

## Learning Objectives

1. Recognize multiple organ system involvement in Anti synthetase syndrome.
2. Recognize myositis as uncommon initial presentation of Anti synthetase syndrome.

## Case Description

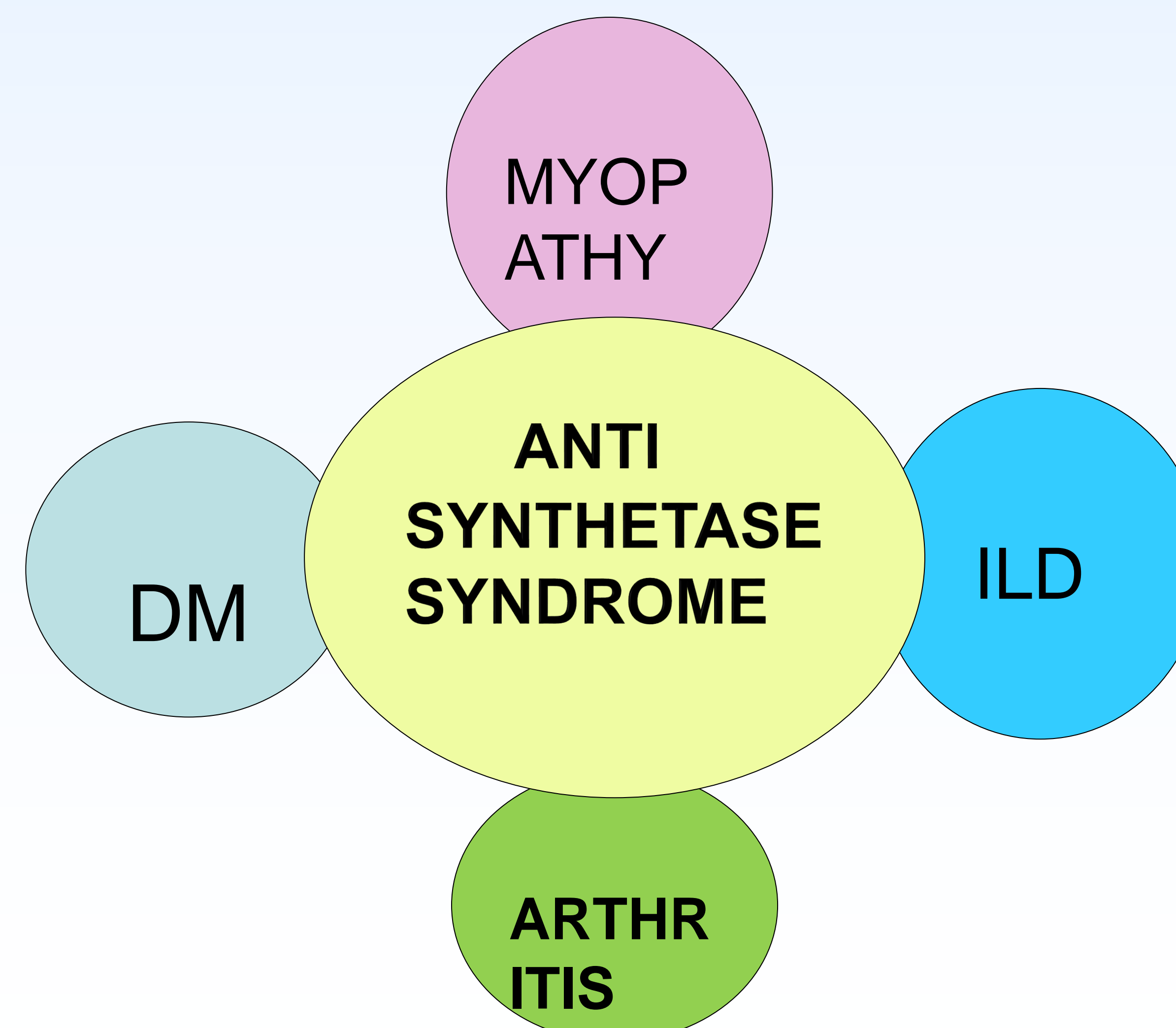
A 62-year-old female with OA and HTN presented with dull, achy pain in both wrists, hands, elbows, knees, and ankle joints, associated with swelling of both wrist joints, fatigue, hair thinning, and Raynaud's phenomenon.

She had bilateral wrist synovitis, a diffusely enlarged thyroid and 5/5 motor power.

Chest X-ray was unremarkable. Bilateral hand radiographs showed degenerative joint space narrowing. Thyroid ultrasound showed a heterogeneously enlarged thyroid. Labs were consistent with a diagnosis of **overlap** of SLE, MCTD and autoimmune thyroid disease.

Blood Test	Result	Blood Test	Result
ANA	1: 12560	Aldolase	21.1 (1-7.5)
Anti-smith antibody	6.5 (<1)	CK	1092 (22-198)
Anti-smith/RNP antibody	>8 (<1)	Thyroid stimulating antibody	191 (<140)
TSH	191 (<140)	Thyroid peroxidase antibody	127 (<9)

Myositis antibodies were obtained, given persistently elevated CK and aldolase levels. Interestingly **Jo-1 antibody was positive** which along with the clinical signs and symptoms confirmed the diagnosis of anti-synthetase syndrome. Prednisone and Hydroxychloroquine were started. She developed muscle weakness with motor power 4/5 in upper extremities and CK trended up, so Azathioprine was later added with good response.



## Review

Anti synthetase syndrome is a rare autoimmune disease characterized by constitutional symptoms, myositis, arthritis, and ILD.

Diagnosis requires the presence of an anti-synthetase antibody plus **two** of the following features: ILD, inflammatory myopathy, or polyarthritis.

Of these presenting features, ILD is the most prevalent.

## Conclusions

Anti synthetase syndrome rarely presents with myositis as the initial manifestation. Our case highlights uncommon presentation of a rare syndrome, as myositis is often absent/subclinical in Anti synthetase syndrome. Anti synthetase syndrome should be considered in atypical cases, as it mimics RA.