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Title: Nine cases of interstitial lung disease associated with anti-CADM140 antibody positive dermatomyositis

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Body: [Background] Anti-CADM140 antibody (CADM) was found in some amyopathic dermatomyositis (DM) patients (pts) in 2005. It was reported that about 50% of CADM positive pts died from acute exacerbation (AE) of DM-related interstitial lung disease (DM-ILD) despite treatment. As compared with CADM negative pts (about 6%), the rate of AE of ILD is clearly high in CADM positive pts. [Objective] To elucidate the clinical characteristics of ILD associated with CADM positive DM. [Methods] Blood examination, arterial blood gas analysis, pulmonary function testing, bronchoalveolar lavage (BAL) analysis and pattern of chest CT were examined in 9 pts who were diagnosed with CADM positive DM in our hospital. [Results] Since the average value of WBC and CRP at the time of the first medical examination was 6629 /μl and 0.56 mg/dl, we thought that inflammation was slight. The value of CK was in the normal range in 7 pts, slightly high (217 IU/l) in 1, and significantly high (26300 IU/l) in 1. The value of KL-6 was in the nearly cut-off range in 8 pts, and significantly high in 1 (3764). Alveolar-arterial oxygen difference was increased in 3 pts. In 4 pts in which BAL was performed, the lymphocyte differentiation was all high. Concerning chest CT image, patchy shadow or patchy shadow and trabecular shadow in outside layer of the lung field were seen in 7 pts, GGO around broncho-vascular bundle in 1, and no abnormal shadow in 1. All of the shade were slight. Although one pt died of AE, 8 pts survive as of February, 2012. [Conclusion] In DM-ILD, even if the CT image and data are slight, whenever the CT image is not typical NSIP pattern, we have to take the possibility of CADM into consideration.