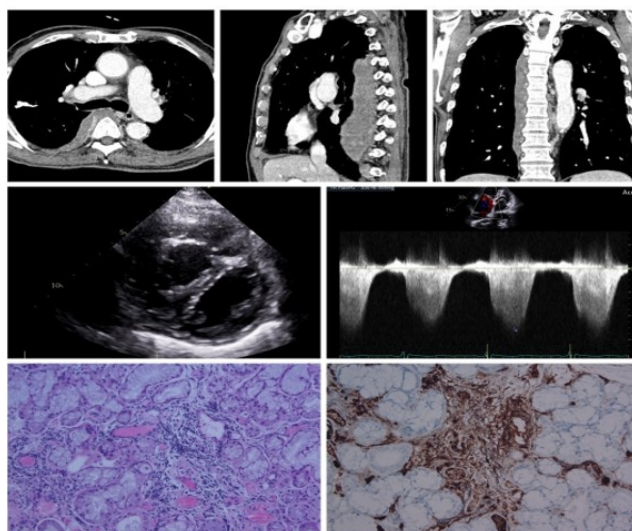


Pulmonary arterial hypertension associated with Thoracic paravertebral IgG4-related disease(IgG4-RD)

원광의대병원 내과¹최지호¹, 정종혁¹

An 81-year old man who previously had COPD was reported in ER for abnormal cardiac enzyme results. An elevation of Troponin T, and BNP, which is a marker for heart failure was noted(1292pg/ml, reference range 0~150) and Echocardiography was performed for evaluation of heart failure. Severe pulmonary hypertension was found(PASP=119.4mmHg, severe criteria : PASP \geq 60). To figure out the etiology, Right heart catheterization was done and chest CT, lung scan for presence of pulmonary thromboembolism. Also, thyroid function, rheumatologic antibody and complement study were performed. Serum IgG elevation was shown(2617mg/dL, reference range 870~1700) and among the subgroups, the elevation of serum IgG4 was prominent(22500mg/L, reference range 30~2010). According to chest CT, there were no signs of thromboembolism. However, peribronchovascular interstitial thickening and nodular/linear pleural thickening with both posterior pleural effusion was found. Also, long segmental paravertebral soft tissue was seen through entire thoracic spine level. To rule out malignancy, biopsy was done via Endobronchial ultrasound-guided Transbronchial needle aspiration(EBUS-TBNA). Biopsy specimens showed a lymphoplasmacytic infiltration and storiform fibrosis with occasional eosinophils. Immunostaining for IgG4 showed that most plasma cells are positive for IgG4, suggesting IgG4-RD. After diagnosis of IgG4-related disease, steroid therapy (Methylprednisolone 30mg) was done. At the blood test f/u, decrease of IgG4 level and patient's subjective extent of dyspnea improved, so the patient was discharged with steroid tapered to oral prednisolone. IgG4-RD is commonly found on pancreas, bile duct, salivary gland, lacrimal gland, lung, kidney and aorta. Few cases were reported about paravertebral involvement of IgG4-RD, and in 2019 Inoue et al. characterized CT findings of Thoracic paravertebral lesions in IgG4-RD, but little studies were done about clinical aspects of paravertebral involvement. In this case, IgG4-RD is highly suggestive for the origin of pulmonary hypertension. Therefore, more studies and investigations should be done about clinical between IgG4-RD and pulmonary hypertension.



Top 3 figures – Figure 1A, 1B, 1C

Middle 2 figures – Figure 2A, 2B

Bottom 2 figures : Figure 3A, 3B

Figure 1A, 1B, 1C : Chest CT with contrast was done, peribronchovascular interstitial thickening and interlobular septal thickening was shown on axial view. On sagittal and coronal view, shows homogeneously enhanced band-like mass, especially on the lower thoracic vertebrae – suggests paravertebral lesion of IgG4-related disease.

Figure 2A, 2B shows Echocardiography : figure 2A shows D-shaped Left ventricle, which implies severe pulmonary hypertension. In figure 2B, severe pulmonary hypertension can be suspected based on high TR value.

Figure 3A, 3B : Biopsy was done by EBUS-TBNA, and lymphoplasmacytic infiltration was detected : also IgG4 shown positive from immunohistochemistry.