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Unilateral Pleural Effusion as a Presentation of Clinically Amyopathic Dermatomyositis

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Dear Editor,

Clinically amyopathic dermatomyositis (CADM) is a subset of dermatomyositis (DM), accounting for 20% of cases[1]. It has been hypothesised that CADM and DM may occur in continuum[2]. Pleural effusion is a very rare manifestation of DM and even rarer in CADM, with only a single previous case report[1].

We investigated a middle aged year old female in our institution for a 7 month history of dyspnoea and a persistent pleural effusion. The pleural effusion had been diagnosed in April 2019 in a peripheral hospital following a short history of right sided pleuritic pain and subsequently a pulmonary embolism was diagnosed. On further discussion, a 9 month history of a bilateral erythematous rash on the dorsal surfaces of the hands, a 7 month history of profound lethargy and 5kg weight loss, and a 4 month history of hair thinning and intermittent migratory polyarthralgia involving her knees, wrists, hands, and shoulder joints were revealed.

Bloods revealed normochromic normocytic anaemia, lymphopaenia and ESR was mildly elevated. Serial serum CK levels were within normal limits. US guided thoracentesis revealed an exudative effusion, abundantly lymphocytic with a white cell count of 3000 cells/mm3 of which 98% were monomorphic cells, no malignant cells were seen. PET CT did not identify any sinister pathology and imaging revealed no evidence of myositis.

ANA was positive at a titre of 1:80 with homogenous staining pattern. ENA panel demonstrated negative anti-ds DNA, anti-Jo1, anti-RO/La, anti-RNP, anti-Sm and anti-SCl 70. Rheumatoid factor, anti-CCP and anti-phospholipid antibodies were negative. ANCA was obscured by nuclear staining but PR3 and MPO were negative. An extended myositis panel demonstrated strongly positive anti-MDA5 antibody and weakly positive anti-NXP2 antibody.

A diagnosis of CADM was made and the patient was commenced on prednisolone 1mg/kg and methotrexate 15mg with good effect. Pulmonary function tests showed a restrictive pattern with an FEV1:FVC of 90%, reduced total lung capacity and reduced diffusing capacity of the lung (DLCO 47% predicted) consistent with a likely diagnosis of interstitial lung disease.

Pleural effusion is a very rare manifestation of DM and even rarer in CADM, with only a single previous case report[1]. The nature and persistence of the pleural effusion leads us to conclude that it was secondary to DM. Venous thromboembolic disease is very common in DM with a hazard ratio of 26.6 in the first year after diagnosis[3] and we believe the pulmonary emboli were also secondary to DM. Pleural effusions have not previously been reported as an isolated phenomenon in DM but as local immune pleuritis associated with ILD[1].

Myositis specific antibodies (MSA) are present in approximately 50% of cases of DM[4]. Anti-MDA5 is associated with amyopathic dermatomyositis and rapidly progressive ILD, while anti-NXP2 is associated with calcinosis, skin disease, and malignancy, often genitourinary, lung and breast. No previous data has shown association between NXP2 and melanoma.

We wish to highlight the importance of CADM as a rare but important consideration of the differential diagnosis of a pleural effusion. An extended myositis panel has both diagnostic and prognostic value. Certain MSAs associate with distinctive clinical manifestations. Patients with anti-MDA5 should be monitored for ILD while those with anti-NXP2 should be evaluated for malignancy.

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