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From Clinical Suspicion to Tissue Diagnosis: The Role of Muscle MRI in Juvenile Dermatomyositis: A Radiological Approach that Enabled the Transition from Clinical Suspicion to a Definitive, Tissue-Based Diagnosis

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Abstract: ***Juvenile dermatomyositis is an inflammatory myopathy characterised by proximal muscle weakness and distinctive cutaneous manifestations. Muscle MRI serves as a diagnostic tool by specifying the extent of inflammatory involvement and assisting targeted muscle biopsy for definitive diagnosis. Clinical Scenario: A 15-year-old male presented with progressive, symmetrical proximal limb weakness and markedly elevated serum creatine phosphokinase (CPK). Muscle MRI revealed diffuse STIR hyperintense edema involving the paravertebral, pelvic-girdle, and thigh muscles, accompanied by perifascial and subcutaneous edema. MRI-guided biopsy of the right vastus lateralis demonstrated histopathological features consistent with dermatomyositis. Strong anti-Mi-2β positivity and characteristic cutaneous lesions further substantiated the diagnosis of juvenile dermatomyositis. Administration of intravenous methylprednisolone followed by oral corticosteroids resulted in significant clinical improvement.***

Keywords: Anti-Mi-2β; inflammatory myopathy; juvenile dermatomyositis; muscle biopsy; muscle MRI; STIR hyperintensity

1. Introduction

Juvenile dermatomyositis (JDM) is an immune-mediated inflammatory myopathy characterized by symmetrical proximal muscle weakness and distinctive cutaneous manifestations. Diagnosis relies on clinical, biochemical, serological, imaging, and histopathological findings.[3,5] Muscle MRI is a valuable noninvasive tool for detecting active inflammation, defining its distribution, and demonstrating fascial and subcutaneous involvement.[2,3,5] It also facilitates targeted muscle biopsy, thereby reducing sampling error.[2,4] In this case, MRI revealed extensive symmetrical muscle, perifascial, and subcutaneous involvement, which supported the diagnosis and guided the selection of the vastus lateralis for biopsy in JDM.

2. Clinical Scenario

A 15-year-old male presented with progressive, symmetrical proximal and neck-flexor weakness, abnormal gait, and markedly elevated creatine phosphokinase (CPK) levels (9,000–14,000 U/L). MRI revealed diffuse STIR hyperintense edema involving the cervical, thoracic, and lumbar paravertebral muscles, while the spinal cord appeared normal. Gluteal and thigh MRI demonstrated symmetrical pelvic-girdle and thigh muscle edema with mild atrophy, as well as perifascial and subcutaneous involvement, findings suggestive of dermatomyositis. MRI-guided biopsy of the right vastus lateralis confirmed the diagnosis. Strong anti-Mi-2β positivity and characteristic facial and anterior chest cutaneous lesions further supported JDM. Treatment with intravenous methylprednisolone followed by oral corticosteroids led to improved muscle strength and a substantial decline in CPK.



Figure 1



Figure 2

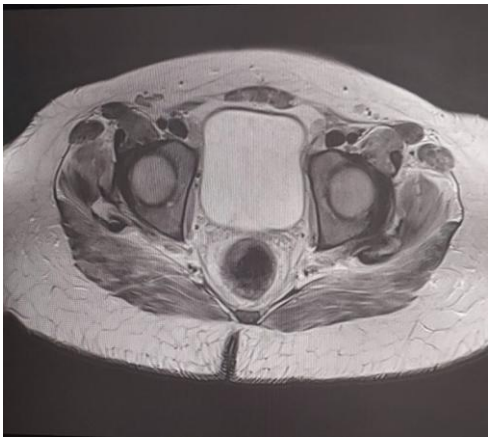


Figure 3

Figure 1. Coronal T1-weighted MRI of the pelvis and thighs demonstrating bilateral symmetrical involvement of the pelvic-girdle and thigh musculature with mild muscle atrophy.

Figure 2. Coronal STIR MRI demonstrating symmetrical diffuse hyperintense edema of the bilateral pelvic-girdle and thigh musculature with associated perifascial and subcutaneous edema, favouring dermatomyositis.

Figure 3. Axial T2-weighted MRI of the pelvis demonstrating bilateral relatively symmetrical signal abnormalities involving the pelvic-girdle musculature, consistent with the diffuse inflammatory muscle involvement demonstrated on fluid-sensitive imaging.

3. Discussion

Role of MRI in Inflammatory Myopathies

Fluid-sensitive sequences, particularly short tau inversion recovery (STIR) and fat-suppressed T2-weighted imaging, reveal active muscle inflammation as areas of increased signal intensity, which reflect elevated tissue water content and edema.[2, 4, 5] In contrast, T1-weighted imaging is effective for identifying chronic structural abnormalities, including muscle atrophy and fatty replacement[2, 4] MRI can also demonstrate fascial and subcutaneous inflammatory involvement, thereby providing detailed information regarding the distribution and extent of disease.[2, 3, 5]

MRI abnormalities may also serve as indicators of disease activity. Barsotti et al. demonstrated a significant association between muscle edema observed on thigh MRI and both serum creatine kinase levels and physician-assessed myositis activity.[1] In JDM, whole-body MRI has identified muscle inflammation not detected during clinical examination, as well as subcutaneous and myofascial involvement. These findings suggest that clinical assessment alone may underestimate the total inflammatory burden[3]

MRI Patterns in Different Inflammatory Myopathies

Although muscle edema is a common feature across inflammatory myopathies, variations in distribution, symmetry, fascial involvement, muscle atrophy, and fatty replacement may offer valuable diagnostic clues. [2, 4] However, MRI findings often overlap between subtypes and should therefore be interpreted in conjunction with the clinical phenotype, autoantibody profile, and histopathology. [2, 4]

Dermatomyositis (DM/JDM)	Predominantly symmetrical proximal muscle edema; perifascial/fascial and subcutaneous edema may be prominent.[2,3,5]
Polymyositis (PM)	Predominantly symmetrical proximal intramuscular edema, with considerable overlap with DM.[2,4]
Immune-mediated necrotizing myopathy (IMNM)	Extensive muscle edema may occur, with atrophy and fatty replacement reflecting structural damage; imaging findings overlap with other inflammatory myopathies.[2,4]
Inclusion body myositis (IBM)	More selective and frequently asymmetric involvement, with prominent chronic atrophy and fatty replacement compared with predominantly proximal inflammatory patterns.[2,4]

These imaging patterns are supportive but not diagnostic. MRI should therefore be used to complement, rather than replace, clinical assessment, muscle enzyme analysis, autoantibody testing, and histopathological evaluation.[2,4]

Significance of MRI in the Present Case

MRI revealed inflammatory involvement that extended beyond clinical findings, including diffuse STIR hyperintense edema of the cervical, thoracic, and lumbar paravertebral muscles, as well as symmetrical pelvic-girdle and thigh muscle edema with mild atrophy, perifascial, and subcutaneous involvement. Whole-body MRI in JDM has similarly demonstrated clinically occult muscle, myofascial, and subcutaneous abnormalities.[3] Although muscle edema is not specific to any one disease, the MRI pattern, when interpreted alongside proximal weakness, markedly elevated CPK, characteristic cutaneous manifestations, and anti-Mi-2β positivity, provided strong support for a diagnosis of JDM[2, 3, 5]

MRI-Guided Tissue Diagnosis

MRI is valuable for identifying actively affected muscle and for assisting in the selection of an appropriate biopsy site, thereby reducing the risk of sampling relatively uninvolved or severely damaged tissue. [2, 4] In this case, MRI guided the selection of the right vastus lateralis for biopsy, with histopathology subsequently confirming dermatomyositis. MRI thus served as a critical link between clinical suspicion and definitive tissue diagnosis.

4. Conclusion

This case illustrates the practical diagnostic value of muscle MRI in juvenile dermatomyositis. [2, 3, 5] Beyond defining the distribution of muscle, fascial, and subcutaneous inflammation, MRI can assist in selecting an appropriate site for muscle biopsy.[2, 4] Integrating MRI findings with clinical, biochemical, serological, and histopathological data enables a targeted and comprehensive approach to diagnosing inflammatory myopathies.[2, 4, 5]

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