



# Acute Pericarditis as the Initial Manifestation of Antisynthetase Syndrome: A Case Report

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## Abstract

Antisynthetase syndrome (AS) is a rare systemic autoimmune disease characterized by the presence of anti-tRNA synthetase antibodies. Cardiac involvement (pericarditis) in AS is uncommon. Here, we present the case of a young African female who presented with acute pericarditis as an initial manifestation of AS. She was diagnosed with anti-Jo1 antisynthetase syndrome based on Connors criteria. Treatment with corticosteroids and azathioprine improved symptoms. This case underlines the importance for clinicians to evoke the possibility of an antisynthetase syndrome in the case of acute pericarditis.

## Subject Areas

Rheumatology

## Keywords

Antisynthetase Syndrome, Acute Pericarditis, Initial Manifestation

## 1. Introduction

Antisynthetase syndrome (AS) is a rare and heterogeneous systemic autoimmune disease characterised by the positivity of anti-aminoacyl-tRNA synthetase (anti-ARS) autoantibodies and by the occurrence of a broad spectrum of clinical features involving many organs, including the muscle, joints, lung, and skin [1]. On the other hand, acute pericarditis is an inflammatory pericardial syndrome with or without pericardial effusion [2], an event lasting <4 to 6 weeks [3]. Pericarditis can be either an isolated form or a cardiac manifestation of a systemic disorder (autoimmune disease) [3].

However, Cardiac involvement (pericarditis) in antisynthetase syndrome is uncommon [4] and only a few cases have been previously reported [5]. Herein,

we present the case of a young African female who presented with acute pericarditis as an initial manifestation of AS.

The aim of this report is to acknowledge the importance of acute pericarditis as an initial manifestation of the antisynthetase syndrome.

## 2. Case Report

A 26-year-old Senegalese female with no significant past medical history has initially admitted to the cardiology department for acute chest pain that evolved for three weeks, this symptomatology was associated with a feeling of muscle weakness and polyarthralgia. She benefited from an electrocardiogram and a chest X-ray which came back normal. However, the echocardiography objectified a moderate circumferential pericardial effusion without other abnormalities. The blood test showed elevated C-reactive protein (CRP) 96 mg/l (normal < 6 mg/l).

The diagnosis of acute pericarditis was made in accordance with the European Society of Cardiology guidelines [2] (Table 1).

She has been initiated on colchicine 1 mg per day. However, the patient reported persistent muscular weakness and polyarthralgia with aggravation during the last week. Thus she was referred to our rheumatology department for etiological research of this symptomatology.

On admission, vital signs including body temperature were normal. Physical examination revealed bilateral proximal muscle weakness of the upper and lower extremities, myalgias and polyarthritides. She had no other symptoms including Raynaud's phenomenon, dry mouth or dry eye, oral ulcers, hair loss, night sweats, weight loss or dermal lesions.

Laboratory findings showed an elevated serum Creatine Kinase at 2130 IU/L (normal < 170), Anti-Jo1 was strongly positive and anti-SSA/Ro52 antibodies were positive, the antinuclear antibody was weakly positive with a titer of 1:100. Anti-Sm, anti-centromere, anti-DNA, anti-U1 RNP and rheumatoid factor were negative.

A diagnosis of Antisynthetase syndrome was made on Connors criteria (Table 2).

**Table 1.** Diagnostic criteria for acute pericarditis [2].

Inflammatory pericardial syndrome to be diagnosed with at least 2 of the 4 following criteria:
1) Pericarditic chest pain
2) Pericardial rubs
3) New widespread ST-elevation or PR depression on ECG
4) Pericardial effusion (new or worsening)
Additional supporting findings:
- Elevation of markers of inflammation (C-reactive protein, erythrocyte sedimentation rate, and white blood cell count)
- Evidence of pericardial inflammation by an imaging technique (CT, CMR)

**Table 2.** Connors criteria for diagnosis of antisynthetase syndrome [1].

Anti-aminoacyl-t RNA synthetase autoantibody plus one among:
Myositis (Bohan and Peter's criteria)
Arthritis (clinic, X-rays, self-report)
Interstitial lung disease
Raynaud's phenomenon
Mechanic' hands
Unexplained fever

We started therapy with Azathioprine (1 mg/kg/day) and prednisolone (0.5 mg/kg/day). After three months of treatment, the patient had an overall improvement in her symptomatology.

### 3. Discussion

Antisynthetase syndrome is a rare autoimmune disease [1], with a global prevalence estimated as 1-9/100,000 [6]. AS more frequently affects females (female to male ratio is estimated to be approximately 7:3) [6].

AS is characterized by the presence of autoantibodies targeting one of several aminoacyl tRNA synthetases [7].

Eight antisynthetase antibodies have been described thus far: anti-Jo-1 (anti-histidyl), anti-PL12 (anti-alanyl), anti-PL7 (anti-threonyl), anti-OJ (anti-isoleucyl), anti-EJ (anti-glycl), anti-KS (anti-asparaginy), anti-YRS/Ha (anti-tyrosyl), and anti-Zo (anti-phenylalyl) [8].

Anti-Jo-1 is the most common anti-synthetase, accounting for 60.3% - 72% of antisynthetase antibodies [9] [10]. Our patient was positive for anti-Jo1 and anti-SSA/Ro52. However, antibodies against Ro (including Ro52) are considered the most common type of association in antibodies in ARS-positive patients, occurring in 30% - 65% of cases [6].

The classic presentation triad of symptoms described in AS includes myositis, arthritis and interstitial lung disease. However, only a minority of patients exhibit the full triad at disease onset [8]. Our patient had muscular and joint manifestations.

Pericardial involvement is common in systemic lupus erythematosus, Sjogren's syndrome, rheumatoid arthritis and scleroderma [2], but is seen more rarely in anti-synthetase syndrome [4] [11].

Indeed, in one large cohort, the prevalence of pericarditis in antisynthetase syndrome is 1.7% [11].

Pericarditis rarely occurs as the initial manifestation of systemic autoimmune diseases [2], particularly during the anti-synthetase syndrome [12].

Our case is special because pericarditis in this disease is rare, and the initial presentation of pericarditis in the clinical picture is exceptional.

Corticosteroids are considered the first-line treatment in antisynthetase syn-

drome, but most of the time, other immunosuppressive agents are needed. Azathioprine or methotrexate are common first-line therapies in AS patients [13] [14]. Our patient was treated with prednisolone and azathioprine. Combination therapy with methotrexate and azathioprine should always be considered even when patients have failed to respond to either agent alone [14]. In refractory cases, treatment escalation from glucocorticoids and immunosuppressants to rituximab is recommended [15].

More recently, anakinra appears to be effective in the treatment of pericarditis during antisynthetase syndrome [5] [16].

#### 4. Conclusion

In conclusion, we report a case of antisynthetase syndrome with an atypical initial manifestation due to the presence of acute pericarditis. It is important for clinicians to consider the possibility of an antisynthetase syndrome in cases of pericarditis and to look for clinical signs suggestive of this disease.

#### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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