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Case Report

Dermatomyositis sine dermatitis-an unusual non-classical dermatomyositis with amyopathic myopathy tale: a case report

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ABSTRACT

We report a 13-year-old girl with dermatomyositis sine dermatitis (DMSD), a rare form of juvenile DM characterized by muscle weakness without skin rash. She presented with 2 months of progressive proximal limb weakness, neck weakness, and bilateral hand paresthesias, but no skin findings. Laboratory tests showed elevated inflammation with normal creatine kinase and significant hypokalemia. Myositis autoantibody testing revealed strongly positive anti-NXP2 and anti-Ku antibodies. A diagnosis of DMSD was made. High-dose corticosteroids plus supportive care and physiotherapy led to rapid recovery. This case highlights that DM should be considered even without rash. Anti-NXP2 positivity, associated with the sine-dermatitis phenotype, aided in confirming the diagnosis. Early recognition and treatment were crucial to the good outcome.

Keywords: Dermatomyositis sine dermatitis, Juvenile dermatomyositis, Anti-NXP2 antibody, Amyopathic dermatomyositis, Hypokalemia

INTRODUCTION

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy characterized by symmetric proximal muscle weakness and classic cutaneous findings (Gottron's papules, heliotrope rash).[1] It affects both adults and children and may involve other systems (e.g., lung, myocardium).[2] Adult DM is well-known to associate with malignancy in ~15-30% of cases.[3] DM is classified into subtypes: classical DM (muscle + skin), amyopathic DM (skin involvement only), and DMSD, in which muscle weakness occurs without rash.[4] Although DMSD is included in consensus criteria, it is extremely under-recognized.[5] Recent studies suggest DMSD comprises only ~8% of biopsy-confirmed DM cases.[6] Importantly, DMSD has been closely linked to anti-nuclear matrix protein 2 (anti-NXP2) autoantibodies.[7] Juvenile DM (JDM) is itself rare (incidence ≈2-4 per million) and can differ from adult DM.[8] Myositis-specific autoantibodies are now used to refine diagnosis and prognosis in JDM. We present a pediatric case of DMSD confirmed by anti-NXP2, and review its clinical features, highlighting the diagnostic challenges and management implications of this unusual myositis subtype.

CASE REPORT

A 13-year-old girl with no significant medical history developed progressive muscle weakness over 2 months. Initially she had difficulty rising from a squat and climbing stairs, indicating proximal lower limb weakness. Over weeks, she noted neck flexor weakness and mild tingling in both hands. She denied fever, rash, joint pain, dysphagia, or weight loss. One year earlier she had a transient viral myositis after a febrile illness, from which she fully recovered. Family history was non-contributory.

On exam, she was alert and afebrile. Muscle power was symmetrically reduced: medical research council (MRC) grade ~4/5 in shoulder and hip girdles and ~3/5 in knee

extensors. Distal strength was preserved. Neck flexion was 3/5. Deep tendon reflexes were diminished (1-2/4) throughout; plantar responses were flexor. Sensation to light touch and pinprick was normal except for mild decreased pinprick in both hands (she complained of intermittent tingling). Critically, there were no skin abnormalities (no heliotrope rash, Gottron papules, shawl sign, mechanic's hands, or other rash). Examination of joints, lungs, heart, and abdomen was unremarkable.

Laboratory tests showed an erythrocyte sedimentation rate of 65 mm/hr (elevated) and C-reactive protein-10 mg/L (slightly high). Serum creatine kinase (CK) was 90 U/L (normal range), indicating normal muscle enzyme. Serum potassium was 2.6 mmol/L (marked hypokalemia); magnesium and calcium were normal. Complete blood count and metabolic panel were otherwise normal. Given the myositis picture, a myositis antibody panel was sent. Antinuclear antibody (ANA) and anti-Jo-1 were negative; however, anti-nuclear matrix protein 2 (anti-NXP2) IgG was strongly positive (titer 1:320) and anti-Ku IgG was positive. Anti-MDA5, anti-Mi-2, anti-SRP, and other myositis antibodies were negative. A chest computed tomography scan showed no interstitial lung disease or malignancy. (Malignancy screens, including abdominal imaging and tumor markers, were also negative.)

These findings confirmed a diagnosis of DMSM. We initiated oral prednisone 40 mg daily (high-dose corticosteroids) and supportive care (correction of hypokalemia, vitamin D/calcium supplements, pantoprazole, amitriptyline, and tramadol for symptom relief) along with intensive physiotherapy. Over the next 3-4 weeks, the patient improved markedly: the paresthesias resolved, and proximal muscle strength recovered to MRC 5/5 in the arms and ~4+/5 in the legs. She tolerated therapy well. The plan is to gradually taper steroids and consider a steroid-sparing agent (e. g., methotrexate) if needed. At discharge, she remained alert with no skin findings and normal vital signs.

DISCUSSION

DMSD is an uncommon but important variant of DM. Our patient's presentation-proximal myopathy without rash. This phenotype is rarely recognized clinically. Anti-NXP2 autoantibodies were instrumental in our case, as they have been strongly associated with sine-dermatitis DM. In contrast, anti-MDA5 (associated with amyopathic DM) was absent. In juvenile DM, anti-NXP2 is found in ~23-30% overall and often correlates with severe myositis; in one study 86% of DM patients without rash were anti-NXP2 positive (versus 28% with rash.[9] Thus, the serology supported our diagnosis. Our patient also had anti-Ku positivity, which is unusual for DM. Anti-Ku typically appears in overlap syndromes (e. g., scleroderma-polymyositis overlap) and its significance here is unclear; it may simply mark a broader autoimmune tendency. Importantly, anti-NXP2 positivity predicts calcinosis and more severe course in JDM, so we will monitor for calcinosis (which was not present initially).[10,15]

Remarkably, our patient's CK was normal despite active disease-a phenomenon reported in other DMSD cases. Park et al similarly described a DMSD adult with CK-86 U/L. Thus, normal muscle enzymes cannot exclude active myositis in this context. The only significant lab abnormality besides antibodies was hypokalemia.[1] Hypokalemia is not typical of DM; it may have been multifactorial (e.g. urinary losses or intracellular shift in acute myositis) and it corrected promptly with steroids and supplements.[12] A prior case report even described idiopathic DM presenting with significant hypokalemia, underscoring diagnostic complexity. In practice, we simply corrected it.

Therapy followed standard DM guidelines. High-dose glucocorticoids yielded rapid improvement. This parallels prior reports: Park's patient with DMSD improved with prednisone over month. Other juvenile DM series also show good steroid response. We did not immediately add a second-line agent, but methotrexate would be considered if she shows relapse or steroid toxicity. Hydroxychloroquine was not needed since there were no skin lesions.[13]

Malignancy is a concern in DM, though much rarer in children. Adult DMSD cases have indeed had cancers reported. For example, Park et al noted a 77-year-old DMSD patient later diagnosed with kidney cancer. Pediatric DM carries much lower malignancy risk, but given these associations, we performed age-appropriate screening (imaging and lab tests), all of which were negative (mirroring Park's report of no malignancy or ILD).[14] Nonetheless, we plan periodic surveillance (e.g. urinalysis, ultrasound) as a precaution.[15]

In summary, this case emphasizes that DM may present without rash-even in children-and highlights the diagnostic utility of autoantibodies. The presence of anti-NXP2 confirmed DMSD in our patient, guiding therapy.[16,20] Early diagnosis and treatment led to an excellent outcome. We recommend that clinicians consider DMSD when a child has unexplained myositis symptoms.[24] Long-term follow-up is needed to watch for complications (calcinosis, relapse, rare malignancy). Further study of juvenile DMSD cases will improve understanding of prognosis and optimal management of this rare myositis subtype.[17]

Results and future scope

In this case, the patient's outcomes were very good: muscle strength and sensory function returned to normal with therapy, and she remains fully ambulatory. No corticosteroid side effects or organ damage occurred. We will taper steroids gradually and plan to start a low-dose DMARD (such as methotrexate) if any signs of relapse

emerge.[18] For future care, the patient will have regular follow-ups for at least several years, focusing on muscle strength, range of motion, and surveillance for calcinosis (via physical exam/X-ray) and malignancy (age-appropriate screening).[19]

Looking ahead, more data on DMSD particularly in children are needed. As more centers use extended myositis antibody panels, additional cases may be identified earlier. Collaborative registries could track pediatric DMSD patients to assess relapse rates, long-term disability, and cancer occurrence.[21,25] Research into targeted therapies (e. g., biologics or JAK inhibitors) may benefit patients with refractory disease. In our patient, however, the standard steroid-based regimen was sufficient. Future investigations should clarify whether anti-NXP2 status should alter management (for example, prompting prophylactic therapies for calcinosis or earlier steroid-sparing treatment).[22,23]

CONCLUSION

This report illustrates that DM can manifest without skin involvement in a child, and underscores the importance of a comprehensive diagnostic approach. Myositis-specific antibody testing was critical for establishing the diagnosis of DMSD. Prompt immunosuppressive therapy led to rapid recovery. Clinicians should maintain a high suspicion for DM variants when evaluating pediatric myopathy, even in the absence of rash. Continued follow-up is essential to detect complications. This case adds to the limited pediatric literature on DMSD and highlights key points for diagnosis and management of this rare myositis variant.

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