The role of magnetic resonance imaging in the diagnostic work-out of myopathies: differential diagnosis between inflammatory myopathies and muscular dystrophies

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Abstract

Objective

The differential diagnosis between idiopathic inflammatory myopathies (IIM) and muscular dystrophies (MD) may be challenging. We analysed the potential role of muscular magnetic resonance imaging (MRI) in the differential diagnosis between IIM and MD.

Methods

MRI of patients (91 IIM and 43 MD), studied with a standardised protocol, have been collected. The presence of oedema, muscular atrophy and intramuscular adipose changes were evaluated. Moreover, we computed a composite score for each MRI item to better discriminate between the two diseases.

Results

Oedema was significantly more prevalent in IIM compared with MD in pelvis muscles ($p<0.001$), anterior lodge and medial lodges ($p=0.044$) of the thighs. Adipose infiltration/substitution and muscular atrophy were more prevalent in MD, in particular adipose tissue was prevalent in all the compartments of the thighs ($p<0.05$), atrophy was prevalent at the thighs and pelvis muscles ($p<0.001$). The probability of IIM increased with higher oedema score and decreased with higher atrophy and intramuscular adipose infiltration/substitution scores.

Conclusion

A different distribution of muscular involvement between IIM and MD has been identified. Muscular MRI may be useful in the differential diagnosis, potentially reducing the number of muscular biopsies that may be reserved only for doubtful cases.

Key words

myositis, dystrophy, oedema, magnetic resonance imaging, differential diagnosis

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Introduction

Idiopathic inflammatory myopathies (IIM) are a group of rare diseases with an autoimmune pathogenesis and unknown aetiology (1). IIMs include polymyositis (PM), dermatomyositis (DM), sporadic inclusion body myositis (IBM) and overlap myositis, where the muscle disorder is associated with other connective tissue disease (usually scleroderma or mixed connective tissue disease) (2). The diagnosis and assessment of disease activity in IIM patients is based on a combination of clinical, serological and biochemical features (3) but, especially in patients without skin rash, the histopathological evaluation of muscle is often necessary (4, 5).

Muscular dystrophies (MD) are a complex and heterogeneous group of genetically determined degenerative muscle diseases (6). Muscle weakness is the common sign of MD with varying muscle involvement patterns, different disease history and progression rate and, often, rather specific pathology at muscle biopsy. The most frequent forms of muscular dystrophies include Duchenne and Becker muscular dystrophies, myotonic dystrophy, facioscapulohumeral dystrophy, limb-girdle muscular dystrophies and congenital muscular dystrophies.

IIM and MD are both characterised by similar clinical features and progressive muscle weakness represents the major shared disease manifestation. For this reason, when exclusive clinical characteristics are absent, such as skin rash, myositis-specific autoantibodies for IIM or a suggestive phenotype or familiar history for MD, the two pathological conditions may be hardly distinguishable only focusing on history case and medical examinations, as both are associated to muscle weakness/pain and raised serum levels of creatine kinase. Muscle biopsy is usually crucial for differential diagnosis and for planning other investigations such as specific DNA testing for MD (6).

Magnetic resonance imaging (MRI) is a non-invasive tool useful in diagnosis and follow-up both in IIM and MD (7, 8). It does not require ionising radiation and allows multiplanar scanning with high natural soft issue contrast (9). Studying the muscle, MRI can easily identify the oedema due to muscular inflammation, the fat infiltration/ substitution and the presence of muscular atrophy. It can distinguish between active muscle inflammation and chronic muscle damage (10) helping in defining the activity of the disease in patients with IIMs. By identifying affected muscles, MRI is useful for targeted muscle biopsy (11) and, depicting specific muscular involvement patterns, might help in differentiating the two diseases amidst overlapping phenotypes (12).

The purpose of the study is to assess whether MRI can identify a significant difference between IIM and MD in terms of muscular involvement distribution and MR signal intensity changes.

Materials and methods

Patient selection

We retrospectively collected data from patients referring to the Rheumatology and Neuromuscular Units of Pisa University Hospital (Pisa, Italy) from January 2008 to December 2018, that received diagnosis of IIM or MD. A total of 134 patients were enrolled and subdivided in two groups: 91 patients affected by IIM diagnosed according to the 2017 EULAR/ACR criteria (4) and 43 by confirmed MD. All patients underwent muscle MRI of thighs and pelvis including paravertebral lodges. In addition, the following clinical data were collected: sex, age at diagnosis, disease duration at the moment of the MRI scan, specific subgroups of diseases (Table I), muscular symptoms (myalgias, weakness, and muscular strength according to Walton-Gardner-Medwin scale (13)), and the peak values of creatinine-kinase (CK) during the disease course.

Local ethics committee approval was not required for the study, because clinical data and muscle MRI were performed as part of a routine protocol used in our clinical practice. All patients signed a statement permitting the use of their data, in anonymous form, for research purposes.

MR Imaging technique

The exams were performed with a 1.5 Tesla MR scanner (General Electric®,

Table I. Detailed diagnosis of patients with IIM and MD.

Idiophatic inflammatory myopathies (IIM)		Muscular dystrophies (MD)	
Dermatomyositis	30	Facioscapulohumeral muscular dystrophy (FSHD)	24
Polymyositis	59	Limb-girdle muscular dystrophies (LGMD)	19
Inclusion body myositis	2		

Hdxt Twin Speed 1.5T) using a twelve-channel phased-array body coil or with a 3.0 Tesla MR scanner (General Electric®, MR 750 Discovery 3.0T) with an eight-channel phased-array receive-only coil (TORSO-PA).

The standard protocol included: STIR (Short Tau Inversion Recovery), DUAL ECHO fast-GRE (in- and out-of-phase) and DWI (diffusion-weighted Imaging) sequences, all acquired in the axial plane. Technical parameters were:

- STIR: repetition time (TR): 3300-3900; echo time (TE): 50–70; inversion time (IT): 150 ms (1.5T) and 200 ms (3T); slice thickness/gap: 10 mm/12 mm; field of view (FOV): 36–44 cm; matrix: 320x192; number of excitation (NEX): 1.
- DUAL ECHO: TR: 100–140; slice thickness/gap: 10 mm/12 mm; FOV: 36–44 cm; matrix 340x360; NEX: 1; IN PHASE TE: 4.5–4.7; OUT OF PHASE TE: 2.2–2.4.
- DWI: TR: 2600-3400; TE: 56–58; b values: 0 and 500 s/mm^2 all directions; slice thickness/gap: 10 mm/12 mm; FOV: 36–44cm; matrix: 136x256; NEX: 5–6.

All the exams included thighs and pelvic muscles. The total scan time range was between 20 and 30 minutes.

Image analysis

STIR and DWI sequences were used to detect inflammatory alterations, meant as oedema at the level of muscular bellies, muscular fasciae and subcutaneous soft tissues (14), known as important markers of disease activity (7). The DUAL ECHO sequence is able to assess the presence of intracellular and extracellular fat because of the different resonance frequency of fat and water protons thus allowing to assess the presence of adipose infiltration and/or substitution within the muscles. This data is suggestive of chronic processes and, therefore, for an advanced disease stage (15). In addition, this sequence quickly provides good anatomic detail and a morphological qualitative evaluation of the muscular trophism.

Muscles were grouped according to their anatomical distribution, as follows:

- Pelvis (paravertebral, gluteus maximus, gluteus medium, gluteus minimus, iliopsoas, piriformis and tensor fasciae latae).
- Thigh's anterior lodge (rectus femoris, vastus lateralis, vastus medialis, vastus intermedius, sartorius and pectineus).
- Thigh's medial lodge (adductor longus, adductor brevis, adductor magnus, obturator externus, obturator internus and gracilis).
- Thigh's posterior lodge (semimembranosus, biceps femoris and semitendinosus).

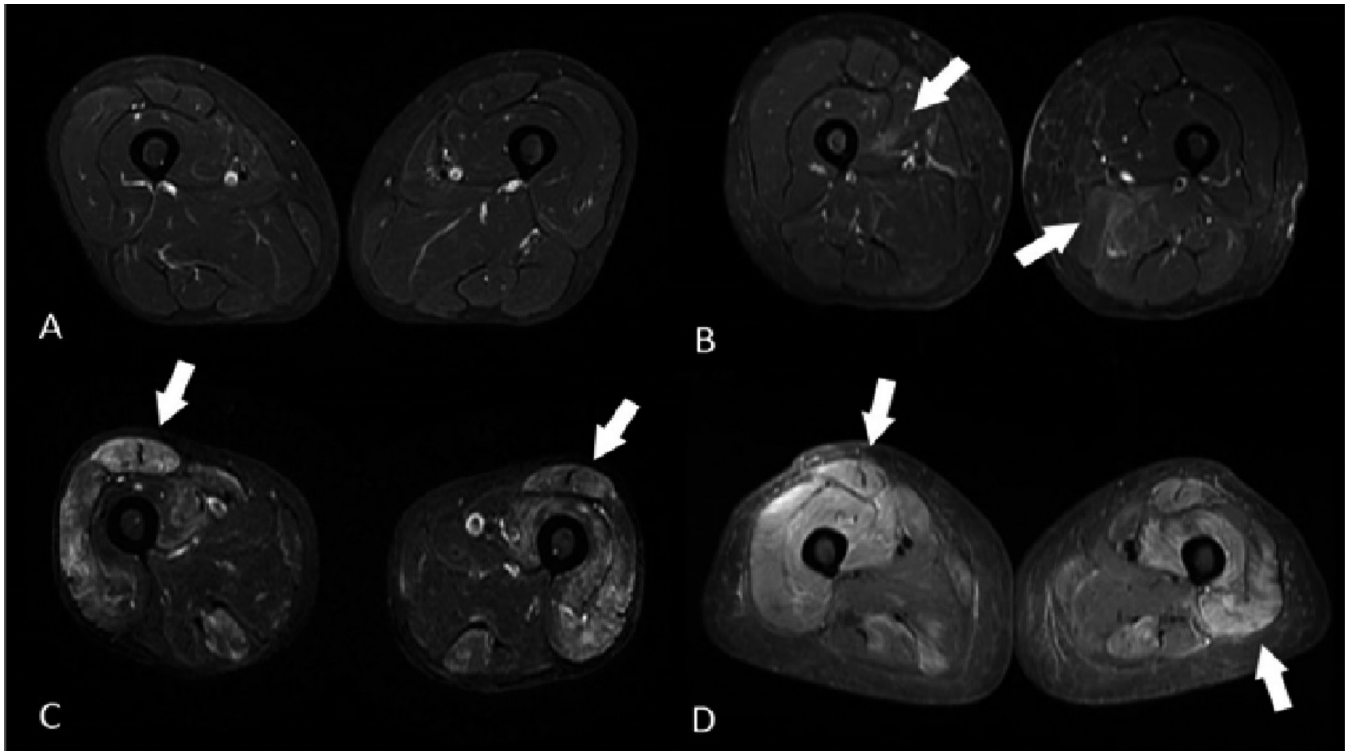

Fig. 1. Axial STIR images of thigh sections of four different patients with IIM. A: no oedema (grade 0). B: shaded oedema (grade 1) of right vastus medialis and left adductor magnus (white arrows). C: moderate oedema (grade 2) of rectus femoris, vastus lateralis, vastus intermedius and semimembranosus. D: widespread and severe oedema (grade 3) of quadriceps.

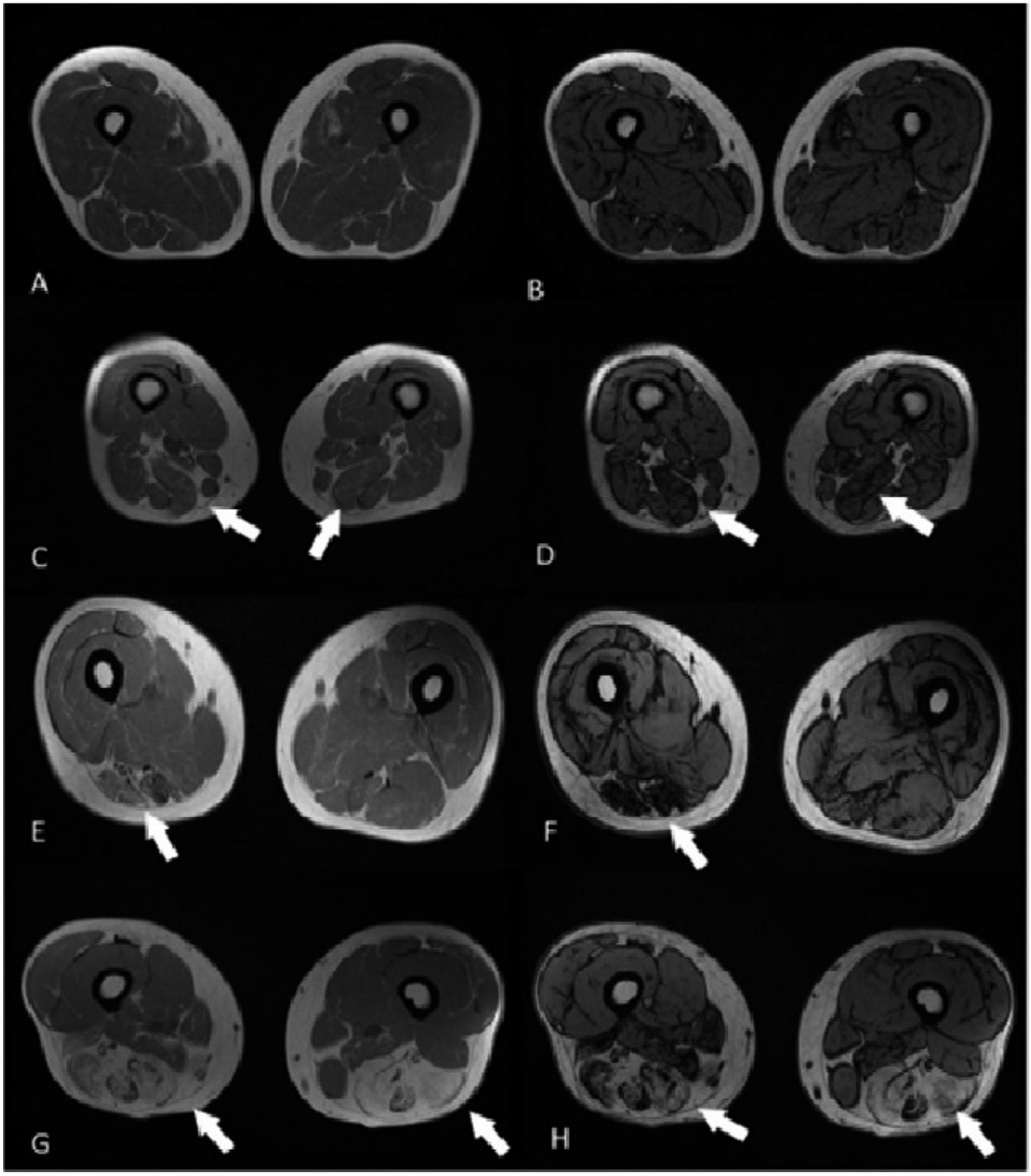

Fig. 2. Axial image of IN- (A, C, E and G) and OUT- of phase (B, D, F and H) sequences of thigh sections of patients with muscular disease. A-B: no atrophy and no fatty infiltration (0 absent). C-D: mild atrophy (grade 1) and mild adipose infiltration (grade 1) of semimembranosus (white arrows). E-F: asymmetric moderate atrophy (grade 2) and adipose infiltration (grade 1) of the right posterior lodge (white arrows). G-H: severe atrophy (grade 3) and adipose substitution (grade 2) of the bilateral posterior lodge (white arrows).

For each muscle and for each anatomic group, oedema, adipose infiltration/substitution and muscular atrophy have been qualitatively evaluated by an experienced radiologist. Muscular oedema (Fig. 1) and atrophy (Fig. 2) were scored according to a 4-points Likert scale (7) as follows: 0 absent, 1 mild, 2 moderate, 3 severe, while fatty intramuscular content was scored as follows: 0 absent, 1 infiltration and 2 substitution (Fig. 2).

To assess the analysis repeatability, the radiologist evaluated all exams two times, four months apart. Finally, to evaluate the reproducibility, a total of 80 randomly selected MRI scans have been evaluated in the same way by a second experienced radiologist, blinded to the clinical diagnosis and to the results of the first reader.

Statistical analysis

All the clinical parameters of the patients and MRI data are expressed as the mean value ± standard deviation. Comparisons between mean values were analysed by Student's t test. The comparison between proportions was made with z-test for proportions. The degree of oedema (4 levels), atrophy (4 levels) and fatty content (3 levels) in each muscle (21 rows) in both IIM and MD was evaluated by 21x4 contingency table. The degree of association between the two variables was assessed by Chi-square test.

In order to identify the diagnostic features that may be helpful for the differential diagnosis between IIM and MD, we defined different probability scores, considering each specific MRI alteration in every single muscle, and using all the clinical and MRI variables. Definition of scores (defined as probability of IIM) for muscular oedema, atrophy and fatty content were performed by Structural Equation Model (SEM) considering each muscle, age at MR and disease duration as linear predictor.

In detail, the score (latent variable) is defined as:

$$y = \sum_{i=1}^{k} rho_i * x_i$$

where:

- rho_i represents the standardised slope of the i-th variable estimated by the SEM model. rho_i is considered as the correlation coefficient between x_i and the variable y;
- x_i is the i-th variable of the k variables used as linear predictors.

The y score has been rescaled with the Rescaling (min-max normalisation) method:

$$y_{std} = \frac{y - \min(y)}{\max(y) - \min(y)} * 100$$

y_{std} has a range of values from 0 to 100 and can be interpreted as the estimate of the probability of IIM as a function of the parameters considered. Values closer to 100 indicate higher probability of IIM.

The strength of direct or inverse association (rho) of each index with the score were defined as: mild if rho <0.3 or rho>-0.3, moderate if 0.3≤rho<0.5 or -0.5<rho≤-0.3, good if 0.5≤rho<0.7 or -0.7<rho≤-0.5, strong if rho≥0.7 or rho≤-0.7. In the results, the variables with rho≥0.7 or rho≤ -0.7 (*i.e.* those that most influence the y scores) will be commented.

The increase in risk of IIM as a function of scores for oedema, atrophy and fat content was evaluated with logistic regression. The increase in risk of IIM as a function of scores for oedema, atrophy and fat content was evaluated with logistic regression. Inter- and intra-observer agreement has been evaluated by Chrombach's alpha test. $p<0.05$ was considered statistically significant.

Results

Epidemiological and demographic data

The demographic profile of the study is summarised in Table II. The percentage of women was significantly higher in patients with IIM compared with MD. In the group with IIM, the average age at the diagnosis was significantly higher compared with MD and, consequently, also the age at the MR scans. On the contrary, the mean disease duration was lower in IIM compared with MD.

Clinical and laboratory data

As summarised in Table III, muscular pain was more frequent in patients with IIM. At the moment of evaluation when they performed MRI, the functional status assessed with the Walton and Gardner-Medwin scale was significantly worst in patients with IIM compared with patients with MD (Table III). The peak values of CK levels during the course of the disease were significantly higher in patients with IIM compared to MD, respectively, 2503 UI/dL and 1601 UI/dL ($p<0.001$).

MRI data

In all muscular groups examined, muscular oedema was significantly more prevalent in IIM compared to MD, particularly in muscles of pelvis ($p<0.001$), anterior lodge ($p=0.044$) and medial lodge of the tights ($p=0.044$) (Table IV). Particularly, oedema resulted to be significantly more prevalent in: gluteus (maximus, medium and minimus), iliopsoas, piriformis, tensor fasciae latae, vastus lateralis, pectineus, adductor longus, adductor brevis, obturator externus and obturator internus. Of interest, oedema score of 3 was never depicted in patients with MD, and only a few of them showed an oedema score of 2 (Supplementary Table S1).

On the contrary, adipose infiltration/substitution and muscular atrophy were significantly more prevalent in MD compared with IIM, except for the pelvis where no significant difference was observed (Table IV).

In particular, in patients with MD, a significant prevalence of the adipose infiltration/substitution and atrophy was present in almost all the muscles considered; exceptions are reported in Supplementary Tables S2 and S3.

Muscular atrophy was significantly more prevalent in MD in all the muscular groups ($p<0.001$) (Table IV). Analysing every single muscle, the only exceptions are the following: iliopsoas, rectus femoris and obturator externus (Suppl. Table S3).

Finally, the most severe degree of atrophy was observed in the MD group and the prevalence of atrophy showed an increase with its severity score in MDs in comparison with IIMs (Suppl. Table S3).

The Cronbach α showed an interobserver agreement of 0.91 and an intraobserver agreement of 0.86.

Definition of scores to discriminate between IIM and MD

The weight of each variable, meant as strength of association (rho), in the three scores was computed, one for muscular oedema, one for adipose tissue infiltration/substitution and one for atrophy (Suppl. Tables S4, S5 and S6). Considering significant the variables with rho greater than 0.7, we identified the findings with higher influence on the scores for their potential role in the

Table II. Epidemiological data of the group of patients with IIM and DM.

	IIM Tot 91	MD Tot 43	*p*-value
Women (%)	64 (70.3)	16 (37.2)	<0.0001
Age at MR scans	58.8 ± 13.2	53.0 ± 13.2	0.02
Age at diagnosis	56.5 ± 14.2	46.7 ± 16.9	0.0007
Disease duration at the MR	45.7 ± 72.7	202.9 ± 157.8	<0.0001

IIM: idiopathic inflammatory myopathies; DM: ; MR:

Table III. muscular symptoms in patients with idiopathic inflammatory myopathies and muscular dystrophies.

	IIM (75 patients)	MD (43 patients)	*p*-value
Muscular pain	28 (37.3%)	2 (6.7 %)	<0.001
Functional status by Walton Gardner-Medwin scale	3.1 ± 2.1	2.1 ± 1.2	0.021

IIM: idiopathic inflammatory myopathies; MD: muscular dystrophies.

Table IV. Muscular groups with detectable oedema, adipose tissue/infiltration and atrophy in idiopathic inflammatory myopathies and muscular dystrophies.

	Muscular group	IIM (%)	MD (%)	*p*-value
Oedema	Pelvis	49 (55.6)	7 (16.7)	<0.001
	Anterior lodge	63 (70.8)	22 (51.2)	0.027
	Medial lodge	52 (58.4)	17 (39.5)	0.042
	Posterior lodge	49 (55.1)	17 (39.5)	n.s.
Adipose tissue	Pelvis	48 (53.9)	27 (64.3)	0.26
	Anterior lodge	29 (32.6)	28 (65.1)	<0.001
	Medial lodge	50 (56.2)	34 (79.1)	0.01
	Posterior lodge	55 (61.8)	35 (81.4)	0.023
Atrophy	Pelvis	18 (20.2)	25 (59.5)	<0.001
	Anterior lodge	15 (16.9)	27 (62.8)	<0.001
	Medial lodge	15 (16.8)	26 (60.5)	<0.001
	Posterior lodge	30 (33.7)	31 (72.1)	<0.001

IIM: idiopathic inflammatory myopathies; DM: muscular dystrophies.

differential diagnosis between the two disease groups, as follows:

- for muscle oedema (presence of muscular oedema in these muscles is more likely associated to IIM): gluteus maximus (rho 0.81), gluteus medium (rho 0.92), gluteus minimus (rho 0.92), iliopsoas (rho 0.83), piriformis (rho 0.89) and tensor fasciae latae (rho 0.89), rectus femori (rho 0.72), pectineus (rho 0.85), adductor longus (rho 0.89), adductor brevis (rho 0.9), gracilis (rho 0.77), obturator externus (rho 0.9), obturator internus (rho 0.9).
- for fatty infiltration / substitution (presence of fatty tissue in these muscles is more likely associated to MD): gluteus medium (rho -0.73), gluteus minimus (rho -0.73), vastus medialis (rho -0.77), vastus intermedius (rho -0.77), gracilis (rho -0.72), adductor magnus (rho -0.70), semimembranosus (rho -0.71), biceps femoris (rho -0.77), semitendinosus (rho -0.72).
- for muscular atrophy (presence of atrophy in these muscles is more likely associated to MD): gluteus maximum (rho -0.87), gluteus medium (rho -0.91), gluteus minimus (rho -0.87), tensor fasciae latae (rho -0.75), vastus lateralis (rho -0.81), vastus medialis (rho -0.84), vastus intermedius (rho -0.82), sartorius (rho -0.78), pectineus (rho -0.79), gracilis (rho -0.81), adductor longus (rho -0.72), adductor magnus (rho -0.73), semimembranosus (rho -0.73), biceps femoris (rho -0.76), semitendinosus (rho -0.74).

The analysis of the ROC curve for the proposed scores was not able to identify a definite cut-off (with sensibility and specificity higher than 80%) for their potential role in the differential diagnosis between IIM and MD. However, based on the presence of the different variables and their cumulative grading, the three scores were correlated to the probability of having IIM according to the following results:

- the increase of 1 point of the oedema score increases the probability of having IIM by 8%;
- the increase of 1 point of the adipose tissue score reduces the probability of having IIM by 5%;
- the increase of 1 point of the atrophy score reduces the probability of having IIM by 13%.

Discussion

In our study we compared the MRI of thighs and buttocks findings in two large monocentric cohorts of patients respectively with IIM and MD, we identified specific MRI patterns of muscular involvement regarding oedema, atrophy and adipose infiltration/substitution. Additionally, we proposed three scores that could be useful in the diagnostic differentiation between IIM and MD. Indeed, there is an increasing interest in MRI "pattern recognition". Limited number of studies have been performed to evaluate whether muscular MRI can identify significant differences in the distribution and in the imaging characteristics of the muscular involvement in IIM and MD (9, 16). However, to date, to our knowledge, no recent studies have been performed on the role of MRI in distinguishing these two muscular diseases.

Oedema-like changes are the first observable findings in case of muscle damage, which eventually develop into adipose replacement and muscle atrophy (17). Our analysis showed that oedema was significantly prevalent in all muscular groups in IIM, compared to MD, and the probability of IIM diagnosis increased in patients with more severe oedema in higher number of muscles. Indeed, consistently with the previous results of Barsotti *et al*., the most severe degrees of oedema were

identifiable only in IIM patients (7); this data has been previously correlated with histopathological changes in DM (18). Anyway, low grades of muscular oedema may be identified in patients with MD due to inflammation caused by the muscular damage (12).

For the assessment of oedematous alterations, we propose to combine both STIR and DWI sequences to increase the sensibility for muscular oedema (19). The oedema score proposed in this article suggests that an increase in the number of muscles affected by oedema, together with its severity, significantly increase the probability of the diagnosis of IIM instead of MD. Only in the posterior lodge of the thigh, the prevalence of muscular oedema was similar comparing IIM and MD. For this reason, if oedema is identified at this level, even of high degree, MD cannot be ruled out.

Intramuscular adipose changes are typically characterised by a progressive evolution with a trend from a mild infiltration in the early phases to a complete adipose substitution in the late phases of the diseases, and they are usually correlated to denervation (20). Actually, adipose infiltration is more commonly identified in patients with MD (21). However, it may be identified also in the chronic advanced stage of IIM, where it reflects largely irreversible damage. Contrarily, oedema more likely represents muscular inflammation and correlates with disease activity (22). In our study, infiltration/substitution shows a significant prevalence in MD in all muscle groups and almost in all the single muscles. A predominant fatty substitution in muscles should lead the radiologist to suspect MD instead of IIM. On the opposite of the oedema score, the proposed adipose tissue score showed a significantly reduced probability of IIM diagnosis with the increased number of muscles affected by adipose infiltration/substitution, together with its severity.

The last aspect considered was the muscular trophism. Atrophy has a significant prevalence in MD in all muscles examined. As for the adipose infiltration/substitution score, the increase of muscles affected by muscular atrophy and its severity – both included in the proposed adipose changes score – significantly reduce the probability of IIM diagnosis. It is well known that MD induces adipose infiltration/substitution of peculiar muscular groups and various studies have been carried out to design diagnosis algorithms based on the MR findings (10, 12). MR imaging can contribute to the differential diagnosis between different MD, but it may be present also in patients with IIM, especially in those with a longstanding disease. Additionally, muscular atrophy and hypotrophy may not be disease-specific and, thus, confounding. They can be associated also with disuse and/or ageing processes, especially in the muscles involved in maintaining the upright position in painful or functional impotence conditions prolonged over time (21).

To our knowledge, our study is the first that proposes a relatively simple way to differentiate IIM from MD. The point of strength of our work is the high number of patients, considering the rarity of the muscular diseases evaluated. In addition, our study was performed with widely available MRI equipment with a short scanning time about 20-30 minutes with a good patients' compliance. Finally, thanks to the use of simple scoring systems, we were able to obtain a good inter- and intra-observer agreement and may lead to short learning curve for radiologists.

Limitations of our work were the retrospective and monocentric nature of the study and the relatively inhomogeneity of the patients enrolled by the rheumatologic and neuromuscular clinics that allowed to compare the groups only for a limited number of clinical variables. Moreover, we performed only a qualitative and semi-quantitative evaluation of muscle MRI; although this aspect may be a limitation, especially for the use of the proposed method in clinical trials, we preferred a simplified approach that may be more useful in the routinely clinical practice. Finally, based on the evaluation of each single muscle of the pelvis and the thighs, the scoring process of all the MRI findings was rather time-consuming; however, novel information technologies developments, such as computer-aided systems and deep-learning methods, could reduce the time needed for the overall evaluation. Further studies are needed to explore the possibility of a MR quantitative approach.

Conclusion

According to MRI parameters, a significant difference in the distribution of muscular involvement between IIM and MD has been observed as well as different patterns of muscular alterations, varying from oedema to fat infiltration and substitution. On MRI, when muscular oedema, especially of high grade, is depicted in more than a muscular compartment the diagnosis of IIM should be considered, while predominant muscular fatty infiltration/substitution suggests the probability of MD. Although our scores should be validated in external cohorts, we identified significant prevalence of oedema in IIM, while adipose infiltration/substitution and atrophy have significant prevalence in MD. Finally, it is noteworthy that similar prevalence of muscular oedema can be found in the posterior lodge of the thigh. Therefore, in this case, MD cannot be ruled out.

In conclusion, muscular MRI may help in the difficult differential diagnosis between IIM and MD and potentially reduce the number of muscular biopsies, which may be reserved only for doubtful cases.

Take home messages

- The differential diagnosis between IIM and MD may be challenging.
- Muscular MRI may identify oedema, muscular atrophy and muscular adipose tissue.
- A semiquantitative analysis of muscular MRI may increase the accuracy of this test.
- Muscular MRI may help in the differential diagnosis between MD and IIM.

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References

1. DALAKAS MC: Inflammatory muscle diseases. *N Engl J Med* 2015; 372: 1734-47. https://doi.org/10.1056/nejmra1402225
2. LUNDBERG IE, MILLER FW, TJÄRNLUND A, BOTTAI M: Diagnosis and classification of idiopathic inflammatory myopathies. *J Intern Med* 2016; 280: 39-51. https://doi.org/10.1111/joim.12524
3. CARDELLI C, ZANFRAMUNDO G, COMETI L *et al*.: Idiopathic inflammatory myopathies: one year in review 2021. *Clin Exp Rheumatol* 2022; 40: 199-209. https://doi.org/10.55563/clinexprheumatol/vskjxi
4. LUNDBERG IE, TJÄRNLUND A, BOTTAI M *et al*.: 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups. *Ann Rheum Dis* 2017; 76(12): 1955-64. https://doi.org/10.1136/annrheumdis-2017-211468
5. BARSOTTI S, DASTMALCHI M, NOTARNICOLA A *et al*.: Performance of the new EULAR/ACR classification criteria for idiopathic inflammatory myopathies (IIM) in a large monocentric IIM cohort. *Semin Arthritis Rheum* 2020; 50(3): 492-7. https://doi.org/10.1016/j.semarthrit.2019.12.001
6. BARSOTTI S, TRIPOLI A, POLLINA LE, MOSCA M, NERI R: Histopathology of the muscle in rheumatic diseases. *Reumatismo* 2018; 70: 133-45. https://doi.org/10.4081/reumatismo.2018.1051
7. BARSOTTI S, ZAMPA V, TALARICO R *et al*.: Thigh magnetic resonance imaging for the evaluation of disease activity in patients with idiopathic inflammatory myopathies followed in a single center. *Muscle Nerve* 2016; 54(4): 666-72. https://doi.org/10.1002/mus.25099
8. MAURER B, WALKER UA: Role of MRI in diagnosis and management of idiopathic inflammatory myopathies. *Curr Rheumatol Rep* 2015; 17(11): 67. https://doi.org/10.1007/s11926-015-0544-x
9. CARLIER PG, MERCURI E, STRAUB V: Applications of MRI in muscle diseases. *Neuromusc Disord* 2012; 22 (Suppl. 2): S41. https://doi.org/10.1016/j.nmd.2012.08.001
10. DIAZ-MANERA J, LLAUGER J, GALLARDO E, ILLA I: Muscle MRI in muscular dystrophies. *Acta Myol* 2015; 34: 95-108.
11. VAN DE VLEKKERT J, MAAS M, HOOGENDIJK JE, DE VISSER M, VAN SCHAIK IN: Combining MRI and muscle biopsy improves diagnostic accuracy in subacute-onset idiopathic inflammatory myopathy. *Muscle Nerve* 2015; 51(2): 253-8. https://doi.org/10.1002/mus.24307
12. STRAMARE R, BELTRAME V, DAL BORGO L *et al*.: MRI in the assessment of muscular pathology: a comparison between limb-girdle muscular dystrophies, hyaline body myopathies and myotonic dystrophies. *Radiol Med* 2010; 115(4): 585-99. https://doi.org/10.1007/s11547-010-0531-2
13. ANGELINI C, SEMPLICINI C, RAVAGLIA S *et al*.: New motor outcome function measures in evaluation of late-onset Pompe disease before and after enzyme replacement therapy. *Muscle Nerve* 2012; 45(6): 831-4. https://doi.org/10.1002/mus.23340
14. KUBÍNOVÁ K, MANN H, VENCOVSKÝ J: MRI scoring methods used in evaluation of muscle involvement in patients with idiopathic inflammatory myopathies. *Curr Opin Rheumatol* 2017; 29: 623-631. https://doi.org/10.1097/bor.0000000000000435
15. KALIA V, LEUNG DG, SNEAG DB, DEL GRANDE F, CARRINO JA: Advanced MRI techniques for muscle imaging. *Semin Musculoskelet Radiol* 2017; 21: 459-69. https://doi.org/10.1055/s-0037-1604007
16. PIPITONE N, NOTARNICOLA A, LEVRINI G *et al*.: Do dermatomyositis and polymyositis affect similar thigh muscles ? A comparative MRI-based study. *Clin Exp Rheumatol* 2016; 34(6): 1098-100.
17. MERCURI E, PICHIECCHIO A, ALLSOP J, MESSINA S, PANE M, MUNTONI F: Muscle MRI in inherited neuromuscular disorders: past, present, and future. *J Magn Reso Imaging* 2007; 25: 433-40. https://doi.org/10.1002/jmri.20804
18. MILISENDA JC, COLLADO MV, PINAL-FERNANDEZ I *et al*.: Correlation between quantitative and semiquantitative magnetic resonance imaging and histopathology findings in dermatomyositis. *Clin Exp Rheumatol* 2019; 37: 633-640
19. FARUCH M, GARCIA AI, DEL AMO M *et al*.: Diffusion-weighted magnetic resonance imaging is useful for assessing inflammatory myopathies. *Muscle Nerve* 2019; 59: 555-60. https://doi.org/10.1002/mus.26438
20. DULOR JP, CAMBON B, VIGNERON P *et al*.: Expression of specific white adipose tissue genes in denervation-induced skeletal muscle fatty degeneration. *FEBS Lett* 1998; 439: 89-92. https://doi.org/10.1016/s0014-5793(98)01216-2
21. FILLI L, WINKLHOFER S, ANDREISEK G, DEL GRANDE F: Imaging of myopathies. *Radiol Clin North Am* 2017; 55: 1055-70. https://doi.org/10.1016/j.rcl.2017.04.010
22. DAY J, PATEL S, LIMAYE V: The role of magnetic resonance imaging techniques in evaluation and management of the idiopathic inflammatory myopathies. *Semin Arthritis Rheum* 2017; 46: 642-9. https://doi.org/10.1016/j.semarthrit.2016.11.001