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Cytosolic 5′-Nucleotidase 1A As a Target of Circulating Autoantibodies in Autoimmune Diseases

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Abstract

Objective—Prior investigations demonstrated that autoantibodies recognizing cytosolic 5′-nucleotidase 1A (NT5C1A) are found in 33–76% of patients with inclusion body myositis (IBM) but are observed only rarely in patients with polymyositis (PM). Thus, anti-NT5C1A may help distinguish IBM from PM. Although 4–21% of patients with dermatomyositis (DM) were shown to be anti-NT5C1A antibody positive, the clinical features of anti-NT5C1A–positive patients with DM have not been described. Furthermore, the prevalence of anti-NT5C1A antibodies in other rheumatic conditions has not been reported. This study was undertaken to define the prevalence and clinical features of anti-NT5C1A–positive patients with DM, PM, IBM, or other systemic autoimmune diseases.

Methods—We screened for anti-NT5C1A autoantibodies in patients with IBM, DM, PM, Sjögren's syndrome (SS), or systemic lupus erythematosus (SLE) and in healthy volunteers. Clinical characteristics were compared between patients who were anti-NT5C1A positive and those who were anti-NT5C1A negative.

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AUTHOR CONTRIBUTIONS

All authors were involved in drafting the article or revising it critically for important intellectual content, and all authors approved the final version to be submitted for publication. Dr. Mammen had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Study conception and design. Mammen.

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Results—Anti-NT5C1A autoantibodies were detected in 71 (61%) of 117 patients with IBM, 2 (5%) of 42 patients with PM, 2 (5%) of 42 healthy volunteers, 24 (15%) of 159 patients with DM, 10 (23%) of 44 patients with SS, and 13 (14%) of 96 patients with SLE. No anti-NT5C1A antibody–positive patients with SS or SLE had muscle involvement. Anti-NT5C1A–positive patients with IBM had a lower prevalence of rimmed vacuoles (62% versus 83% of antibody-negative patients; P = 0.02). No differences in the clinical characteristics of antibody-positive and antibody-negative patients with DM, SS, or SLE were observed.

Conclusion—Anti-NT5C1A is a common target of circulating autoantibodies, especially in IBM but also in several different autoimmune diseases. In SLE and SS, anti-NT5C1A autoreactivity is not associated with muscle disease.

Introduction

Circulating autoantibodies are common in patients with a variety of autoimmune diseases. In some cases, autoantibodies are tightly associated with a specific syndrome. For example, autoantibodies recognizing anti–histidyl–transfer RNA synthetase (i.e., anti–Jo-1) are specifically found in patients with a unique clinical phenotype known as the antisynthetase syndrome, which is characterized by myositis, interstitial lung disease, arthritis, fever, and/or characteristic skin lesions ([1,2]). In contrast, other autoantibodies are found in a broad spectrum of autoimmune diseases. These include autoantibodies recognizing Ro 52, which are found in the majority of patients with Sjögren's syndrome (SS) (67–75%) and also in a substantial minority of patients with systemic lupus erythematosus (SLE) (27–43%), patients with primary biliary sclerosis (28%), and patients with systemic sclerosis (19%) (for review, see ref. [3]).

The idiopathic inflammatory myopathies are a heterogeneous group of diseases including dermatomyositis (DM), polymyositis (PM), and inclusion body myositis (IBM) ([4]). Of note, 2 recent studies identified autoantibodies recognizing cytosolic 5′-nucleotidase 1A (NT5C1A) in 33–70% of patients with IBM but in only 4–14% of patients with PM, depending on the cut-offs chosen to define a positive result ([5,6]). In light of these findings, positive results of an assay for anti-NT5C1A antibodies may help distinguish patients with IBM from those with PM. Making this distinction is important, because patients with PM (but not those with IBM) are likely to benefit from immunosuppressive therapy.

Interestingly, anti-NT5C1A antibodies were also detected in 4–21% of patients with DM, but testing was performed in only relatively small cohorts of 36 patients ([5]) and 24 patients ([6]). In those studies, no clinical differences between IBM patients with and those without these antibodies were detected, and the clinical features of the anti-NT5C1A–positive patients with DM were not compared with those of antibody-negative patients. Furthermore, the prevalence of anti-NT5C1A autoantibodies in other connective tissue diseases has not been previously reported.

In this study, we confirmed the high frequency of anti-NT5C1A antibodies in patients with IBM compared with patients with PM, thereby validating a novel immunoblotting assay for detecting these antibodies. We observed that the prevalence of rimmed vacuoles on muscle biopsy was lower in antibody-positive IBM patients compared with antibody-negative IBM

patients. We also confirmed that anti-NT5C1A antibodies are present in 15% of patients with DM, but we could not identify a unique clinical phenotype associated with this immunospecificity. Furthermore, we observed that anti-NT5C1A autoantibodies were detectable in 14% of patients with SLE and in 23% of patients with SS. None of the anti-NT5C1A–positive patients with SLE or SS had muscle disease, and we could not identify antibody-associated clinical characteristics in these patients. Similar to Ro 52, NT5C1A appears to be a common target of the immune response in a variety of autoimmune diseases.

Patients and Methods

Patients and sera

Serum samples were collected from 117 consecutively enrolled patients with a diagnosis of IBM according to data-derived criteria ([7]) and/or "probable" or "definite" IBM according to the European Neuromusuclar Centre criteria ([8]), 42 patients with a diagnosis of probable or definite PM according to the criteria described by Bohan and Peter ([9],[10]), 96 patients with SLE, 44 patients with SS, and 42 healthy volunteers. We included sera from 159 of 160 patients with DM who were described in detail in a previous report ([11]); serum from 1 of these patients was not available for testing in the current study. Informed consent was obtained from all subjects, and all samples were obtained under the auspices of Johns Hopkins Institutional Review Board–approved protocols. Clinical information for all patients with DM, SLE, or SS was obtained by medical record review.

Anti-NT5C1A antibody assay

Lysates of HEK 293 cells transfected with NT5C1A and nontransfected HEK 293 cell lysates were obtained from Applied Biological Materials. Equal protein amounts of these lysates were electrophoresed on 8% sodium dodecyl sulfate–polyacrylamide gels, transferred to nitrocellulose membranes, and immunoblotted with either a positive control rabbit polyclonal antibody recognizing NT5C1A (Applied Biological Materials) or human sera diluted 1:1,000 for 1 hour. Human sera that recognized the ~43-kd NT5C1A protein in NT5C1A-transfected cells but not in nontransfected cells were considered to be positive for anti-NT5C1A antibodies.

Statistical analysis

The chi-square test or Fisher's exact test was used to compare frequencies, as appropriate. The unpaired *t*-test was used to compare means, and Wilcoxon's rank sum test was used to compare non–normally distributed continuous variables (expressed as the median and first and third quartiles) between 2 groups. *P* values less than or equal to 0.05 were considered significant.

Results

Validation of the anti-NT5C1A immunoblotting assay

We screened for anti-NT5C1A antibodies by immunoblotting patient sera against NT5C1A-transfected and nontransfected HEK 293 cell lysates (Figure 1). Sera that recognized the ~43-kd NT5C1A protein present in transfected (but not nontransfected) lysates were

considered positive for anti-NT5C1A autoantibodies. Consistent with observations in studies using other methods of anti-NT5C1A detection ([5,6]), the immunoblotting assay detected these autoantibodies in 71 (60.7%) of 117 patients with IBM, 2 (4.8%) of 42 patients with PM, and 2 (4.8%) of 42 healthy volunteers. The 2 anti-NT5C1A–positive patients with PM had proximal muscle weakness and did not have distal or asymmetric patterns of weakness consistent with IBM. Thus, sensitivity of the immunoblotting assay for anti-NT5C1A was 60.7% in IBM, and specificity was 95.2% when this assay was used to distinguish IBM from PM.

Lower prevalence of rimmed vacuoles in anti-NT5C1A–positive patients with IBM

In the original description of these autoantibodies, no clinical features in IBM patients were identified that correlated with the presence or absence of anti-NT5C1A autoantibodies ([5]). We did not identify demographic or clinical features that distinguished these 2 groups of IBM patients (Table 1), and none of the patients had documented improvement in muscle strength, regardless of treatment. However, rimmed vacuoles were more frequently observed in muscle biopsy specimens obtained from patients without anti-NT5C1A autoantibodies (83% versus 62% of patients with anti-NT5C1A autoantibodies; $P = 0.02$) (Table 1).

No association of anti-NT5C1A autoantibodies with a distinctive phenotype in patients with DM

We screened a cohort of 159 patients with DM and observed that 24 patients (15%) were anti-NT5C1A positive. We compared the demographic features of patients with and those without anti-NT5C1A antibodies and observed no significant differences in sex, race, age at diagnosis, or disease duration between the 2 groups (Table 2). We also compared several clinical features in patients with and those without anti-NT5C1A antibodies. In general, the clinical characteristics of the patients in both groups were remarkably similar.

Interestingly, among the 14 anti-NT5C1A–positive patients with DM in whom comprehensive myositis-associated autoantibody testing was performed, a myositis-related autoantibody was identified in 10 patients (71.4%) (4 with anti–Mi-2, 2 with anti–nuclear matrix protein 2, 2 with anti–PM-Scl, 1 with anti–Jo-1, and 1 with both anti–Mi-2 and anti–Tif-1).

Presence of anti-NT5C1A autoantibodies in patients with SLE and patients with SS

We next sought to determine whether anti-NT5C1A autoantibodies are present in patients with other autoimmune diseases, including those with relatively low rates of myositis. To this end, we screened sera from 96 patients with SLE and 44 patients with SS and observed that 13 (13.5%) and 10 (22.7%), respectively, were anti-NT5C1A positive. Among the patients with SLE, 5 (5%) did have myositis; however, none of these patients was positive for anti-NT5C1A. In the SLE group, there was no correlation between the presence of anti-NT5C1A antibodies and Raynaud's disease or interstitial lung disease (data not shown). None of the SS patients had myositis or muscle weakness, and there was no correlation between the presence of anti-NT5C1A antibodies and sex, positive lip biopsy results, positive SSA/SSB serology, a Schirmer's test result of 5 mm in 5 minutes, leukopenia, a low C4 level, hypergammaglobulinemia, rheumatoid factor status, or an antinuclear antibody

titer ≥1:320 (see Supplementary Table 1 available on the *Arthritis Care & Research* web site at http://online-library.wiley.com/doi/10/1002/acr.22600/abstract).

Discussion

The 5-nucleotidases are a family of 7 enzymes that dephosphorylate noncyclic nucleoside monophosphates to nucleoside and inorganic phosphate ([12]). These enzymes regulate the balance of intracellular nucleotide pools and probably play crucial roles in energy balance, metabolism regulation, and cell replication ([13],[14]). Of note, one member of the 5′-nucleotidase family, NT5C1A, was recently shown to be a prevalent target of the immune response in IBM, but not in PM ([5],[6]). Thus anti-NT5C1A antibodies may be useful in making a correct diagnosis of IBM, a disease that is commonly mistaken for PM.

Interestingly, anti-NT5C1A autoantibodies were also found at a prevalence of 4–21% in small cohorts of DM patients ([5],[6]). In the current study, we screened a large cohort of 159 patients with DM and confirmed the presence of anti-NT5C1A antibodies in 15% of these patients. Although we analyzed clinical and demographic information from these DM patients, we could not identify specific features that were associated with the presence of anti-NT5C1A autoantibodies. Rather, DM patients with and those without these antibodies were very similar, with nearly equivalent frequencies of DM rashes, weakness, fever, inflammatory arthropathy, Raynaud's phenomenon, mechanic's hands, interstitial lung disease, and calcinosis. This finding also parallels what has been observed in IBM patients, in whom no clinical differences between those with and those without NT5C1A autoreactivity have yet been described ([5]). In the current study, we did observe a modestly increased prevalence of rimmed vacuoles in patients without anti-NT5C1A antibodies; this histologic feature was not assessed in the prior studies. Given that we performed multiple comparisons and the *P* value was only 0.02, our findings should be considered preliminary until they are validated in other cohorts. However, we hypothesize that NT5C1A-positive patients may generate a specific immunologic response against muscle cells with a high content of rimmed vacuoles, considering that these vacuoles have been reported to have increased NT5C1A reactivity ([5]).

To determine whether antibodies against NT5C1A might be present in other autoimmune diseases, including those without prevalent muscle involvement, we screened for this autoantibody in cohorts of patients with SLE and patients with SS. Surprisingly, anti-NT5C1A autoantibodies were observed in 14% of patients with SLE and in 20% of patients with SS, none of whom had myositis. As in patients with DM, we were unable to identify specific disease features that were associated with anti-NT5C1A autoantibodies in either patients with SLE and those with SS.

Our results demonstrate that, as with anti–Ro 52 autoantibodies, anti-NT5C1A autoantibodies are found in a broad spectrum of autoimmune diseases. As the current study confirms, anti-NT5C1A autoantibodies are most common in IBM, reinforcing an emerging appreciation that this disease has an autoimmune component despite its resistance to traditional immunosuppressive approaches (for review, see ref. [15]). Although the mechanisms underlying the initiation of NT5C1A autoimmunity are unknown, it is

noteworthy that NT5C1A, which normally has a cytosolic localization, is found in areas of myonuclear degeneration and rimmed vacuoles in IBM but not in normal muscle ([5]). This has suggested the possibility that aberrant localization of NT5C1A might contribute to its antigenicity in IBM. Because patients with SS and patients with SLE without muscle involvement also have NT5C1A immunoreactivity, it would be of interest to study localization of NT5C1A in the target tissue of patients with these diseases.

The current study has several limitations. First, differences between anti-NT5C1A–positive and anti-NT5C1A–negative patients with DM may not have been discovered because the relevant features were not analyzed. For example, it has been reported that anti–Ro 52–positive patients with type 1 autoimmune hepatitis have especially poor prognoses ([16]); long-term outcome data for our patients with DM were not available to allow a similar comparison. Second, because we studied a relatively small cohort of patients with SS (n = 44), clinically relevant associations may have been missed. For example, there was a trend toward anti-NT5C1A–positive SS patients having a white blood cell count of <4,000/μl (30% versus 6% of anti-NT5C1A–negative patients; $P = 0.07$). If this represents the actual prevalence of leukopenia in these populations, our sample size would have needed to be more than twice as large (n = 95) to have an 80% chance of showing a true association with a P value of less than 0.05 (for additional information see Supplementary Table 1, available on the *Arthritis Care & Research* web site at http://onlinelibrary.wiley.com/doi/10/1002/acr.22600/abstract). Finally, we categorized patients as being either positive or negative for anti-NT5C1A autoantibodies, and it may be that specific autoantibody titers are correlated with distinct disease features.

Notwithstanding these limitations, our study shows that anti-NT5C1A autoantibodies are common not only in patients with IBM and patients with DM but also in patients with other autoimmune diseases without muscle involvement. It remains to be determined how NT5C1A becomes an antigenic target and whether the localization and/or function of this protein may be aberrant in extra-muscular tissue targeted by the immune system.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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Significance and Innovations

- We demonstrate that anti–cytosolic 5′-nucleotidase 1A (anti-NT5C1A) antibodies are found in 14% of patients with systemic lupus erythematosus and 23% of patients with Sjögren's syndrome but are not associated with muscle disease or other known clinical characteristics in these patients.
- We found no unique demographic or clinical characteristics associated with the 15% of anti-NT5C1A–positive patients with dermatomyositis.
- We showed that anti-NT5C1A antibodies are common in a large cohort of patients with inclusion body myositis (IBM) (61%) but are rare in patients with polymyositis (5%), confirming that the presence of these antibodies may help differentiate patients with these 2 diseases.
- Rimmed vacuoles were more common in anti-NT5C1A–negative IBM patients than in antibody-positive IBM patients.

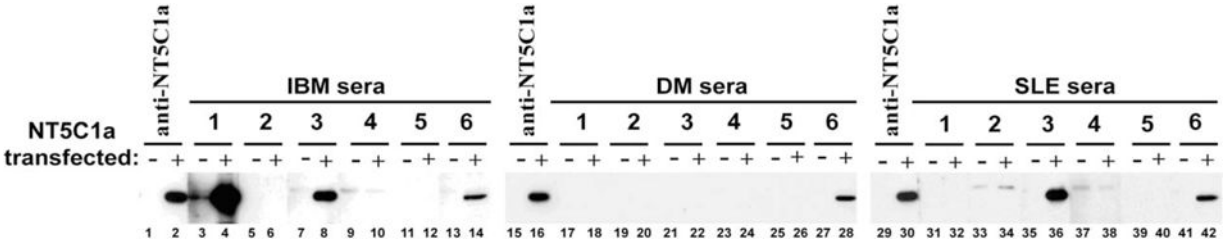

Figure 1.

An immunoblotting method for detecting autoantibodies recognizing cytosolic 5′-nucleotidase 1A (anti-NT5C1A) autoantibodies. Odd numbered lanes were loaded with lysates from nontransfected HEK 293 cells and even numbered lanes were loaded with lysates from HEK 293 cells transfected with NT5C1A. The first two lanes of each blot (1, 2, 15, 16, 29, and 30) were immunoblotted with rabbit anti-NT5C1A, demonstrating the presence of NT5C1A protein only in the transfected cell lysate. Sera from 3 IBM (#1, 3, and 6), 1 DM (#6), and 2 SLE (#3 and 6) subjects recognized proteins in the transfected but not nontransfected cell lysates; only these 6 sera were considered to be anti-NT5C1A positive.

Table 1

Demographic, clinical, and histologic characteristics of the patients with IBM according to anti-NT5C1A antibody status*

Characteristic	Anti-NT5C1A positive (n = 71)	Anti-NT5C1A negative (n = 46)	*P*
Female sex	20 (28)	16 (35)	0.4
Race			
White	54 (76)	36 (78)	0.8
African American	7 (10)	3 (7)	0.5
Other	10 (14)	7 (15)	0.9
Age at onset, mean ± SD years	58.9 ± 9.3	57.9 ± 10	0.6
Disease duration, mean ± SD years	7.8 ± 6.3	6 ± 4.3	0.1
Maximum creatine kinase level, median (IQR) IU/liter	642 (371–1,188)	578 (349–1,300)	0.7
Knee extensors weaker than hip flexors	37 (52)	29 (63)	0.2
Wrist flexors weaker than wrist extensors	48 (68)	35 (76)	0.3
Proximal weakness	30 (42)	23 (50)	0.4
Focal invasion	44 (64)	26 (58)	0.5
Endomysial inflammation	69 (97)	46 (100)	0.3
Rimmed vacuoles	44 (62)	38 (83)	0.02

*Values are the number (%) except where indicated otherwise. IBM = inclusion body myositis; anti-NT5C1A = anti–cytosolic 5′-nucleotidase 1A; IQR = interquartile range.

Table 2

Demographic and clinical features of DM patients according to anti-NT5C1A antibody status*

Characteristic	Anti-NT5C1A positive (n = 24)	Anti-NT5C1A negative (n = 135)	*P*
Sex			
Male	4 (17)	38 (28)	0.3
Female	20 (83)	97 (72)	0.3
Race			
White	20 (83)	111 (82)	1
African American	3 (13)	16 (12)	1
Asian	0 (0)	5 (4)	1
Other	1 (4)	3 (2)	0.5
Age at diagnosis, mean ± SD years	41.3 ± 15.5	45.4 ± 16.0	0.3
Disease duration, mean ± SD months	14.7 ± 15.0	28.5 ± 55.0	0.2
Gottron's papules or sign	17/24 (71)	98/134 (73)	0.8
Heliotrope rash	10/24 (42)	65/135 (48)	0.7
Weakness	23/24 (96)	123/135 (91)	0.7
Fever	2/21 (10)	23/130 (18)	0.5
Inflammatory arthropathy	4/19 (21)	36/131 (27)	0.8
Raynaud's phenomenon	6/21 (29)	40/127 (31)	1
Mechanic's hands	4/24 (17)	31/134 (23)	0.6
Interstitial lung disease	2/24 (8)	22/133 (17)	0.5
Calcinosis	3/23 (13)	18/134 (13)	1

*Values are the number of patients/number of patients assessed (%) except where indicated otherwise. DM = dermatomyositis; anti-NT5C1A = anti–cytosolic 5′-nucleotidase 1A.