RHEUMATOLOGY

Rheumatology 2022;61:185–194
doi:10.1093/rheumatology/keab340
Advance Access publication 11 April 2021

Original article

A novel grey scale and Power Doppler ultrasonographic score for idiopathic inflammatory myopathies: Siena Myositis Ultrasound Grading Scale

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Abstract

Objectives. No clear-cut guidelines exist on the use of diagnostic procedures for idiopathic inflammatory myopathies (IIM) and only minimal and conflicting data report the use of ultrasound (US). In this regard, we aimed to assess if grey-scale (GS) and Power Doppler (PD) US, graded with a 0–3-point scale, may be a reliable tool in a cohort of patients affected by IIM.

Methods. All patients underwent US examination of both thighs in axial and longitudinal scans. Oedema and atrophy, both assessed in GS and PD, were graded with a 0–3-point scale. Spearman's test was used to identify the correlations between US and clinical and serological variables.

Results. A total of 20 patients were included. Six and two patients were evaluated twice and three times, respectively. Muscle oedema was found to be directly correlated with physician global assessment (PhGA), serum myoglobin and PD and negatively with disease duration. PD score was positively correlated to PhGA and negatively to disease duration. Muscle atrophy directly correlated with Myositis Damage Index, disease duration and patient's age. The single-thigh sub-analysis evidenced a direct correlation between PD score and Manual Muscle Test.

Conclusions. In our cohort, we found that oedema and PD are strictly related to early, active myositis, suggesting that an inflamed muscle should appear swollen, thickened and with Doppler signal. Conversely, muscle atrophy reflects the age of the patient and the overall severity of the disease. Such findings shed a new, promising, light on the role of US in diagnosis and monitoring of IIMs.

Key words: myositis, Power Doppler, ultrasonography, connective tissue diseases, autoimmunity

Rheumatology key messages

- We propose a GS and PD US 0-3-point scale for patients affected by IIM.
- Oedema and PD are correlated with disease activity, while muscle atrophy is correlated with patients' age and damage extent.
- Muscle US may be a useful, easy-to-perform and promising tool for the assessment of IIM.

Introduction

Idiopathic inflammatory myopathies (IIM) are autoimmune, severe, rare and chronic diseases typically affecting striate muscles and also variously involving other organs and systems [1].

IIM are usually distinguished between DM and PM, while inclusion body myositis represents a completely different subset of disease, in terms of pathogenesis, signs and symptoms, and response to treatment [2].

Aside of traditional and outdated classification criteria proposed by Bohan and Peter in 1975 [3, 4], more recent classifications have been developed by the ACR and EULAR in 2017, including stricter histological definitions and the presence of anti-Jo1 [5]. Further updates

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Submitted 7 December 2020; accepted 5 April 2021

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CLINICAL SCIENCE

will probably include the positivity of other myositis-specific antibodies (MSAs), scarcely considered in the 2017 ACR/EULAR criteria.

Nevertheless, the use of other, less invasive, diagnostic procedures has not been included: aside of electromyography (EMG), MRI is gaining a position in the routine baseline assessment of patients with suspected IIM [6], but no clear-cut guidelines exist on how to use this procedure in the daily clinical practice [7]. Similarly, only minimal and conflicting data report the use of muscle ultrasonography (US) in these conditions [8] and just a few described the role of Power Doppler (PD) [9–12].

In this regard, we aimed to assess if US examination, with both grey-scale (GS) and PD examination, may be a reliable tool in a cohort of patients affected by various IIMs. At the same time, we propose a new 0–3-point scale for oedema, atrophy, and PD in order to better stratify the different US findings in patients affected by IIM.

Methods

In this monocentric cross-sectional study, we prospectively collected, from July 2020 to January 2021, all patients referred to the Vasculitis and Myositis clinic, Rheumatology Unit, University of Siena, for suspected IIM, as well as patients with a previous, definite diagnosis of IIM and evaluated during follow-up or referred from other centres for a second opinion.

Clinical and serological data

For every patient, age, sex, definite diagnosis, duration of disease (expressed in months), biopsy, and MRI findings (if performed and expressed dichotomously as presence or absence of active IIM or muscle atrophy), antibodies, current treatment including Prednisone dosage, erythron-sedimentation rate, CRP, creatine phosphokinase (CPK), myoglobin and aldolase were recorded. Myositis specific (MSA) and associated antibodies (MAA) were assessed in all patients by our Autoimmunity Laboratory, using Myositis Euroline Immunoassay (Euroimmune, Lubeck, Germany).

Disease activity was evaluated measuring physician global assessment (PhGA) in 0–4 Likert scale and manual muscle testing (MMT) scored with Kendall scale, while disease damage using Myositis Damage Index (MDI) extent of damage, all belonging to International Myositis Assessment and Clinical Studies Group (IMACS) core set measures [13, 14].

Ultrasonography

US examination was carried out by an expert rheumatologist sonographer, blinded to clinical and serological data of the patients, with an Esaote MyLab Twice (Genoa, Italy) machine equipped with a broadband 6–18 MHz (LA435) linear transducer, with standardized B-mode and Doppler settings, which were optimized for all examinations. Doppler parameters were pulse repetition frequency within 500–1000 Hz and Doppler frequency within 5.9–11.1 MHz, maintaining colour gain just under the artifact limit. Focus was localized at rectus femoris level. The probe was held horizontal to the body surface, in order to keep the rectus femoris fascia perpendicular to the beam. PDUS assessment of anterior thigh muscles was performed on each patient, in supine position and on both sides. Grey-scale image, as well as the colour box for Doppler examination, were extended to comprise all muscle thickness until cortical profile of femoral diaphysis.

Quadriceps femoris muscle was always studied with anterior axial and longitudinal scans approximately at the junction of the median third with distal third. The rectus femoris was assessed as referring muscle, and vastus intermedius was assessed as comparator for echogenicity and thickness. Other muscle groups, vastus lateralis, adductors and hamstrings on posterior thigh, were assessed if specifically requested by the clinician, based on symptoms, history, physical examination, or MRI findings. The evaluation and grading of affected muscles were made also in comparison with unaffected sites in case of focal or unilateral IIM (Supplementary Fig. S1, available at *Rheumatology* online).

Siena Myositis Ultrasound Grading Scale (SMUGS)

A 0–3-point GSUS grading scale was proposed for both muscle oedema and muscle atrophy (fibro-adipose involution), and a 0–3-point PDUS scale was proposed for assessing muscle vascularity (Table 1; Figs 1 and 2). The US scales were applied separately by two rheumatologist sonographers on both axial and longitudinal scans, on each assessed muscle. The highest score obtained for each item and for each leg was computed as the pathologic US score for that patient.

Statistical analysis

The statistical analysis was conducted using GraphPad version 9.0.1 and SPSS version 27.0. Variables' normality was checked with Shapiro–Wilk test while Spearman's test was used to study variable correlations.

Initially, each component of SMUGS (oedema, PD and atrophy), each summed for both thighs, was separately tested with CPK, aldolase, myoglobin, PhGA, MDI and disease duration. These variables were also tested between themselves. Subsequently, we summed oedema and PD of both thighs and this total score was tested with CPK, aldolase, myoglobin, PhGA, MDI and disease duration. Moreover, we investigated the relationship between oedema, PD and atrophy and MMT by conducting a left/right sub-analysis for each leg: first considering the variables separately, then using the sum of oedema, atrophy and PD. Finally, we correlated oedema, PD and atrophy with MRI findings. P was considered statistically significant when below <0.05.

Inter-observer agreement for oedema, atrophy and PD was assessed using weighted kappa coefficient with a quadratic scale.

Table 1 Siena Myositis Ultrasound Grading Scale (SMUGS).

	Grey-scale oedema	Grey-scale atrophy	Power Doppler
0	Normal muscle echotexture (homogeneous echogenicity, with slightly more echoic appearance of rectus femoris with respect to vastus intermedius), with hyperechoic septa and hypoechoic muscle fibres, conserved thickness (comparable thickness of rectus and vastus intermedius).	Normal muscle echotexture (homogeneous echogenicity, with slightly more echoic appearance of rectus femoris with respect to vastus intermedius), with hyperechoic septa and hypoechoic muscle fibres, conserved thickness (comparable thickness of rectus and vastus intermedius, the overall thickness of thigh muscles is thicker than hypoderm).	No PD signal.
1	Focal hypoechoic areas, where septa are less evident. Conserved thickness.	Focal heterogeneously hyperechoic areas, where septa are thicker and more evident, and muscle fibres are thinner. Conserved muscle thickness (rectus femoris not thinner than vastus intermedius; the overall thickness of thigh muscles is thicker than hypoderm).	One or two PD signals in at least one muscle (PD vascular spots, small vessels of homogeneous diameters, vessel diameters approximately not superior to fibrous intramuscular septa).
2	Diffuse and heterogeneous hypoechogenicity (rectus femoris as hypoechoic or more than vastus intermedius), septa diffusely less evident. Conserved thickness.	Diffuse and heterogeneously hyperechoic muscle, with thicker septa and thinner muscle fibres. Conserved muscle thickness (rectus femoris not thinner than vastus intermedius; the overall thickness of thigh muscles is comparable or thicker than hypoderm).	>2 PD signals for each muscle (as vascular spots, small vessels of homogeneous diameters, vessel diameters approximately not superior to fibrous intramuscular septa).
3	Diffuse and heterogeneous hypoechogenicity (rectus femoris as hypoechoic or more than vastus intermedius), septa diffusely less evident. Increased thickness (rectus femoris became thicker than vastus intermedius).	Diffuse and heterogeneously hyperechoic muscle, with thicker septa and thinner muscle fibres. Reduced muscle thickness (rectus femoris thinner than vastus intermedius; the overall thickness of thigh muscles is thinner than hypoderm).	>2 PD signals for each muscle with larger diameter of the vessel (at least superior to fibrous intramuscular septa), or vessels with different diameters or branched vessels.

Ethics

All patients gave their written informed consent. The research was carried out in compliance with the Declaration of Helsinki, but no ethical committee approval was necessary, as US is part of the common clinical practice in our Vasculitis and Myositis clinic.

Results

A total of 20 patients (2 males, 18 females, median age 56.05 (15.88) years) were included in our study. Six and two of them were evaluated twice and three times, respectively, at baseline and within 12 months from the start of the treatment, for a total of 30 US examinations. Nine of them were affected by DM, six by PM, four by anti-synthetase syndrome (ASS) and one by scleromyositis (SM). Eleven subjects underwent thighs MRI, while in those remaining diagnosis was made with muscle biopsy or clinical findings. Clinical features of the patients, as well as US and MRI findings, are summarized in Table 2.

The sum of muscle oedema of both thighs positively correlated with PhGA ($r = 0.53$; $P = 0.002$), serum myoglobin ($r = 0.51$; $P = 0.02$) and PD signal ($r = 0.61$; $P = 0.0003$) and negatively with atrophy ($r = -0.36$; $P = 0.04$) and disease duration ($r = -0.54$; $P = 0.002$). The sum of PD score of both thighs was positively related to PhGA ($r = 0.69$; $P < 0.0001$) and negatively to disease duration ($r = -0.45$; $P = 0.01$).

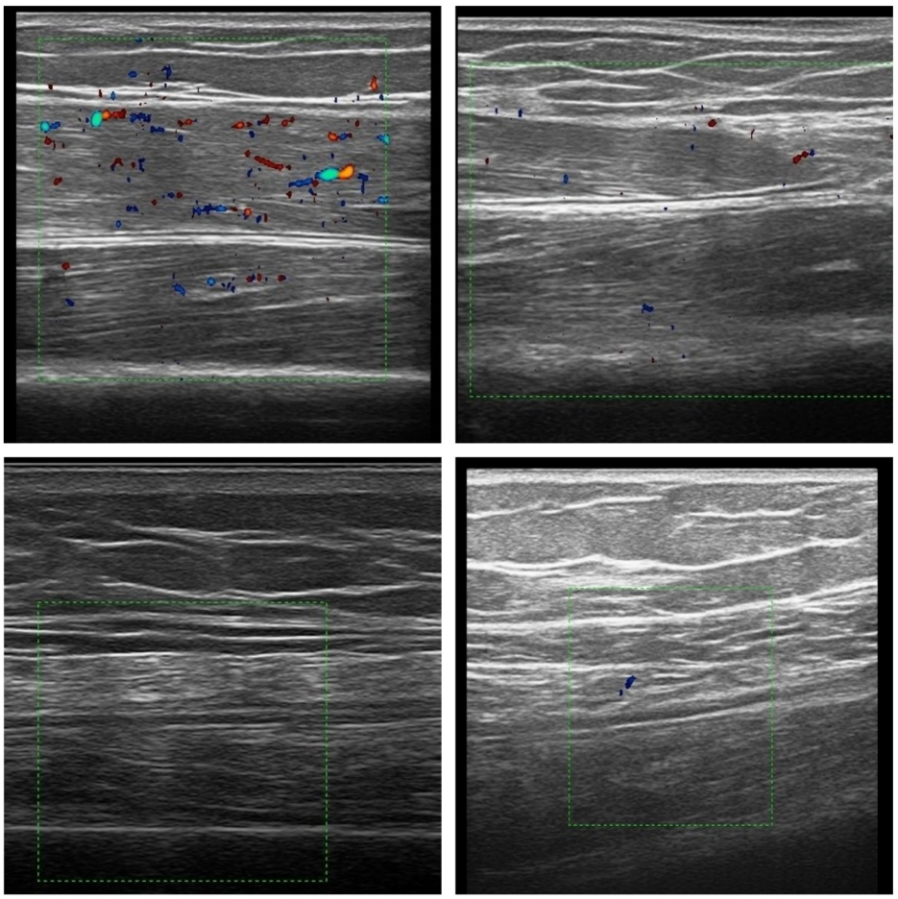
The overall sum of oedema and PD for both legs evidenced a positive correlation with PhGA ($r = 0.67$; $P < 0.0001$) and myoglobin ($r = 0.49$; $P = 0.03$) and a negative correlation with disease duration ($r = -0.55$; $P = 0.001$).

The sum of muscle atrophy correlated positively with MDI ($r = 0.71$; $P < 0.0001$), age ($r = 0.50$; $P = 0.004$) and disease duration ($r = 0.38$; $P = 0.03$) and negatively with oedema ($r = -0.36$; $P = 0.04$). (Supplementary Table S1, available at *Rheumatology* online).

The single-thigh sub-analysis (Supplementary Table S2, available at *Rheumatology* online) evidenced an inverse correlation between PD score and MMT (Right thigh: $r = -0.46$; $P = 0.009$. Left thigh: $r = -0.38$; $P = 0.03$). The sum of oedema, atrophy and PD of each leg, compared with MMT of the corresponding leg, evinced a high inverse correlation for both right ($r = -0.51$; $P = 0.004$) and left thigh ($r = -0.43$; $P = 0.01$).

A direct correlation was found between MRI findings of active IIM (e.g. muscle and fascial oedema and contrast enhancement) and both oedema ($r = 0.5245$; $P = 0.0122$) and PD ($r = 0.5106$; $P = 0.0152$). A non-statistically significant correlation was conversely found between MRI and US findings of atrophy.

Fig. 1 Different Power Doppler (PD) findings in longitudinal anterior scans of the thigh



Clockwise: PD 3 in a patient with a recent diagnosis of anti-Mi2 DM; PD 2 in the same patient after 1 month of treatment with steroids and methotrexate; PD 1 in a patient affected by anti-SAE DM, with a suspected disease flare; PD 0 in a patient affected by an advanced PM diagnosed in 2000, currently not in treatment.

An excellent inter-observer agreement was achieved for all three items: k-value was 0.893 (95% CI: 0.761, 1.025) for atrophy, 0.857 (95% CI: 0.698, 1.016) for oedema, and 0.954 (95% CI: 0.898, 1.011) for PD.

Discussion

The availability of new diagnostic procedures in IIM has dramatically modified the approach of the clinicians facing such rare, difficult-to-diagnose and difficult-to-treat [2] conditions, still strongly related to serological and histological findings.

Among the various imaging procedures employed to date in IIM, MRI has a paramount role, in terms of available evidences and increasing experience. Nevertheless, MRI is a time-consuming, expensive, and not always feasible procedure, particularly in the case of patients in whom an urgent diagnosis is required; for such reasons, MRI is still far from being included in the common clinical practice [7].

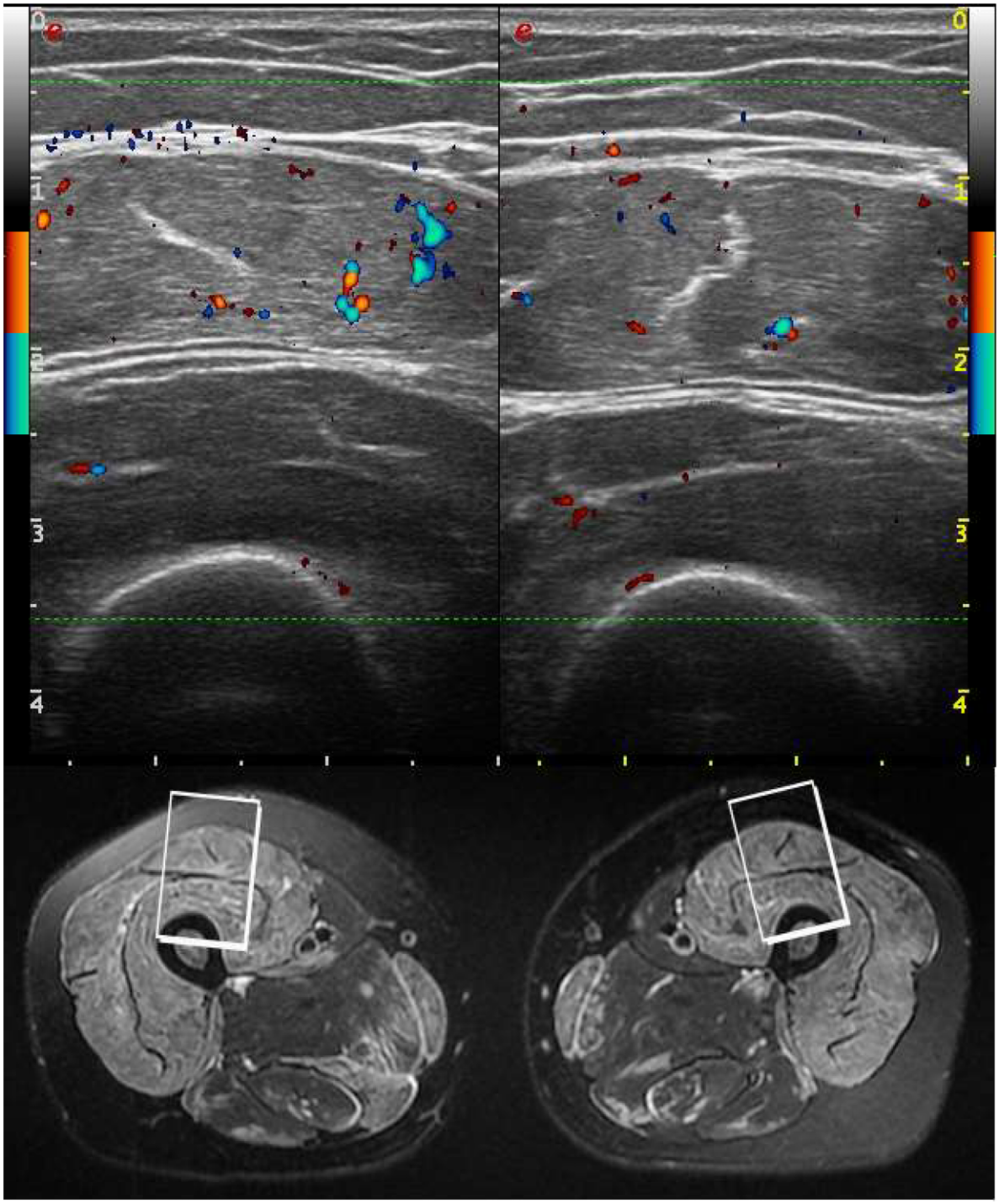
Similarly, ^{18}fluorodeoxyglucose positron emission tomography (FDG-PET) has been recently proposed in the evaluation of IIM [14], as an elevated uptake of tracer may correlate with an inflammatory condition. Nevertheless, the routine use of FDG-PET is burdened by the high cost and exposure to ionizing radiation.

If uncertainty exists about the preferred noninvasive diagnostic procedures to be employed at diagnosis, no clear-cut guidelines or recommendations exist about the optimal follow-up of patients affected by IIM; signs or symptoms are often treacherous and distinguishing between a loss of strength due to active inflammation from steroid-induced and/or immobilization myopathy may be difficult [15, 16].

Similarly, markers of inflammation or myonecrosis may underestimate a concomitant disease activity in case of normal or slightly elevated values at baseline, while antibodies, despite being particularly useful at diagnosis, do not seem to correlate with disease activity.

FDG-PET, although interestingly correlated to disease activity [17], may not be able to distinguish an inflammatory condition from reparative processes arising after the resolution of the disease [18]. Finally, no robust data

Fig. 2 Ultrasonography (US) and MRI of a recently diagnosed DM in a 33-year-old female



The patient presented with a marked elevation of serum creatine kinase (4817 UI/l) and myoglobin (1296 UI/l) and was not in treatment at the time of US and MRI. PD was graded 3 in both thighs, while oedema 2 in the right and 3 in the left thigh. MRI (axial scans, STIR T2) showed diffuse and limited oedema of quadriceps and hamstrings, respectively. The boxes within MRI images indicate the areas covered by the sonographic scans.

exist about the use of MRI during follow-up, which can hardly be proposed as a suitable follow-up technique, due to the high cost and low availability in common clinical practice.

Muscle US, too, has been proposed for diagnosis and follow-up of patients affected by IIM, but conflicting data have limited its extensive use in trials and clinical practice [8].

Table 2 Clinical and US features of the patients

Patient	Sex	Age (years)	Duration of disease (months)	Oedema (R and L)>		Atrophy (R and L)		PD (R and L)		Antibodies	Diagnosis	MMT (R and L)		PhGA	MDI	MRI findings
1	F	33	0	2	3	0	0	3	3	Mi2 SSA	DM	7	8	4	0	Oedema, enhancement
–			1	2	2	1	1	2	2	Mi2 SSA	DM	8	9	3	0	–
2	F	54	5	1	1	2	2	3	3	TIF1g	DM	7	6	4	0	Oedema
–			6	1	1	3	3	3	3	TIF1g	DM	8	7	3	1	–
3	F	41	24	0	0	0	0	0	0	JO1 SSA	ASS	10	10	0	0	–
4	F	27	48	0	0	0	0	0	0	PL7	ASS	10	10	0	0	Normal findings
–			54	3	0	0	0	2	0	PL7	ASS	9	10	2	0	–
5	F	61	48	0	1	2	2	0	1	JO1 SSA	ASS	9	8	2	3	Atrophy
6	F	66	1	3	3	2	2	3	3	MDA5	DM	8	7	4	0	Diffuse oedema, enhancement, slight atrophy
–			6	1	1	3	3	1	1	MDA5	DM	10	10	2	4	–
–			12	1	1	3	3	1	0	MDA5	DM	10	10	1	4	–
7	F	55	120	0	0	3	3	1	1	Pm/Scl 75	DM	6	4	3	3	–
8	F	80	36	1	1	3	3	1	2	TIF1g	DM	8	9	2	3	–
9	F	52	156	2	2	3	3	1	1	–	PM	8	9	2	3	Focal oedema, atrophy
–			160	1	1	2	2	1	1	–	PM	8	10	1	3	–
10	F	66	240	1	0	3	3	1	0	–	PM	2	2	4	10	Severe atrophy
11	M	51	120	1	1	1	1	1	1	JO1 SSA	ASS	10	10	0	0	–
12	M	67	72	1	1	1	1	1	1	NXP2	PM	10	10	1	5	Atrophy
13	F	71	0	3	2	1	1	3	1	TIF1g	DM	7	9	3	0	–
–			1	1	1	2	2	2	0	TIF1g	DM	8	9	2	0	–
–			3	1	0	2	2	1	0	TIF1g	DM	8	9	1	0	–
14	F	82	60	1	1	3	3	2	2	NXP2	PM	5	5	4	8	–
15	F	55	24	1	1	2	2	0	1	SRP	PM	10	10	0	2	Slight oedema without enhancement
16	F	22	12	2	3	0	0	2	2	Pm/Scl 75–100	PM	8	7	3	0	Focal oedema and diffuse enhancement
17	F	69	312	1	1	2	2	0	0	Pm/Scl 75	SM	10	10	1	5	–
18	F	56	24	2	2	2	2	2	1	SAE	DM	4	5	3	5	–
19	F	55	3	2	2	0	0	3	2	MDA5	DM	8	9	4	0	–
–			4	2	1	0	0	2	0	MDA5	DM	9	10	3	0	–
20	F	58	1	3	3	0	0	2	1	TIF1g	DM	3	3	4	0	Diffuse oedema, enhancement, muscle tear
–			2	2	1	0	0	0	0	TIF1g	DM	5	5	3	0	–

ASS: anti-synthetase syndrome; F: female; L: left; M: male; MDI: myositis damage index; MMT: Manual Muscle Test; PD: Power Doppler; PhGA: physician global assessment; R: right; SM: scleromyositis.

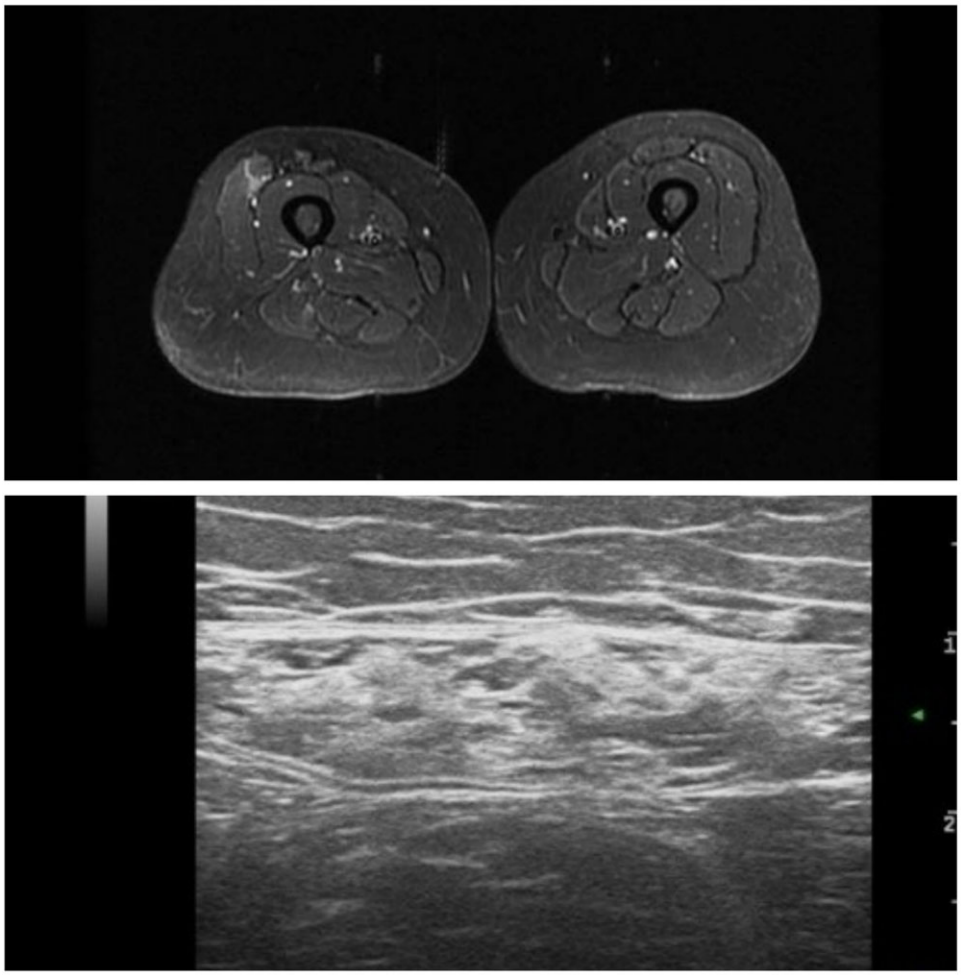
In normal conditions, long striated muscles appear as a relatively hypoechoic beam, surrounded by a thin hyperechoic line without acoustic shadow, corresponding to epimysium. Within the muscle, perimysium appears as a network of hyperechoic lines or spots, respectively in longitudinal or transverse scan [8, 12]. In the case of fatty replacement or fibrosis, muscle appears hyperechoic [19] and the bone below is less distinguishable: Heckmatt 4-point scale, although outdated, is still employed for the quantification of such US findings [20]. Conversely, uncertainty relies on how an inflamed muscle should appear on US: many authors have described a hyperechoic pattern [8, 21–24], similar to the one evidenced in case of fibrosis and fat replacement, while others suggest that intramuscular oedema should look hypoechoic [25].

Thus, due to such deep differences and uncertainties in the assessment of the very first phases of IIM, US alone is far from being considered a reliable technique, particularly if compared with MRI, in which muscle oedema is clearly characterized [26].

For these reasons, more advanced procedures have been variously proposed in order to augment sensitivity and specificity of US: contrast-enhanced US (CEUS) has displayed a 71% sensitivity and 91% specificity in PM and DM, slightly lower than MRI, whose sensitivity and specificity were 88% and 100%, respectively [27]. Nevertheless, no other papers to date have further investigated the role of CEUS, which is also burdened by a low feasibility in common clinical practice.

Similarly, only a few papers have investigated the role of PDUS in IIM. In 2001, Meng et al. [9] evaluated 37 IIM patients, all treated with immunosuppressants, and 6 healthy controls: PD positivity was significantly correlated with a shorter course of the disease, while no correlation was found between PD and serum creatine kinase (CK). Such findings were confirmed only by a more recent paper: in 2018, Sousa Neves et al. [12] performed muscle

Fig. 3 Axial anterior scan of the right thigh of a patient affected by PM



The patient was diagnosed 13 years ago and, currently not in treatment, was re-evaluated for a suspicion of disease flare. Atrophy and localized areas of oedema were found on thigh MRI (axial STIR scans). Ultrasonography evidenced areas of atrophy (GS 3) within rectus femoris muscle and focal areas of oedema (GS1) with loss of fibrillar texture.

US in 15 patients affected by IIM and, among various grey-scale alterations, one patient, with a shorter course of disease, presented moderate PD signal in both limbs.

Interestingly, a hypervascularization of the fascia was found in six out of seven patients affected by DM: in four of these, signs of fasciitis were found also on MRI. Nevertheless, no PDUS within muscle fibres was assessed nor evaluated, as it was beyond the scope of this paper [11].

Conversely, no PD signal was found by Adler [10], who proposed a 0–4 scale in six patients affected by IBM: none of them displayed a significant hyperemia.

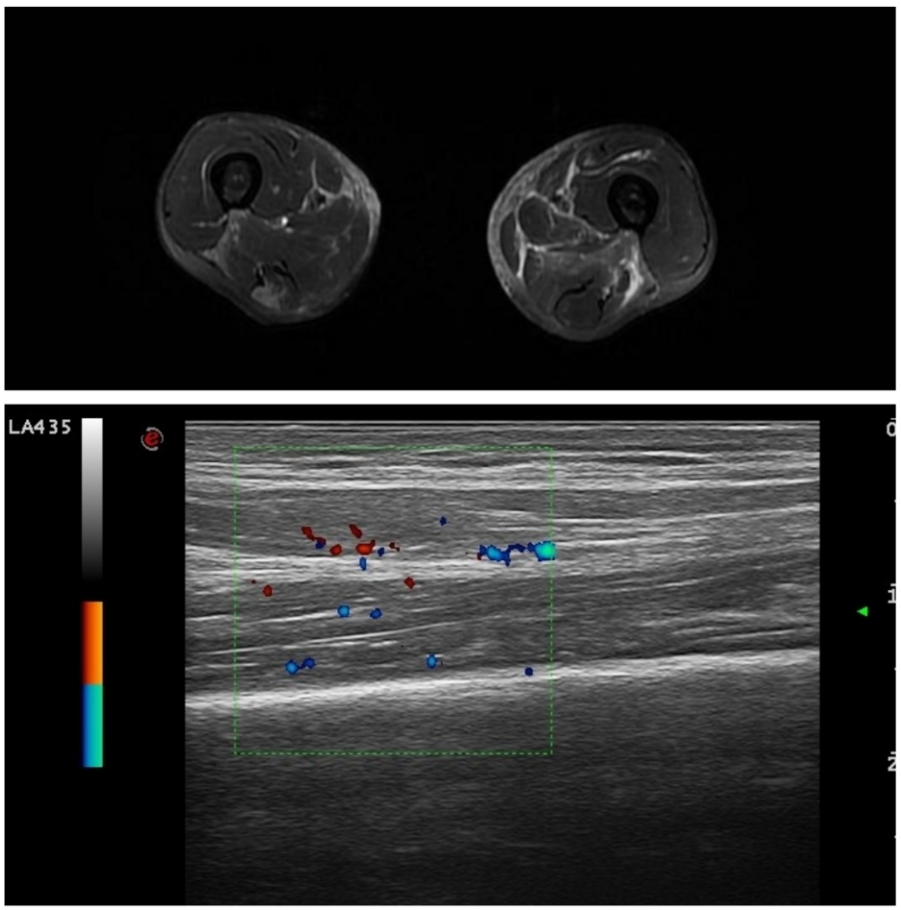
In this regard, due to the paucity and notable variability of the studies that have to date investigated the role of PDUS in IIM, we aimed to assess if this technique may still be considered a reliable diagnostic and prognostic tool in this subset of patients.

To the best of our knowledge, our study represents the largest cohort of patients affected by IIM who underwent both GS and PD US. According to our data, which should always be interpreted in the context of a real-life scenario, acute myositis, which on MRI appears as oedema, is frankly hypoechoic and swollen at US scan: this is underlined by the positive correlation between oedema and both PhGA and myoglobin. These findings do not confirm those, coming from other recent papers, that report a hyperechoic appearance of the inflamed muscle, while, conversely, are in line with the findings reported by Weber [25].

Conversely, oedema is negatively correlated with duration of disease: in case of extremely long duration of disease (Fig. 3), the advanced fibrosis may be de facto indistinguishable from concomitant oedema, even in case of a concomitant flare.

PD, too, reflects disease activity, as evidenced by the positive correlation with PhGA. PD is also positively correlated with oedema, suggesting that both these measures are concomitantly expression of early, active disease, as evidenced by the positive correlation of oedema and PD with MRI findings of active IIM. At the same time, both oedema and PD did not display any correlation with MDI, which is expression of aggressive and/or advanced IIM.

FIG. 4 Longitudinal anterior scan of the left thigh, evidencing atrophy (GS 3), oedema (1) and PD signal (3) in both rectus femoris and vastus intermedius



The patient, a 54-year-old female, suffered from an aggressive anti-TIF1γ DM, diagnosed 6 months before and active despite immunosuppression. Further examinations revealed the presence of a metastatic ovarian carcinoma. Diffuse atrophy and oedema are evidenced also on MRI (axial T2 scans).

Such findings were confirmed when oedema and PD were considered as a whole, as a positive correlation was found with serum myoglobin and PhGA and a negative correlation with disease duration.

Conversely, muscle hyperechoicity and reduced thickness, as expression of atrophy and fat replacement, is not correlated to PhGA or PD but only to MDI and duration of disease. This finding may underline the strict correlation between atrophic change of the muscle and destructive and systemic effects of IIMs (Fig. 4).

Finally, atrophy was found to be positively correlated to patient's age: this is not surprising, as muscle aging is physiologically associated with fat replacement and fibre thinning [28]. More importantly, no correlation was found between PD or oedema and patient's age, thus suggesting that PD and oedema remain useful in the diagnosis of IIM, also in elderly patients.

When thighs were evaluated singularly, coupling MMT with US findings, an inverse correlation was found between MMT and PD. Considering that a strong ($P < 0.0001$) inverse correlation was found also between MMT and PhGA, we may suggest that PD is a precious complementary tool in the clinical assessment of active IIM, which should always be employed in association with GS findings. Conversely, we did not find a statistically significant correlation between MMT and atrophy or oedema alone: a negative correlation was found only for oedema in the right thigh and when oedema, atrophy and PD were summed together. We may hypothesize that atrophy or oedema alone, as assessed on US, are not able to lead to a clinically evaluable muscle weakness, while only the sum of all findings, expression of a flare on a concomitant chronic myositis, is evaluable on clinical examination. Further data from larger cohorts may clarify whether this is due to the low size of our sample or to an intrinsic limitation of this procedure.

There are a number of limitations in our paper: first, the relatively low number of patients, so further confirmation from larger, multicentric studies, are needed. Second, our study was conducted in a real-life scenario, thus including subjects affected by various IIMs (DM, PM, ASS, SM) and displaying 11 different autoantibodies: further papers should evaluate if differences exist among such conditions, stratifying the patients according to autoimmune and histological profile. Third, only a limited number of patients contemporarily underwent MRI and US; it would be of interest to evaluate whether US may be an effective and reliable substitute for MRI, not only at diagnosis but also during follow-up. Finally, an intrinsic limit is the cross-sectional nature of our study: only a reduced number of patients underwent US during follow-up, thus further efforts should be addressed in the long-term monitoring, in order to assess US variations in IIM patients.

In conclusion, our study is the first to demonstrate the usefulness of muscle US, integrated with PD, in the assessment of IIM at different stages of the disease, describing how an inflamed muscle should appear. In our paper, we provide an easy and repeatable 0–3-point scale, which can be particularly useful in the stratification and follow-up of patients affected by different IIM.

We strongly believe that a better awareness of the potential role of US in these conditions, including, but not limited to, elastography and US-guided muscle biopsy [8] will meet many of the unmet needs of IIM in real-life clinical practice, in which the availability of more sensitive but less feasible diagnostic techniques, such as MRI, is still strongly limited.

Acknowledgements

E.C. and P.F. conceived the study; E.C., P.F. and S.G.A.K. wrote the manuscript; P.F. performed ultrasonography; E.C. and P.F. analysed ultrasonographic findings; S.G.A.K. and J.S. performed statistical analysis; E.C., C.B., M.B., F.B. and L.C. collected clinical data; L.C. and B.F. supervised the study. All authors approved the final manuscript.

Patients consent for publication

All patients gave their written consent for publication.

Funding: No specific funding was received from any bodies in the public, commercial or not-for-profit sectors to carry out the work described in this article.

Disclosure statement: The authors have declared no conflicts of interest.

Data availability statement

All data relevant to the study are included in the article or uploaded as supplementary information.

Supplementary data

Supplementary data are available at *Rheumatology* online.

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