Should all patients with anti-centromere antibodies be referred for a rheumatology assessment?

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Background: Anti-centromere antibodies (ACA) are commonly associated with systemic sclerosis (SSc). The presence of ACAs in patients with SSc is known to increase the likelihood of developing Pulmonary Hypertension, which has a high mortality rate. ACAs are also seen in patients with other connective tissue diseases (CTD) and can sometimes be identified after testing for antinuclear antibodies in patients who have not reported any rheumatological symptoms. It is possible that connective tissue diseases are being under diagnosed due to a lack of awareness by physicians of the clinical significance of ACAs.

Objectives: To investigate whether patients with ACAs are being appropriately referred to the Rheumatology service.

Methods: All patients who were positive for ACAs in Southend university hospital between April 2016 and October 2018 were included in this single centre retrospective observational study. We identified patients' demographics, diagnosis, ANA titre, additional diagnosis and immunosuppressive therapy. We also captured their monitoring with pulmonary function testing and echocardiography.

Results: A total of 75 patients were identified with ACAs. The average age was 65 years, 61 females, 14 male. Fifty-six patients were referred to rheumatology team and were found to have the following diagnosis, LcSSc (10), undifferentiated connective tissue disease (UCTD) (6), Rheumatoid arthritis (RA) (5), ANCA associated vasculitis (AAV) (3), Raynaud’s phenomenon (3), Lupus (2), Antiphospholipid syndrome (APS) (1), Primary biliary cirrhosis with Sjögren’s syndrome (SS) (1), Juvenile idiopathic arthritis (1), Antisynthetase syndrome (AS) (1), Autoimmune hepatitis (1) and osteoarthritis (1). Of those 33 patients had a routine screening with an Echocardiogram and 26 had pulmonary function tests. One patient with LcSSc developed pulmonary hypertension. The remaining 19 patients were not referred and did not have the adequate screening for pulmonary hypertension. ANA titre was 1:80 in one patient, 1:320 for 4 patients, unknown for three and 1:640 for the remaining 67. 11 patients were treated with Hydroxychloroquine (4 SS, 4 UCTD, 1 lupus, 1 APS and 1 Lc SSc), 5 on Methotrexate (4 RA and 1 AS), 2 on MMF and steroid (RAPS).

Conclusion: Nearly all patients with ACAs that were seen in the Rheumatology clinic had an autoimmune rheumatic disease. However, we found that 25% of people with ACAs were not referred to the Rheumatology service. For the reasons for this are unclear. It is possible that patients did not report symptoms that would have prompted a referral. Some of the CTDs in which ACAs are typically found (LcSSc and SS) are associated with symptoms that can be mild and might not be reported by the patients or general physicians do not associate them with rheumatological disorders (e.g. sicca symptoms, gastro-esophageal reflux and Raynaud’s phenomenon). Early diagnosis might enable earlier treatment and prevent complications from these diseases. General physicians should therefore be more aware of these antibodies and the disorders that they can be associated with.

Disclosure of Interests: None declared