 1

Figure 1: Clinical presentation. (a) The white arrow shows Lipodystrophy over the right cheek, and the black arrow shows calcinosis over the neck. The black arrow shows Psoriasis vulgaris lesions over the scalp. (c) The black arrow shows Psoriasis vulgaris lesions, and the white arrow shows calcinosis over the trunk. (d) The black arrow shows Koebner's phenomenon over the lower back

Investigations

Laboratory investigations revealed the following:

Parameter	Value	Reference Range
Creatine kinase	268 U/L	26-192 U/L
Lactate dehydrogenase	174 U/L	135-214 U/L
Aspartate transaminase	30 U/L	0-40 U/L
Alanine transaminase	28 U/L	0-40 U/L

Table 1

Table 1: Laboratory parameters at presentation

Myositis-specific antibody profile demonstrated strong positivity to anti-NXP2 antibodies. Anti-nuclear antibody (ANA) and extended autoantibody profile by line immunoassay were negative. Serum complement levels (C3, C4) were within normal limits. High-resolution computed tomography (HRCT) of the chest revealed no evidence of interstitial lung disease. Histopathological examination of the scaly cutaneous lesions from the trunk demonstrated features consistent with psoriasis vulgaris, including parakeratosis, dermal edema, perivascular inflammatory infiltrate, and Munro's microabscesses.

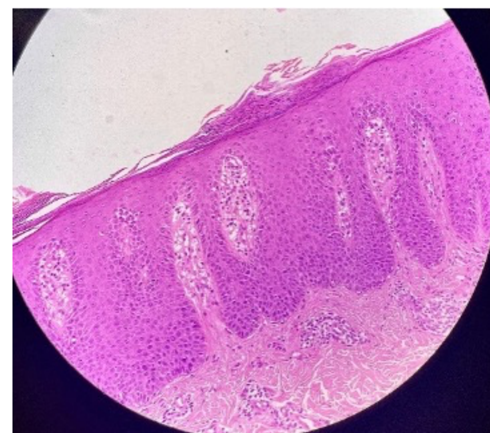

Figure. 2

Figure 2: Histopathological examination of scaly cutaneous lesions from the trunk. Red arrow shows parakeratosis, the Orange arrow shows dermal edema, the green arrow shows perivascular infiltrate, and the black arrow Munro's microabscess, consistent with psoriasis vulgaris

Treatment and outcome

In view of severe cutaneous manifestations and progressive muscle weakness, the patient was treated with rituximab 1 gram intravenous infusion. Notably, improvement in the psoriatic lesions over the neck, abdomen, and trunk was observed secondary to intravenous methylprednisolone, which was administered as premedication prior to rituximab therapy. She was discharged on oral prednisolone 0.5 mg/kg/day and methotrexate 0.3 mg/kg/week.

On follow-up evaluation over the past year, the patient has not developed new skin lesions, and muscle strength has improved significantly, with no progression of calcinosis.

DISCUSSION

In searching the literature for cases of coexisting juvenile dermatomyositis and psoriasis, we found very limited data in the form of scattered case reports [3,4,14]. Kim et al. detailed the clinical trajectories of three pediatric female patients exhibiting a rare overlap of juvenile dermatomyositis and psoriasis [3]. The first patient, an 18-year-old with a 10-year disease duration, developed psoriasis subsequent to JDM diagnosis. While her classical JDM manifestations—proximal muscle weakness and heliotrope rash—achieved remission on intravenous methylprednisolone and oral prednisone, her psoriatic lesions demonstrated only partial response to topical corticosteroids and methotrexate. Conversely, an 8-year-old female presented with a reversed sequence, developing JDM one year after her initial psoriasis diagnosis. Exhibiting similar JDM symptomatology, she showed minimal initial response to methotrexate; however, her JDM ultimately improved following an aggressive, multi-agent regimen comprising oral and intravenous corticosteroids, subcutaneous methotrexate, etanercept, mycophenolate mofetil, and cyclosporine [3]. Our patient's presentation shares similarities with the first case reported by Kim et al., with psoriasis developing after established JDM diagnosis. However, our case is distinguished by the presence of severe cutaneous complications of JDM, including extensive calcinosis universalis and lipodystrophy, along with positive anti-NXP2 antibodies. Anti-NXP2 antibodies are associated with increased risk of calcinosis in JDM, particularly in patients with early disease onset before 5 years of age, and are predictive of worse disease outcomes [16].

Highlighting the complexities of managing overlapping conditions, Perna et al. reported a 20-year-old female whose corticosteroid-responsive JDM was destabilized by psoriasis treatment [4]. Following failed trials of topical agents and prednisolone for her psoriasis, the introduction of secukinumab (an IL-17A inhibitor) and methotrexate led to significant worsening of her underlying JDM, necessitating discontinuation of secukinumab [4]. In contrast to this report, our patient demonstrated improvement in both psoriatic lesions and JDM manifestations. The improvement in psoriasis was initially noted with methylprednisolone premedication, with sustained benefit following methotrexate therapy. This observation underscores the variability in treatment responses and the importance of careful agent selection in managing this dual pathology.

Immunopathological considerations

JDM and psoriasis share overlapping immune cascades driven by plasmacytoid dendritic cell (pDC) activation, elevated cutaneous IL-17, and cytokines including interferons (IFNs), TNF-α, and IL-23 [17,18]. However, they exhibit distinct local immunological signatures. JDM features prominent IFN-γ-inducible chemokines and muscle/mononuclear cell TNF-α overexpression [20], whereas psoriasis is primarily driven by IFN-α and the IL-23/IL-17 axis mediating keratinocyte hyperproliferation [18]. These overlapping yet distinct pathogenic mechanisms create therapeutic challenges. While agents such as methotrexate and cyclosporine demonstrate efficacy for both conditions, targeted psoriasis treatments including phototherapy and TNF-α inhibitors can paradoxically trigger or exacerbate underlying dermatomyositis [5,6]. Similarly, TNF-α inhibitors have been associated with dermatomyositis induction or exacerbation, possibly through increased type I interferon production, which plays a central role in dermatomyositis pathogenesis [6]. Our patient demonstrated clearing of psoriatic lesions with methylprednisolone followed by sustained improvement with methotrexate,

supporting the utility of agents with dual efficacy in managing this overlap syndrome [18].

Rituximab in JDM-psoriasis overlap

Montoya et al. reported the occurrence of erythrodermic psoriasis following rituximab use in a case of amyopathic JDM coexisting with psoriasis [14].

In our case, rituximab was well tolerated without exacerbation of psoriatic lesions. Previous studies have demonstrated significant improvement in cutaneous disease activity in refractory adult dermatomyositis and JDM following rituximab administration, with particular benefit observed in erythroderma, erythematous rashes, heliotrope rash, and Gottron's papules [19,20]. Our patient's favorable response to rituximab, with resolution of both muscle weakness and psoriatic lesions, supports its potential utility in carefully selected cases of JDM-psoriasis overlap.

Unique features of the present case

While the coexistence of juvenile dermatomyositis and psoriasis has been reported globally, our case is distinctive in several aspects. The presence of severe cutaneous complications of JDM, including extensive calcinosis universalis and lipodystrophy, distinguishes this presentation. Positive anti-NXP2 antibody status is associated with increased risk of calcinosis and poor disease outcomes [16]. Our patient demonstrated favorable response to rituximab without exacerbation of psoriatic manifestations. There was a long delay between initial JDM diagnosis and development of psoriasis (10-year interval). Finally, the patient achieved successful disease control with combination therapy avoiding agents known to exacerbate either condition.

CONCLUSION

The coexistence of juvenile dermatomyositis and psoriasis represents a rare and therapeutically challenging clinical entity. Our case is notable for the presence of severe cutaneous complications of JDM, including extensive calcinosis universalis, lipodystrophy, and positive anti-NXP2 antibody status. Careful selection of immunosuppressive agents that do not aggravate either condition is essential for achieving optimal therapeutic outcomes.

Learning points

The co-occurrence of juvenile dermatomyositis and psoriasis is extremely rare but represents a significant diagnostic and therapeutic challenge requiring multidisciplinary expertise. Clinicians managing patients with JDM-psoriasis overlap must exercise caution in selecting immunosuppressive agents, avoiding therapies that may exacerbate either condition, particularly phototherapy and TNF-α inhibitors. Anti-NXP2 antibody positivity in JDM is associated with increased risk of calcinosis development and worse disease outcomes, necessitating aggressive early treatment and close monitoring. Combination therapy with corticosteroids, methotrexate, and rituximab can be effective in managing both JDM and psoriatic manifestations, although careful monitoring for adverse effects is essential. Long-term follow-up is critical in patients with JDM, as late-onset complications including calcinosis and coexisting autoimmune conditions may develop years after initial diagnosis.

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