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ORIGINAL RESEARCH

Cardiovascular events in patients with myositis: results from a French retrospective cohort

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ABSTRACT

Introduction Idiopathic inflammatory myositis (IIM) are systemic diseases, including dermatomyositis (DM), inclusion body myositis (IBM), immune-mediated necrotising myopathy (IMNM), antisynthetase syndrome (ASSD) and overlap myositis (OM). Patients with IIM have an increased risk of premature death, largely due to cardiovascular events (CVE). The aim of this study was to describe specific and non-specific cardiac involvement in patients with IIM, and to assess the occurrence of CVE.

Methods We conducted a retrospective observational cohort study of patients with IIM from Saint Antoine University Hospital, Paris, between 1997 and 2020. Cardiac involvement was defined as abnormalities at baseline on ECG, Holter ECG, transthoracic echocardiography, cardiac MRI or elevated cardiac biomarkers. CVE were defined as heart failure due to ischaemia, arrhythmia or conductive block, inflammatory myocarditis or resuscitation department admission.

Results 78 patients were included (median age 49 years; 67% female); 33 (42%) had DM, 18 (23%) ASSD, 12 (15%) OM, 11 (14%) IMNM and 4 (5%) IBM. Cardiac involvement at diagnosis was present in 12 (15%) patients; 15 (19%) had a CVE during follow-up. Patients with versus without cardiac involvement at diagnosis were more likely to present a CVE (6 (50%) vs 9 (14%); p=0.01). Median (IQR) time to CVE was shorter in patients with cardiac involvement (9 (0–34) vs 84 (26–156) months; p<0.01).

Conclusion Patients with cardiac involvement at myositis diagnosis are at increased risk of CVE and experience them earlier than patients without and should be carefully followed up, particularly during the first months after diagnosis.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Patients with idiopathic inflammatory myositis have an increased risk of premature death, largely due to cardiovascular events (CVE).

WHAT THIS STUDY ADDS

⇒ Patients with cardiac involvement at the time of myositis diagnosis have a significantly increased risk of CVE, and experience CVE earlier in the disease course than patients without cardiac involvement.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ Our findings emphasise the importance of carefully monitoring patients with cardiac involvement, particularly during the first months after diagnosis, and even if cardiac involvement is not specific to myositis disease.

INTRODUCTION

Myositis comprises a group of rare, heterogeneous diseases first described as polymyositis.[1] Since then, knowledge on idiopathic inflammatory myositis (IIM), its terminology and its classification have evolved, leading to the current classification of dermatomyositis (DM), inclusion body myositis (IBM), immune-mediated necrotising myopathy (IMNM), antisynthetase syndrome (ASSD) and overlap myositis (OM).[2 3] Muscle weakness is the most frequent symptom, but various organs can be involved including the skin, lungs, heart and joints.[4 5] Cardiac involvement in myositis is rare but is a leading cause of death.[6–8] Subclinical cardiac anomalies assessed on ECG, transthoracic echocardiography (TTE) and cardiac MRI (CMR) can be frequent, but evidence from large prospective studies is needed to determine the clinical impact of these findings.[9 10] Patients with myositis are at increased risk of death compared with the general population, with cardiovascular events (CVE) being a key cause.[8 11–13] Several studies have reported an increased rate of coronary artery atherosclerosis in patients with myositis,[13–15] but the prevalence of these risk factors, along with their management and clinical outcomes, are uncertain.

The aim of this retrospective cohort study of patients with IIM was to describe specific and non-specific cardiac involvement in patients with IIM, and to assess the occurrence of CVE.

Patients and methods

Study design

We conducted a monocentric retrospective observational cohort study of patients with IIM from the Department of Internal Medicine, Saint Antoine University Hospital, a tertiary hospital in Paris, France. We followed the Strengthening the Reporting of Observational Studies in Epidemiology recommendations to design our methodology and draft our manuscript.[16]

Population

Patients with IIM were identified by reviewing electronic medical records using diagnostic CIM 10 codes for IIM. Data from 1997 to December 2021 were collected. Patients aged 18 years and older at the time of IIM diagnosis were enrolled. Diagnosis of IIM was assessed according to the international classification criteria.[2] Medical files were retrieved (CK) and patients were classified as having DM, IBM, IMNM, ASSD or OM.[2 3] Patients with infectious or drug-induced myositis were excluded.

Data collection

The baseline was defined as the time of IIM diagnosis. The time of CVE was defined as the date of the first CVE if one occurred. The follow-up period was defined as the time between the date of diagnosis and the date of the CVE or the date of the last medical contact. We collected data on demographics, clinical and biological features, treatment related to IIM and occurrence of CVE. Recorded variables included sex, age, cardiovascular risk factors, history of cardiovascular diseases and treatments, features of IIM. Data were collected at baseline, occurrence of CVE and last medical contact.

Cardiac involvement

To define cardiac involvement at baseline, we first defined the significant abnormalities on ECG, Holter ECG, TTE, CMR and cardiac biomarkers. Significant abnormalities on ECG and Holter ECG were arrhythmias, conduction abnormalities (other than first-degree atrioventricular block) and signs of myocardial ischaemia (acute or sequela). Echocardiographic abnormalities were left ventricular dysfunction (left ventricular ejection fraction <40%), significant circumferential pericardial effusion and pulmonary hypertension confirmed by right heart catheterisation. Abnormalities on CMR were the presence of myocarditis (according to Lake and Louise criteria[17]), localised hypokinesia, signs of infarction and significant pericardial effusion. Clinical features of cardiac involvement were defined as clinical symptoms of heart failure (dyspnoea, orthopnoea, leg oedema, jugular venous distention and auscultation crepitation without other explanation) and chest pain suggesting angina. Two independent investigators (CK and AM) retrospectively reviewed the data to characterise cardiac involvement as 'specific cardiac involvement' (patients whose cardiac involvement was directly related to the inflammatory myopathy) or 'non-specific cardiac involvement' (when another cause was present that could explain cardiac abnormalities, such as ischaemic or hypertensive heart disease). Cardiac involvement was classified as clinical or subclinical.

Cardiovascular events

CVE were defined as the occurrence during follow-up of acute myocardial infarction, acute heart failure due to second- or third-degree atrioventricular block, acute heart failure due to a documented arrhythmia, occurrence of inflammatory myocarditis absent at diagnosis assessed on CMR, and admission to a resuscitation department for a cardiac or circulatory reason.

Statistical analysis

Continuous variables are described as medians with IQRs and binary variables as numbers (percentages). Continuous variables and binary variables were compared using Kruskal-Wallis tests and Fisher's tests, respectively. Cumulative incidence curves of CVE were generated using the Kaplan-Meier method and were compared using the log-rank test. The follow-up period was defined as the time between the date of diagnosis and the date of the CVE or the date of the last medical contact. Two-sided testing was used and $p<0.05$ was considered statistically significant. All analyses were performed using R software V.4.2.2 for Mac (Foundation for Statistical Computing, Vienna, Austria).

RESULTS

Study population

98 patients were identified in the database. Among them, 16 did not fulfil IIM diagnosis criteria, and 4 were excluded due to lack of data (figure 1). Finally, 78 patients were analysed; 67% were female and the median age was 49 (37–62) years at diagnosis. 33 patients (42%) had DM, 18 (23%) ASSD, 12 (15%) OM, 11 (14%) IMNM and 4 (5%) IBM. Median follow-up was at 72 (18–156) months.

Clinical characteristics at myositis diagnosis

The clinical features at diagnosis in the overall population and according to myositis subgroup are presented in table 1. Overall, 65% of patients experienced muscle weakness and 27% had a severe initial presentation with dysphagia. Extra-muscular involvement was present in 53 (68%) patients, of which 47% had interstitial lung disease, 70% cutaneous rash and 43% arthralgia. Five

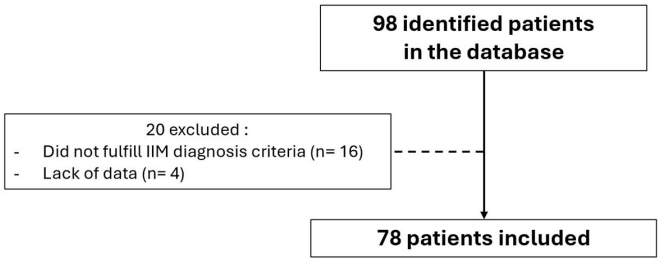

Figure 1 Flowchart. IIM, idiopathic inflammatory myositis.

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Table 1 Characteristics and treatment of patients at myositis diagnosis

Variable	Overall population (N=78)	ASSD (n=18)	DM (n=33)	IBM (n=4)	IMNM (n=11)	OM (n=12)	P value*
Women	52 (67)	12 (67)	23 (70)	2 (50)	7 (64)	8 (67)	0.95
Age, years	49 (37–62)	45 (43–54)	50 (33–61)	57.3 (55–61)	66 (44–72)	50 (31–56)	0.24
Cardiovascular risk factors	41 (53)						
Current smoker	13 (17)	3 (17)	6 (18)	1 (25)	1 (9)	2 (17)	0.95
Hypertension	20 (26)	4 (22)	9 (27)	1 (25)	2 (18)	4 (33)	0.93
Dyslipidaemia	15 (19)	3 (17)	6 (18)	2 (50)	3 (27)	1 (8)	0.42
Diabetes	11 (14)	2 (11)	5 (15)	1 (25)	2 (18)	1 (8)	0.90
History of myocardial infarction	3 (4)	0	3 (9)	0	0	0	0.37
Peripheral artery occlusive disease	1 (1)	0	1 (3)	0	0	0	0.85
Non-ischaemic cardiopathy	2 (3)	0	0	0	1 (9)	1 (8)	0.30
Cardiovascular treatment							
AEI or ARA	15 (19)	4 (22)	7 (21)	0	2 (18)	2 (17)	0.88
Beta-blocker	6 (8)	0	3 (9)	1 (25)	1 (9)	1 (8)	0.51
Statin	12 (15)	2 (11)	2 (6)	3 (75)	4 (36)	1 (8)	<0.01
Antiplatelet	7 (9)	0	3 (9)	0	2 (18)	2 (16)	0.38
Autoantibodies							
Anti-Mi-2	3 (4)	0	3 (9)	0	0	0	0.37
Anti-SAE	1 (1)	0	1 (3)	0	0	0	0.85
Anti-TIF-1-gamma	2 (3)	0	2 (6)	0	0	0	0.66
Anti-MDA-5	3 (4)	0	3 (9)	0	0	0	0.37
Anti-HMG-CoA	6 (8)	0	0	0	6 (55)	0	<0.01
Anti-SRP	4 (5)	0	0	0	4 (36)	0	<0.01
Anti-JO-1	13 (17)	13 (72)	0	0	0	0	<0.01
Anti-PL-7	2 (3)	2 (11)	0	0	0	0	0.14
Anti-Ku	1 (1)	0	0	0	0	1 (8)	0.23
Anti-PM-Scl	2 (3)	0	0	0	0	2 (17)	0.15
Anti-RNP	4 (5)	0	0	0	0	4 (33)	<0.01
Anti-DNA	2 (8)	0	1 (3)	0	0	1 (8)	0.65
Seronegative	31 (40)	0	22 (66)	4 (100)	1 (9)	4 (33)	<0.01
Organ involvement							
Lung	25 (32)	15 (83)	6 (18)	0	1 (9)	3 (25)	<0.01
Muscle weakness	51 (65)	8 (44)	25 (76)	3 (75)	9 (82)	6 (50)	0.10
Arthralgia	23 (30)	11 (61)	8 (24)	0	1 (9)	3 (25)	0.01

Continued

Table 1 Continued

Variable	Overall population (N=78)	ASSD (n=18)	DM (n=33)	IBM (n=4)	IMNM (n=11)	OM (n=12)	P value*
Dysphagia	21 (27)	2 (11)	11 (33)	0	3 (27)	5 (42)	0.22
Cutaneous rash	37 (47)	8 (44)	24 (73)	0	3 (27)	2 (17)	<0.01
Active neoplasia	5 (6)	1 (6)	4 (12)	0	0	0	0.06
Laboratory values							
Troponin≤10 ng/L	10 (10–41)	10 (10–432)	10 (10–35)	NA	10 (10–18)	10 (10–185)	0.87
B-type natriuretic peptide≤100 ng/L	32 (17–89)	59 (21–102)	33 (15–63)	NA	32 (29–89)	22 (17–33)	0.95
C reactive protein≤5 mg/L	11 (4–31)	42 (35–64)	7.50 (4–17)	3 (3–3)	5 (4.5–14)	13 (8–24)	0.03
Creatine phosphokinase≤200 U/L	1504 (269–4558)	1400 (223–5801)	1333 (187–3851)	700 (434–1803)	4965 (1191–6000)	3006 (969–3193)	0.25
TTE findings							
LVEF, %	60 (60–60)	60 (60–60)	60 (60–60)	NA	60 (60–68)	58 (53–60)	0.13
Pericardial effusion	3 (4)	2 (11)	0	NA	0	1 (8)	–
Systolic PAP, mm Hg	26 (21–32)	25 (22–29)	24 (20–36)	NA	23 (22–25)	31 (29–34)	0.51
Treatments							
Corticoids	67 (86)	16 (89)	30 (91)	2 (50)	10 (91)	9 (75)	0.17
Methotrexate	31 (40)	6 (33)	12 (36)	1 (25)	7 (64)	5 (42)	0.48
Rituximab	20 (26)	7 (39)	7 (21)	0	3 (27)	3 (25)	0.49
Immunoglobulins	31 (40)	5 (28)	14 (42)	2 (50)	4 (36)	6 (50)	0.75
Cardiac involvement	12 (15)	5 (28)	2 (6)	0	0	5 (42)	<0.01
Specific	5 (6)	2 (11)	1 (3)	0	0	2 (17)	0.35
Non-specific	7 (9)	3 (17)	1 (3)	0	0	3 (25)	<0.01

Data are count (percentage) or median (IQR).
*Between myositis subgroups.
AEI, ACE inhibitors; ARA, angiotensin receptor antagonist; ASSD, antisynthetase syndrome; DM, dermatomyositis; IBM, inclusion body myositis; IMNM, immune-mediated necrotising myopathy; LVEF, left ventricular ejection fraction; NA, not available; OM, overlap myositis; PAP, pulmonary artery pressure; TTE, transthoracic echocardiography.

(6%) had neoplasia or recurrence of neoplasia. 41 patients (53%) had cardiovascular risk factors: 13 (17%) were active smokers, 20 (26%) had arterial hypertension, 15 (19%) dyslipidaemia and 11 (14%) diabetes. Three (4%) patients had a history of myocardial infarction, 3% had a known cardiac disease from other causes than ischaemia (hypertensive cardiopathy) and 1% had peripheral artery occlusive disease. ACE inhibitors or angiotensin receptor antagonists were taken by 15 (19%) patients at baseline, 6 (8%) were on a beta-blocker and 13 (17%) on another antihypertensive drug. 12 (15%) patients were taking a statin and 7 (9%) antiplatelet.

Data on biological features were available for 62 patients. Complete echocardiography data were available in 36 patients (8 with ASS, 17 with DM, 6 with IMNM and 5 with OM). During follow-up CMR were performed in 36 patients, 16 at myositis diagnosis (7 in patients with DM, 3 with ASS, 3 with IMNM and 3 with OM).

Cardiac involvement was present in 12 patients (15%). Five presented specific cardiac involvement, two of whom had clinical symptoms. The remaining seven patients presented non-specific cardiac involvement, three of whom had clinical symptoms. Baseline data for patients with and without cardiac involvement are shown in table 2. Patient-level data for those with cardiac involvement are shown in online supplemental file 1. Five (42%) patients with ASSD, 42% with OM and 17% with DM had cardiac involvement versus none with IBM or IMNM.

At myositis diagnosis, patients with cardiac involvement were more likely to present arterial hypertension (58% vs 20%; p=0.01). No additional difference was noted for the other cardiovascular risk factors, and no difference was found for lung, skin or articular manifestations. However, patients with cardiac involvement had a higher median systolic pulmonary artery pressure (p=0.04) and presented higher concentrations of acute phase reactants (C reactive protein 35 vs 7 mg/L, p=0.01; gamma globulins 18.5 vs 12 g/L, p=0.02). Rituximab and intravenous immunoglobulins were more likely to be used in patients with cardiac involvement (both p<0.01).

Follow-up data

At the last available follow-up visit, patients with cardiac involvement had significantly higher concentrations of serum troponin and B-type natriuretic peptide. They also had lower left ventricular function (left ventricular ejection fraction (LVEF) 48% vs 60%, p=0.004) and higher pulmonary artery pressure (47 (31–57) vs 26 (21–30) mmHg, p=0.019).

Cardiovascular events

Time-to-event curves in patients with and without cardiac involvement are shown in figures 2 and 3. During the study, 15 (19%) patients experienced a new CVE: 5 (6%) myocardial infarction, 3 (4%) heart failure from causes other than ischaemia, 5 (6%) resuscitation admission and 2 (3%) inflammatory myocarditis with specific cardiac involvement (table 3). Median time to CVE occurrence was 72 (13–144) months.

Patients with versus without cardiac involvement at diagnosis were more likely to present a CVE (6 (50%) vs 9 (14%); p=0.01). Patients with specific cardiac involvement at diagnosis did not have more CVE than those with non-specific cardiac involvement (2 (40% vs 13 (18%), respectively; p=0.53). CVE were more frequent in patients with versus without non-specific cardiac involvement at diagnosis (4 (57%) vs 11 (16%); p=0.03). Time to CVE was shorter in patients with versus without cardiac involvement (9 (0–34) vs 84 (26–156) months; log-rank test p<0.01) (figure 2). Time to CVE was shorter in the subgroup with specific cardiac involvement (log-rank test p<0.001; figure 3).

DISCUSSION

In this case series, we describe cardiac features in patients with myositis, with several notable results. First, the prevalence of overall cardiac involvement at diagnosis is rare, at 15%, and specific inflammatory heart disease is present in only 6%. Second, the risk of cardiovascular disease appears important, as 53% of patients at diagnosis had traditional risk factors and 19% experienced a CVE during follow-up. Third, the risk of CVE appears higher and occurs earlier when cardiac involvement is present at diagnosis.

The demographic and clinical characteristics of our cohort of 78 patients with IIM from a French tertiary hospital are consistent with the recent European literature.[6 7 10] Indeed, our data confirm the preponderance of women (67%) and the median age at diagnosis of 49 years. DM was the most common subgroup, followed by ASSD and OM, whereas IMNM and IBM were infrequent.

The prevalence of cardiac involvement in patients with myositis ranges from 6% to 75% in the literature, highlighting the lack of consensus over the definition.[6 8 15 18] This heterogeneous rate is explained by differences in the inclusion criteria, the definition of cardiac involvement and the modalities used to detect cardiac involvement. Clinical cardiac involvement, represented mostly by heart failure, is rare in patients with myositis.[19] Subclinical changes are more frequent in series and are evolving with the availability of more sensitive modalities such as the use of strain in TTE and access to CMR.[19 20] Myocardial biopsies have shown similar inflammatory changes than in muscle biopsies, suggesting a specific inflammatory involvement of the cardiac muscle in myositis-associated myocarditis.[6 21] Currently, CMR is increasingly used in suspected cardiac involvement of IIM and can reveal a wide range of anomalies.[9] More precisely, mapping T1 and T2 relaxation time parameters have shown good performances in distinguishing patients with specific cardiac involvement in IIM from healthy patients.[22] Interestingly, a recent Chinese study by Sun *et al* published in 2021 included 51 newly diagnosed IIM patients without

Table 2 Characteristics and treatment of myositis patients with and without cardiac involvement at diagnosis

Variable	Cardiac involvement (n=12)	No cardiac involvement (n=66)	P value
Women	7 (58)	45 (68)	0.74
Age, years	55 (48–65)	49 (36–61)	0.13
Myositis subgroup			0.01
ASSD	5 (42)	13 (20)	–
DM	2 (17)	31 (47)	–
IBM	0	4 (6)	–
IMNM	0	11 (17)	–
OM	5 (42)	7 (11)	–
Autoantibodies			
Anti-Mi-2	2 (17)	1 (2)	0.09
Anti-SAE	0	1 (2)	1
Anti-TIF-1-gamma	1 (8)	2 (3)	0.95
Anti-MDA-5	0	3 (5)	1
Anti-HMG-CoA	0	6 (9)	0.61
Anti-SRP	0	4 (6)	0.87
Anti-JO-1	3 (25)	10 (15)	0.42
Anti-PL-7 or PL-12	1 (8)	1 (2)	0.70
Anti-Ku	0	1 (2)	1
Anti-PM-Scl	1 (8)	2 (3)	0.95
Anti-RNP	0	4 (6)	0.73
Anti-DNA	0	2 (3)	1
Anti-SSA	2 (17)	5 (8)	0.64
Seronegative	3 (25)	28 (42)	0.42
Cardiovascular risk factor			
Active smoker	2 (17)	11(17)	1
Arterial hypertension	7 (58)	13 (20)	0.01
History of myocardial infarction	2 (17)	1 (2)	0.09
Diabetes	2 (17)	9 (14)	1
Dyslipidaemia	4 (33)	11 (17)	0.34
Laboratory values			
C reactive protein, mg/L	35 (10–72)	7 (4–20)	0.01
Gamma globulins, g/L	18.5 (15-19)	12 (10–16)	0.02
Creatine phosphokinase, U/L	2783 (1183–3354)	1400 (231–4773)	0.59
Troponin, ng/L	120 (10–445)	10 (10–12)	0.07
B-type natriuretic peptide, ng/L	98 (24.75, 209.00)	29 (13–56)	0.02
Organ involvement			
Muscle weakness	6 (50)	45 (68)	0.38
Dysphagia	4 (33)	17 (26)	0.85
Lung involvement	6 (50)	19 (29)	0.27
Arthralgia	4 (33)	19 (29)	1
Cutaneous involvement	5 (42)	32 (49)	0.90
Active neoplasia	1 (8)	2 (3)	0.76
TTE findings			
LVEF, %	60 (50–60)	60 (60–61)	<0.01
Circumferential pericardial effusion	3 (25)	0	0.17
Systolic PAP, mm Hg	31 (27–39)	23 (20–27)	0.04

Continued

Table 2 Continued

Variable	Cardiac involvement (n=12)	No cardiac involvement (n=66)	P value
Treatment at baseline			
Corticoids	11 (92)	56 (85)	0.86
Methotrexate	3 (25)	28 (42)	0.42
Rituximab	8 (67)	12 (18)	<0.01
Immunoglobulin	10 (83)	21 (32)	<0.01
Mycophenolate mofetil	3 (25)	3 (5)	0.06
Cardiovascular event during follow-up	6 (50)	9 (14)	0.01
Time delay to CVE, months	9 (0–34)	84 (26–156)	<0.01

Data are count (percentage) or median (IQR).
ASSD, antisynthetase syndrome; CVE, cardiovascular events; DM, dermatomyositis; IBM, inclusion body myositis; IMNM, immune-mediated necrotising myopathy; LVEF, left ventricular ejection fraction; OM, overlap myositis; PAP, pulmonary artery; TTE, transthoracic echocardiography.

cardiac involvement suspected, and found a 27.5% positive LGE rate.[23]

In our series, cardiac involvement at myositis diagnosis was identified in 15% of patients, 4% had clinical cardiac involvement and 6% had specific IIM-related involvement. The results from recent European national cohorts showed similar rates of cardiac involvement to ours: 11.5% in a 2021 Slovenian cohort[10] and 20% in a 2017 cohort from Spain.[12] Cardiac involvement was more frequent in patients with ASSD and OM, consistent with the literature. A Chinese study reported cardiac involvement in 25% of their ASSD cohort of 96 patients,[24] and an Italian study published in 2024, retrospectively analysed 53 patients with ASSD and identified myocarditis proved by CMR in 30% of the cases.[25] In ASSD patients, retrospective studies have shown worse survival in anti-JO-1 positive patients. Survival difference, however, appears to be related to interstitial lung involvement.[26 27]

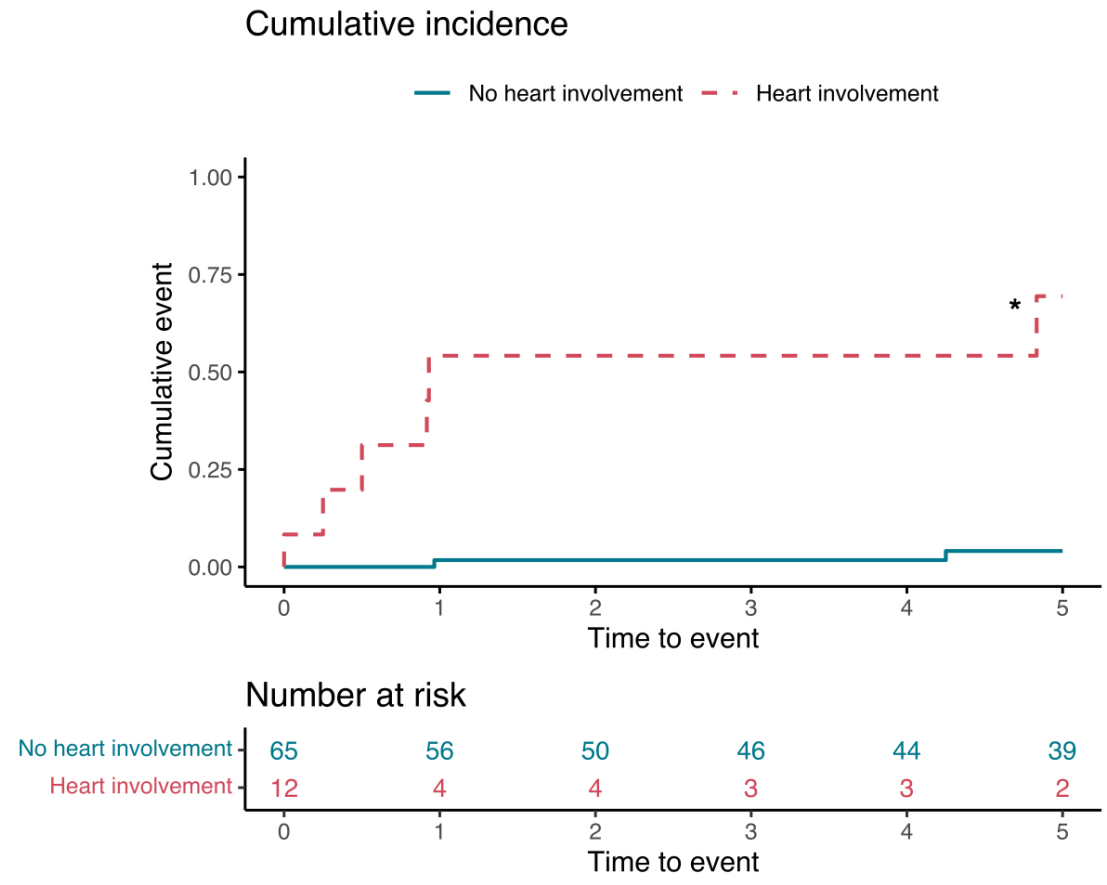

Figure 2 Time to cardiovascular event in myositis patients with cardiac involvement and without cardiac involvement at diagnosis. *P<0.001 using log-rank test.

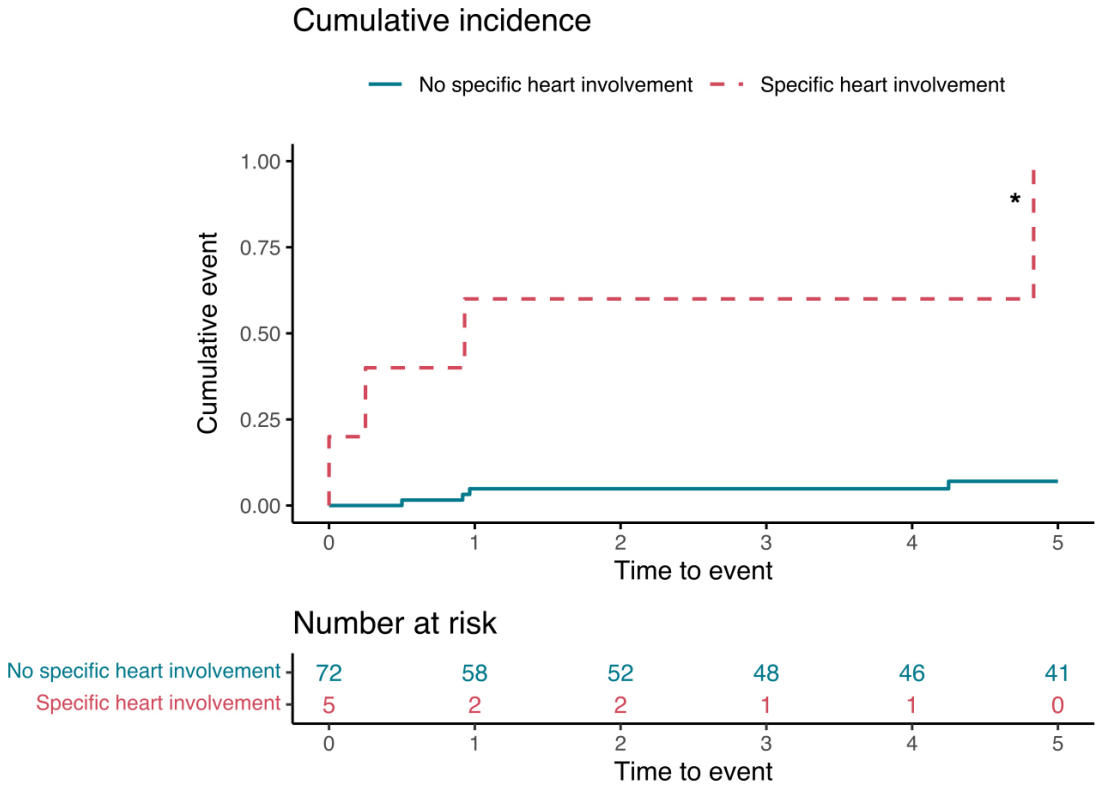

Figure 3 Time to cardiovascular event in myositis patients with specific cardiac involvement and without specific cardiac involvement at diagnosis. *P<0.001 using log-rank test.

To our knowledge, cardiac involvement in antisynthetase syndrome is not associated with any specific antibody.[25 28] In our cohort, we did not find any significant association between cardiac involvement and myositis-specific antibody.

A Hungarian cohort retrospectively examined clinical features in 39 patients with OM,[29] and identified cardiac involvement in 41%. Cardiac involvement was also associated with pulmonary hypertension, as highlighted in our cohort. In our study, none of the patients with IBM or IMNM presented cardiac involvement. Whereas IBM is not known to be associated with cardiac involvement,[30] it has been described in IMNM patients with positive anti-SRP in retrospective cohorts, with heterogeneous rates ranging from 2% to 69%.[31 32]

Patients with myositis, particularly in the first year after diagnosis, present a higher risk of traditional cardiovascular disease, such as atherosclerosis and myocardial infarction.[15 33 34] Two meta-analyses reported that IIM patients have a higher risk of presenting CVE independently of traditional risk factors.[15 35] Chronic inflammation has been described as a major player in the pathogenesis of CVE in rheumatic diseases such as rheumatoid arthritis and systemic lupus erythematosus,[36] and

Table 3 Cardiovascular events during follow-up

Variable	Overall population (N=78)	ASSD (n=23)	DM (n=42)	IBM (n=5)	IMNM (n=14)	OM (n=15)	P value*
Cardiovascular events	15 (19)	5 (28)	2 (6)	1 (25)	3 (27)	4 (33)	0.42
Myocardial infarction	5 (6)	1 (6)	1 (3)	1 (25)	1 (9)	1 (8)	–
Heart failure due to arrhythmia	2 (3)	0	1 (3)	0	1 (9)	0	–
Heart failure due to atrioventricular block	1 (1)	0	0	0	0	1 (8)	–
Myocarditis	2 (3)	0	0	0	0	2 (17)	–
Resuscitation admission	5 (6)	3 (17)	1 (3)	0	1 (9)	0	–
Overall death	3 (4)	0	0	0	2 (18)	1 (8)	0.06

Data are count (percentage).
*Between myositis subgroups.
ASSD, antisynthetase syndrome; DM, dermatomyositis; IBM, inclusion body myositis; IMNM, immune-mediated necrotising myopathy; OM, overlap myositis.

may explain the increased cardiovascular risk in patients with myositis. In our cohort, patients with cardiac involvement had a higher concentration of C reactive protein at baseline, which has also been reported in Chinese patients with ASSD.[24]

In our study, CVE occurred in 19% of the population, consistent with the rate from a 2017 Spanish cohort (22%).[12] A 2013 US study found that 20% of 50 000 hospitalisations in patients with dermatomyositis were for an atherosclerotic cardiovascular diagnosis or procedure.[37] Interestingly, a recent Czech cross-sectional study found no increase in cardiovascular risk in patients with myositis versus a matched control population.[38] This surprising result is explained in part by the absence of a specific tool to estimate cardiovascular risk in the myositis population.

Our study is subject to the limitations and bias inherent to a small, retrospective, single-centre study. Disease classification, CVE definitions and cardiac involvement remain heterogeneous, and require the establishment of an agreed definition. Our criteria were therefore defined based on the most recent publications, and we tried to build a coherent definition of cardiac involvement, while acknowledging that the non-specific features we describe may have unknown mechanisms and may not be directly related to myositis. We were unable to include certain interesting markers, such as global longitudinal strain on TTE, which reflects subclinical systolic ventricular dysfunction.[39] Indeed, these data were not available for many patients in our cohort, as they were often diagnosed before the common use of strain imaging. For most of our patients, data on troponin I was not available, leading us to use troponin T, despite its lack of cardiac specificity.[40] Considering myositis is a rare disease, we were able to include a significant number of patients in this single-centre study. Further multicentre studies with a larger cohort of patients are needed to confirm our results.

In conclusion, our research suggests that patients with cardiac involvement at the time of myositis diagnosis have a significantly increased risk of CVE. Furthermore, we show that these patients experience CVE earlier in the disease course than myositis patients without cardiac involvement. Given that CVE are a leading cause of morbidity and mortality in myositis, our findings emphasise the importance of carefully monitoring patients with cardiac involvement, particularly during the first months after diagnosis, and even if cardiac involvement is not specific to myositis disease.

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