Objective(s): Anti-NXP2 are myositis-specific autoantibodies, described in idiopathic inflammatory myopathies. To our knowledge, these autoantibodies have not been associated with other rheumatic diseases. We wish to report three cases of isolated seronegative polyarthritis, attributed to anti-NXP2 autoantibodies, without myositis or dermatomyositis (DM) rash.

Method(s): Written informed consent from each patient was obtained as part of their participation in the Canadian Inflammatory Myopathy Study. Approval by our local ethics committee was not required for case reports under four patients.

A 51-year-old female presented with acute hands polyarthritis. Examination did not show muscle weakness or rash. She had normal inflammatory markers and creatine kinase (CK). Serological workup demonstrated positive ANA (1/640 speckled) and anti-NXP2 (3+) on a myositis panel (Euroimmun). Thighs MRI did not show inflammatory hypersignals. A nailfold capillaroscopy showed dystrophic capillaries without a definite DM pattern. Age appropriate cancer screening was negative. Her polyarthritis resolved with a short course of prednisone followed by methotrexate (MTX) and hydroxychloroquine (HCQ).

A 58-year-old female presented with acute polyarthritis regarding her hands, wrists and ankles. Muscle strength was normal and there was no rash. She had normal inflammatory markers and CKs. Serological workup demonstrated positive ANA (1/320 speckled) and anti-NXP2 (3+). MRI and EMG were normal. Nailfold capillaroscopy showed dystrophic capillaries without a specific DM pattern. Extensive cancer screening was negative. She was successfully treated with prednisone, followed by MTX and HCQ.

A 22-year-old female developed polyarthritis and edema of both hands. She also reported mild upper limb myalgia over the last 3 months. Examination did not reveal muscle weakness or rash. She had normal inflammatory markers and mildly elevated CKs (390 U/L, normal range 24-184). Serological workup demonstrated positive ANA (1/640 diffuse) and anti-NXP2 (2+). Muscle MRI, EMG, nailfold capillaroscopy and quadriceps muscle biopsy were normal. Full-body PET scan was negative. Her polyarthritis was successfully treated with a combination of prednisone, MTX, HCQ and tofacitinib.

Result(s): Our three cases presented with a unique phenotype of acute polyarthritis, normal inflammatory markers and no cutaneous or muscular features of DM.

Conclusion(s): Thus, anti-NXP2 autoantibodies may present with seronegative polyarthritis as the sole clinical manifestation. This antibody could be sought in the presence of isolated positive ANA and normal inflammatory markers. Although none of our patients had cancer, clinicians should remain careful as this antibody has been associated with a higher risk of cancer.