

Positive Rheumatoid Arthritis in Antisynthetase Syndrome

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Case Report

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Abstract: Positive Anticyclic Citrullinated Peptid Antibody in the antisynthetase syndrome should not lead to systematic diagnosis of association Antisynthetase syndrome-Rheumatoid arthritis. However their positivity is a marker of severity provider of destructive changes in joint damage in the antisynthetase syndrome without any diagnosis of Rheumatoid Arthritis established.

Keywords: Antisynthetase syndrome; Rheumatoid arthritis; positive ACPA.

INTRODUCTION

The Antisynthetase syndrome (ASS) is characterized by a series of clinical manifestations such as myositis, fever, mechanic's hands and diffuse interstitial lung disease (ILD), all associated with positivity of antisynthetase antibodies.

We report the case of an ASS with positive anticyclic citrullinated peptid antibody (ACPA) and we will try to explain the meaning of this association.

CASE REPORT

It's about a 52-year-old man patient without pathological history. He presents since 6 years, chronic distal symmetrical and bilateral polyarthritis interesting wrists, metacarpo-phalangeal, proximal interphalangeal, respecting the distal interphalangeal, without notion of swelling associated with a Raynaud, diffuse myalgia, deficit of scapular and pelvic belts and dyspnea; all evolving in a context of weight loss at 6kg over one year.

Examination of the musculoskeletal system notes, painful stiffness of the shoulders, pain at the pressure of muscular masses and painful mobilisation without limitation or synovitis of the joints peripheral devices. Mechanic's hand with trophic disorders of the pulp of fingers (Figure-1).

Pleuro-pulmonary examination: crepitating thoracic basal, the neurological examination: a deficit 4/5 at the level of the 2 arms.

A number of laboratory tests were obtained and the results are as follows: Sedimentation rate (CR) at 75 mm in the first hour, C reactive protein at 1.84 mg/L (RV: 0-6); LDH: 1000 UI/L (RV: 125-220); CK: 8450 UI/L (RV: 30-200); AST: 164 UI/L (RV: 5-34); ALT: 115 UI/L (0-55); Rheumatoid factor (RF): 336 U/ml (RV: 7-14), ACPA: 300 UI/ml (RV < 12

UI/ml); Antinuclear antibody (ANA): positive (1/160, cytoplasmic fluorescence); positive JO1 antibody; SCL70, PM-SCL100, DNA, SSA, SSB antibodies: negative;

X-ray of the hands notes demineralisation in bands and resorption of phalangeal tusocks without erosions (Figure-2), ultrasound of hands looking for synovitis or erosions is normal.

At the pulmonary level, in comparison with the previous pulmonary imaging, we note the emergence of a bilateral basal interstitial syndrome confirmed by CT scan thoracic (Figure-3) associated with a restrictive syndrome. Electromyogram detects myogenic involvement and muscle biopsy shows an inflammatory muscle infiltrate.



Fig-1: Mechanic's hand with trophic disorders of the pulp of fingers



Fig-2: Demineralisation in bands and resorption of phalangeal tufts without erosions



Fig-3: Interstitial lung disease (ILD)

The diagnosis retained is an AAS with positive rheumatoid serology but the possibility of overlap (AAS- RA) although rare is not to be dismissed.

The patient received steroids at a dose of 1 mg / kg / day combination with cyclophosphamide with good tolerance and improvement of biologic tests (CR-CK- AST/ALT) at one month.

DISCUSSION

Arthritis, interstitial lung disease (ILD), and Raynaud phenomenon are symptoms encountered in both ASS and RA [1-4]. Our patient meets the diagnostic criteria of ASS (myositis, mechanic's hands, diffuse interstitial lung disease (ILD), arthralgia and positive antisynthetase antibody) and generally to those of RA. However, some elements are against the diagnosis of RA:

- The importance of systemic signs, particularly Raynaud with trophic disorders, dyspnea and deficit of proximal belts.
- The absence of episode of synovitis throughout the duration of evolution.
- The absence of destructive lesions after 6 years of evolution under no background treatment.
- The discrepancy between positive rheumatoid serology (RF, ACPA) and a benign evolution of RA.

Meyer *et al.*, [5] described, that one third of positive ACPA –ASS patients were first diagnosed as having solely RA. However, during the course of the follow-up, all but 2 patients developed at least 1 additional sign of ASS, including typical dermatological signs and/or myositis.

Indeed, joint involvement affected 50% of patients with ASS, presenting joint pain or arthritis with or without bone erosions. All the patients had joint symptoms early in the disease [6].

Typically, the arthritis in idiopathic inflammatory myopathies is light, nondeforming, and nonerosive [7]. Although a subgroup of patients might develop an erosive and/or subluxing joint disease. The following joints are commonly affected: distal and proximal interphalangeal joints, metacarpophalangeal joints, wrists, elbows, and knees.

Meyer *et al.*, [5] described 40 patients with ASS and joint manifestations and they were classified into three categories: subluxing joint disease of thumb distal interphalangeal joints (n = 6); symmetrical polyarthritis with or without erosion (n =24) and isolated arthralgia (n= 10). Oddiset *al.*, [8] described non erosive joint disease of proximal and distal interphalangeal joints (floppy thumb).

RF and ACPA are not very specific and exclusively positive in RA. Several reports evaluating the meaning of anti-CCP antibodies in connective tissue diseases other than RA; in systemic sclerosis (SSc) and systemic lupus erythematosus (SLE), these antibodies are markers of erosive arthritis, as we suggest in antisynthetase syndrome. In contrast, in Sjögren's syndrome this association is lacking, although anti-CCP seem to be closely associated with the occurrence of synovitis.

The cases described by Nagashima *et al.*, [9] all patients with erosive arthritis also had clinical and biopsy-proven inflammatory myositis; we also confirmed the association between erosivity and anti-CCP in anti-Jo-1-positive patients.

Our patient was initially considered to have a positive serology RA (RF, ACPA), he was treated with

methotrexate at the dose of 25 mg stopped after 4 years of use for the appearance of an interstitial lung disease which was falsely assigned to him. Indeed, the diagnosis of this patient was reconsidered and we have finally selected an ASS with positive ACPA. The possibility of a SAA-PR association remains unlikely.

In the current series, the positive-ACPA-ASS patients experienced more frequently DMARDs refractory. Although there is no consensual therapeutic strategy for such patients in ASS. However, it has recently been reported that anti-TNF agents may not be effective in ASS patients and may even trigger myositis and/or ILD in ASS [10, 11].

CONCLUSION

ASS is an uncommon clinical entity and therefore little known by rheumatologists. The positivity of ACPA during this syndrome further complicates the diagnosis. Nevertheless, the significance of this association according to the literature is in favor of a marker of severity, the possible association ASS-RA is rare.

Consent

Written informed consent was obtained from the patient for publication of this case report.

Conflict of interests

The authors declare no conflict of interest.

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