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Radio-Diagnosis

SEEING THROUGH SKIN: HOW ADVANCED MRI TRANSFORMS DERMATOMYOSITIS DIAGNOSIS

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ABSTRACT Juvenile dermatomyositis (JDM) is a rare systemic autoimmune connective tissue disorder marked by distinctive cutaneous manifestations and progressive proximal muscle weakness. While the Bohan and Peter criteria historically guided diagnosis, magnetic resonance imaging (MRI) has emerged as a non-invasive alternative that is increasingly supplanting muscle biopsy for both confirmation and ongoing monitoring of JDM. Three pediatric patients, aged 4 to 8 years, presented with characteristic cutaneous findings and muscle weakness and underwent whole-body MRI (WBMRI). These cases were selected for their representative clinical features and the diagnostic complexity associated with juvenile dermatomyositis (JDM). Each patient exhibited distinct clinical manifestations, all confirmed by established JDM diagnostic criteria. American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) scores ranged from 7 to 9, reflecting the variability and severity of symptoms in pediatric populations. WBMRI with short tau inversion recovery (STIR) sequences enabled comprehensive assessment, revealing symmetric muscle involvement with T2/STIR hyperintensity in proximal muscles, subcutaneous fat reticulation, and fascial involvement. One patient demonstrated large calcifications on WBMRI, while the other cases showed diffuse inflammation and perivascular enhancement in the pelvis and thighs. WBMRI findings correlated with clinical severity, elevated muscle enzyme levels, and positive autoimmune markers, underscoring its value as a non-invasive diagnostic modality. In summary, WBMRI is an essential non-invasive modality for diagnosing juvenile dermatomyositis (JDM). Early detection of subclinical involvement enables objective assessment of disease activity, facilitating timely diagnosis and effective treatment strategies. This approach contributes to improved long-term patient outcomes.

KEYWORDS : Whole-body MRI, Magnetic Resonance Imaging, Juvenile Dermatomyositis, Dermatomyositis, STIR WB MRI

INTRODUCTION

Juvenile dermatomyositis (JDM) is a rare autoimmune disorder of connective tissue, accounting for approximately 85% of idiopathic inflammatory myopathies in children, with an incidence of 2 to 4 cases per million children annually.[2] The condition is more prevalent in females and demonstrates bimodal age peaks at 5 to 14 years and 45 to 64 years. Principal symptoms include symmetrical, progressive muscle weakness and distinctive cutaneous changes, such as a violaceous rash on the eyelids and Gottron's papules on the finger joints. JDM may also involve the gastrointestinal, cardiovascular, respiratory, renal, and ocular systems. Late complications, including lipodystrophy and calcinosis, have become rare due to early intervention.[3-5]

Historically, the diagnosis of juvenile dermatomyositis (JDM) was based on the Bohan and Peter criteria established in 1975, which require characteristic cutaneous manifestations in addition to three of four further diagnostic features.[6]

Magnetic resonance imaging (MRI) detects alterations in signal intensity within the skin, subcutaneous tissue, fascia, and muscles, enabling quantitative assessment of subtle inflammation that may not be apparent on physical examination.

The adoption of whole-body MRI (WB MRI), particularly STIR WBMRI (Short tau inversion recovery Whole Body Magnetic Resonance Imaging) represents a significant advancement in the assessment of JDM. This technique enables comprehensive evaluation of the entire muscular system and addresses the limitations of traditional thigh-only MRI protocols, which may miss scapular girdle involvement in up to 30% of patients.[7] Early detection through WBMRI guides therapeutic decisions and provides objective measures for monitoring treatment response.

This case series describes three pediatric patients with juvenile dermatomyositis and their imaging findings on MRI. Inclusion criteria comprised characteristic dermatological features and muscle weakness, confirmed by JDM diagnostic criteria. Exclusion criteria included incomplete medical records or overlapping autoimmune conditions. The primary objective is to demonstrate the diagnostic utility of STIR WB-MRI as a comprehensive modality for detecting muscular and extra-muscular manifestations, as well as for disease assessment and monitoring.

Case Description

Case One

An 8-year-old girl had worsening thigh muscle weakness over two years, with a marked decline in the past 1.5 months. She struggled to rise from sitting and could not climb stairs. She also had scaly lesions on finger joints, a red rash on the right cheek, and firm swellings in the axilla. Family history was unremarkable.

She met the Bohan and Peter criteria for dermatomyositis. She had high muscle enzymes and myopathic changes on EMG. Her American College of Rheumatology European League Against Rheumatism score (ACR-EULAR) score was 7, indicating probable idiopathic inflammatory myopathy.

The physical examination showed discrete scalp alopecia, right cheek erythema with dilated vessels, a firm 1×2 cm axillary lesion with calcinosis, violaceous scaly Gottron's papules on the fingers, and cuticular/nail changes. Muscle strength was symmetrically reduced to 3/5 at the hips. Muscle bulk, tone, reflexes, neurological exam, joints, and other systems were unremarkable.

Laboratory tests showed elevated CPK, LDH, and SGOT, mild anemia, and positive AMA-M2. Other autoimmune, metabolic, and endocrine markers were normal.

Nerve conduction was normal. EMG revealed myopathic changes.

Radiological Findings

Plain radiography of the hips and thighs showed diffuse nodular and sheet-like calcifications in the soft tissues of the hip muscles and lateral distal thighs (Figure 1 A&B).

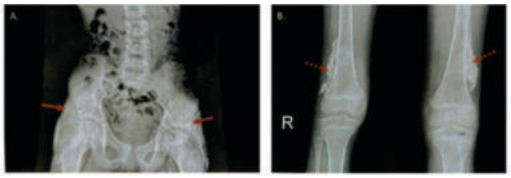

Figure 1: Xray abdomen (A) and bilateral thigh (B) AP view shows, diffuse nodular, and sheet like calcification seen in the soft tissue of the muscle of bilateral hip (red arrow in A) and bilateral distal thigh on lateral aspect (dotted red arrow in B).

Chest X-ray revealed soft tissue calcifications in the chest wall and axillae; lungs were clear.

MRI of the thighs evaluated the superficial tissues, muscles, myofascial changes, and calcifications. It showed T2/STIR hyperintensity and subtle post-contrast enhancement in the skin and subcutaneous fat of the posterior mid and distal thighs, predominantly on the left side. Nodular signal alterations appeared in the vastus lateralis and vastus intermedius muscles on both sides. (Figure 2 A-E) These were hypointense on T1 and hypointense centers with hyperintense rims on T2/STIR. Peripheral rims post-contrast enhancement on T1 fat-suppressed images. Blooming artifacts on gradient-echo confirmed calcifications. Patchy hyperintense areas were observed in the remaining vastus lateralis and hamstring muscles on T2/STIR, which showed post-contrast enhancement.

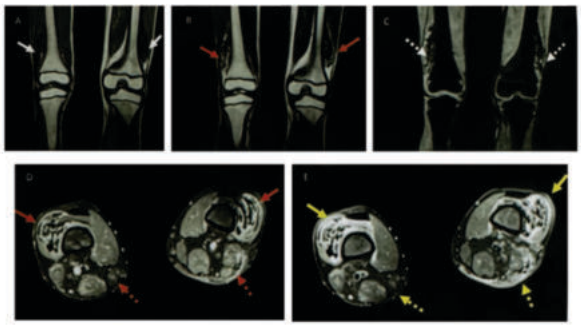

Figure 2 (A-E): Coronal T1W, T2W and medic sequence with axial PDFS, T1WFS and post contrast image of same patient at the level of distal thigh show the calcification in the intermuscular plane of bilateral vastus lateralis and intermedius muscle appearing hypotension and blooming on medic sequence. They appear hypo intense on T2 W oblique and PDF S with peripheral hyperintense rim (red arrow in B&D) and post contrast enhancement of the peripheral intense rim (yellow arrow in E). Reticulation in the subcutaneous fat in bilateral posterior leg left more than right and showing hyperintensity on PDFS image (dotted arrow in D) with post contrast enhancement is also noted (dotted yellow arrow in E).

Treatment

The patient was started on oral prednisolone, methotrexate, hydroxychloroquine, topical corticosteroids, and sunscreen, with laboratory and clinical monitoring.

Case Two

The patient received a probable dermatomyositis diagnosis based on Bohan and Peter classification criteria, demonstrating characteristic cutaneous manifestations, proximal symmetric muscle weakness, and elevated skeletal muscle enzymes. According to the ACR-EULAR criteria for idiopathic inflammatory myopathy, the patient achieved a score of 9, corresponding to definite classification.

The exam showed discrete scalp alopecia, confluent facial malar erythema showing exacerbation upon sun exposure, violaceous scaly Gottron's papules on the fingers, and periorbital puffiness and bilateral lower extremity pitting edema. Muscle strength was symmetrically reduced to 2/5 at the hips and (3/5) in the upper extremity. There was tenderness in the bilateral thigh and knee joints. Muscle bulk, tone, reflexes, neurological exam, and other systems were unremarkable.

Laboratory tests showed elevated CPK, LDH, and SGOT, mild anemia, and positive Jo-1 antibodies. Other autoimmune, metabolic, and endocrine markers were normal.

Nerve Conduction and EMG were Normal.

Radiological Findings

Magnetic resonance imaging of bilateral thighs was performed to assess oedema and inflammation of muscle, myofascial, and subcutaneous fat involvement. It demonstrated bulky thigh muscles with bilateral intramuscular T2/STIR hyperintense areas involving anterior and medial compartments, exhibiting patchy post-contrast enhancement predominantly in perivascular distributions. Additional findings included patchy, discrete hyperintense areas in bilateral adductor and hamstring muscle groups against a background of otherwise normal muscle signal intensity (Figure 3A-D). Imaging also revealed prominent reticulation appearing as T2/STIR hyperintensity within subcutaneous fat at the pelvic level posteriorly and posteromedial upper thigh compartments, which showed post-contrast enhancement. Bilateral symmetric intramuscular diffuse T2-weighted/STIR hyperintensity was identified in pelvic musculature, including gluteus maximus, obturator externus and internus, and pectineus muscles, all showing post-contrast enhancement.

Treatment

The patient was started on oral prednisolone, methotrexate, hydroxychloroquine, topical corticosteroids, and sunscreen, with laboratory and clinical monitoring

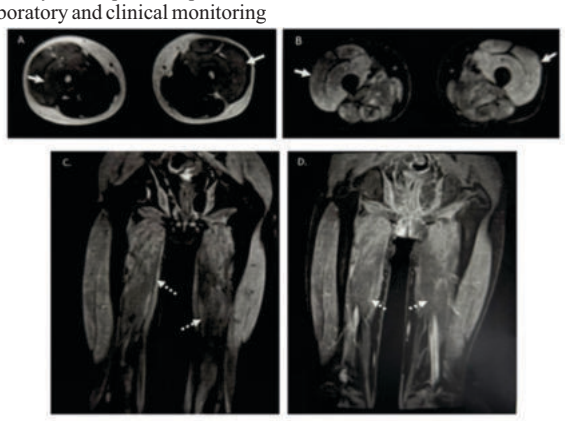

Figure 3(A-D): Axial T2W (A) and STIR (B)image at the level of mid-thigh show bulky muscles with diffuse T2/STIR hyperintensity in the rectus femoris, vastus medialis, intermedius and lateralis muscle (arrows in A&B). Patchy discrete area of hyper intensity is seen in bilateral adductor and the hamstring muscles in the background of normal muscle signal intensity.

Coronal STIR WBMRI (C) and T1WFS post contrast(D) show diffuse hyperintensity with post contrast enhancement of vastus medialis with patchy hyper intensity and post contrast enhancement of adductor muscle (dashed arrows in C &D) predominantly in perivascular location.

Case Three

A 4-year-old girl presented with an 8-month history of bilateral lower extremity weakness and pain involving the hips and knee joints, accompanied by violaceous rash affecting bilateral upper and lower limbs. One month after initial symptom onset, she developed difficulty climbing stairs and rising from seated positions.

The exam showed confluent facial malar erythema, which worsens with sun exposure, and violaceous scaly Gottron's papules on the dorsal surfaces of bilateral upper and lower extremities. Muscle strength was symmetrically reduced to 2/5 at the hips and (3/5) in the upper extremity. There was tenderness in the bilateral thigh and knee joints. Muscle bulk, tone, reflexes, neurological exam, and other systems were unremarkable.

Laboratory tests showed elevated CPK, LDH, and SGOT, mild anemia, and positive Jo-1 antibodies, AMA-M2 antibodies, TIF-1 gamma, and NXP2. Other autoimmune, metabolic, and endocrine markers were normal.

Nerve Conduction and EMG were Normal.

Imaging Findings

Plain radiographic examination was normal. STIR whole-body magnetic resonance imaging (STIR WB-MRI) (Figure 4 A-D) revealed inflammatory changes as diffuse increased signal intensity within the muscles, superficial and deep fascia, skin, and subcutaneous tissue.

Treatment

The patient was started on oral prednisolone, methotrexate, hydroxychloroquine, topical corticosteroids, and sunscreen, with laboratory and clinical monitoring.

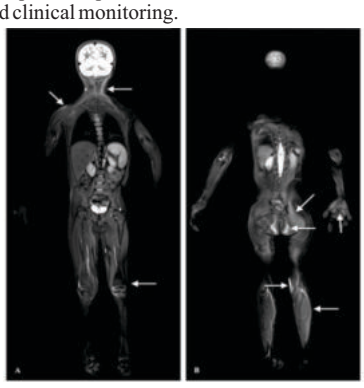

Figure 4 (A&B): Coronal STIR WBMRI image show diffuse distribution of muscle inflammation in the form of hyperintensity in bilateral gluteus muscles (*) thigh muscles (white arrow) and visualized calf muscles (white small arrow). Patchy discrete area of hyper intensity is seen in neck and left palm muscles (arrows). Mild patchy hyperintensity is seen in visualized skin, superficial fascia and intermuscular fascia.

DISCUSSION

Juvenile dermatomyositis (JDM) presents with a more acute onset than adult dermatomyositis, characterized by earlier skin manifestations followed by progressive muscle weakness and inflammation. This rare autoimmune disorder affects approximately 1 per 100,000 individuals annually, with a female predominance of 2:1 and a bimodal age distribution peaking at 5-14 years and 45-64 years. JDM demonstrates distinctive features, including increased propensity for calcification in muscles, skin, and subcutaneous tissues, where calcium deposits are characteristically firm and may lead to contractures and localized atrophy.

The pathognomonic cutaneous manifestations typically precede muscle weakness and include heliotrope rash, Gottron's papules over the knuckles, elbows, and knees, the V-sign, and the shawl sign. Less common features encompass calcinosis cutis, periungual telangiectasias, and poikiloderma. Progressive proximal muscle weakness affects both the upper and lower extremities, with relative sparing of the trunk, accompanied by muscle tenderness and stiffness. Systemic complications may involve cardiovascular, pulmonary (including interstitial lung disease), and gastrointestinal systems.

The etiopathogenesis involves humoral immunity-mediated vasculopathy, where specific autoantibodies target the endomysial capillary endothelium, activating the complement cascade and leading to the deposition of the membrane attack complex. This process results in endothelial swelling, capillary necrosis, and perivascular inflammation, leading to muscle ischemia and characteristic perifascicular atrophy. Additional pathogenic mechanisms include genetic associations with specific HLA types (B8, DR3, DQA1) and TNF-α gene polymorphisms, as well as infectious triggers through molecular mimicry involving influenza, parainfluenza, hepatitis B, and group A streptococci.

Diagnostic criteria have evolved from the original Bohan and Peter criteria to the 2017 EULAR/ACR classification system, which incorporates age of onset, muscle weakness patterns, cutaneous manifestations, laboratory parameters (including anti-Jo1 positivity, elevated CK/LDH, and AST/ALT), and muscle biopsy features. Patients are classified based on cumulative scores as definite Idiopathic Inflammatory Myositis (IIM) (≥90%), probable IIM (55-89%), possible IIM (50-54%), or not IIM (<50%). The 2006 revised JDM criteria, as outlined by Brown et al., emphasized the clinical utility of MRI findings and muscle biopsy as key diagnostic tools, alongside muscle weakness, skin rash, and elevated enzymes.

Imaging plays a crucial role in JDM assessment across multiple modalities. Conventional radiography serves as a baseline investigation, showing soft tissue thickening and poor muscle-subcutaneous tissue delineation in the acute phase, progressing to characteristic calcification patterns in the chronic stage: superficial plaques, deep nodular deposits, fascial plane involvement, and extensive surface calcium deposition. Computed tomography demonstrates superior sensitivity for detecting deeper calcifications and ossifying lesions compared to radiography.

Magnetic resonance imaging with high resolution and multiplanar capability offers optimal soft tissue characterization and excellent sensitivity for inflammatory changes. The study by Kaufman et al.[8] was one of the earliest to assess the role of MRI for JDM. MRI guides optimal biopsy site selection, preferentially targeting areas with muscle edema and minimal fatty infiltration. The modality effectively monitors disease activity, treatment efficacy, and longitudinal progression. Characteristic MRI findings include bilateral symmetric muscle involvement with a predilection for the pelvic girdle and thigh muscles. The various patterns of T2W/STIR hyperintensity are diffuse, patchy, peripheral, and reticular. Subcutaneous fat demonstrates reticular involvement due to perivascular inflammation, while dystrophic calcification appears hypointense with a hyperintense peripheral rim and enhancement. Collectively, these patterns highlight the crucial role of MRI in assessing disease severity and informing clinical decisions, thereby bridging the gap between empirical data and practical interpretation in the management of JDM. In imaging, muscle inflammation appears hyperintense on fluid-sensitive sequences such as T2-weighted or short tau inversion recovery (STIR) scans, while unaffected muscles appear hypointense. To enhance the visibility of abnormalities, two main fat-suppression methods are used: T2-weighted images with fat suppression (T2W/FS) and STIR, which uses a long echo time to suppress fat. T1-weighted images are better for showing fat replacement in chronic cases, indicating chronicity and tissue damage. MRI also helps select the best location for a muscle biopsy if needed. This improves diagnostic accuracy, especially since inflammation can be patchy and biopsies alone may be less reliable.

Whole-body MRI (WB-MRI) represents a paradigm shift in comprehensive JDM assessment, providing a sensitive and non-invasive evaluation of the total inflammatory burden throughout the musculoskeletal system. STIR WBMRI sequences effectively detect disease activity across the entire body, identifying previously unsuspected involvement sites and enabling assessment of extra-muscular manifestations, including interstitial lung disease and systemic malignancy. [9,10,11]This technique facilitates baseline documentation, treatment monitoring, and screening for steroid-induced complications such as osteonecrosis. A contrast study is not required in the evaluation of muscle disease, as it is well demonstrated on STIR images. Yoshipovitch et al. described three adult patients with dermatomyositis (DM) in whom STIR MRI was used to establish the diagnosis and for follow-up.[7]

WBMRI demonstrates superior diagnostic capability compared to conventional approaches, revealing muscle groups that are not captured by standard protocols and providing a comprehensive assessment of inflammatory burden. The technique demonstrates a strong correlation with clinical activity measures, including Manual Muscle Testing (MMT), Disease Activity Score (DAS), Childhood Myositis Assessment Score (CMAS), and muscle enzyme elevation. Muscle biopsies from MRI-positive sites contain significantly more inflammatory cells than those from unaffected areas. STIR sequences prove particularly valuable, potentially eliminating the need for muscle biopsy when demonstrating clear inflammatory changes. MRI was considered an important diagnostic criterion and, along with muscle biopsy, was rated the most clinically useful investigative criterion for JDM by 78 respondents in a survey of members of the Network for JDM and the Pediatric Rheumatology International Trials organization.[8]

Differential diagnosis considerations include necrotizing fasciitis, viral myositis, pyomyositis, myositis ossificans, metabolic myopathies, medication-related myositis, and other rheumatologic conditions. The characteristic bilateral and symmetric proximal appendicular distribution distinguishes JDM from isolated muscle involvement patterns. Whole-body MRI (WBMRI) findings can help differentiate JDM from these conditions by highlighting the unique imaging features of each disorder. For instance, necrotizing fasciitis typically presents with diffuse subcutaneous edema and fascial enhancement, while viral myositis often shows more focal muscle involvement without the calcification seen in JDM. Pyomyositis is usually characterized by muscle abscess formation, often lacking the diffuse inflammation and symmetric distribution found in JDM. Myositis ossificans will typically demonstrate calcified masses in the subacute phase, contrasting with the sheet-like calcifications seen in JDM. Metabolic myopathies might show lipid accumulation in muscle on MRI, and medication-related myositis often lacks the inflammatory changes visible in JDM. Comparing these imaging characteristics is crucial for clinicians in making accurate diagnoses.

Treatment typically involves first-line oral corticosteroids with gradual tapering over 10-12 months. Refractory cases require adjuvant immunosuppressive agents, including methotrexate, azathioprine, or cyclophosphamide. Established calcinosis remains challenging to treat despite high-dose prednisolone and methotrexate therapy.

CONCLUSION

Following treatment, patients were closely monitored to evaluate their responses to imaging-guided management strategies. Imaging outcomes demonstrated significant improvements in muscle signal intensity, indicating reduced inflammation. Follow-up assessments revealed that all three cases experienced improved muscle strength and reduced skin rashes. The patient with extensive calcifications exhibited a marked reduction in calcific deposits. These findings highlight the importance of early imaging and targeted therapy in optimizing patient outcomes. Imaging monitoring enables assessment of treatment response through normalization of muscle signal intensity and supports prediction of clinical outcomes at diagnosis, thereby guiding therapeutic intensity and improving long-term management.

Abbreviations

JDM.	Juvenile dermatomyositis

MRI.	Magnetic resonance imaging
WBMRI.	Whole-body magnetic resonance imaging
T2W.	T2-weighted images
STIR.	Short tau inversion recovery
T2W/FS	T2-weighted images with fat suppression
CPK.	Creatine phosphokinase
LDH.	Lactate dehydrogenase
ESR.	Erythrocyte sedimentation rate
EMG.	Electromyography
DM.	Dermatomyositis
ACR-EULAR	American College of Rheumatology European League Against Rheumatism score
DAS.	Disease Activity Score
MMT	Manual Muscle Testing
CMAS	Childhood Myositis Assessment Score

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