厚生労働科学研究費補助金（難治性疾患政策研究事業）
分担研究報告書

肺高血圧症を呈した Airway-centered Fibroelastosis （症例報告）

研究分担者　井上義一
国立病院機構近畿中央呼吸器センター 臨床研究センター 臨床研究センター長

研究要旨

特発性 Idiopathic Pleuroparenchymal Fibroelastosis(PPFE)は特発性間質性肺炎の 1 つに分類されるまれな病態である。上肺優位に Fibroelastosis が認められ緩徐に進行し予後不良である。近年、気道中心性に Fibroelastosis が認められる事が報告され注目されている。当院で経験した肺高血圧を呈した Airway-centered Fibroelastosis の患者を経験したので報告する。症例は、26 歳の男性、胸部 CT で、気管支血管束に沿って胸膜下領域に高密度の陰性が指摘された。右 S5 からの外科的肺生検で、肺胞破壊を伴う胸膜下および中枢気道領域に線維弾性症を示し MDD の結果、気道中心性の iPPFE と診断した。 ピルフェニドン、ニンテダニブで治療を行ったが、肺高血圧症を発症し、6 年後に死亡した。

A. 研究目的

病気道中心性の Fibroelastosis 患者に認められた肺高血圧症について検討する。

B. 研究方法

肺高血圧症を呈した Airway-centered Fibroelastosis の症例について、外科的肺生検所見、画像所見、心臓カテーテル検査所見について検討し肺高血圧症との関連について文献を含めて考察する。

C. 研究結果

肺の線維化の進行と共に肺高血圧症を発症し、抗線維加薬を投与するも効果無かった。肺高血圧症の薬物療法は行わなかった。病理にて肺動脈の変化は明らかではなかった。肺病変に伴う肺高血圧症と考えられた。

D. 考察

Airway-centered Fibroelastosis はこれまで１０例ほどの報告であるが、Idiopathic PPFE との関連は不明である。３群の肺高血圧症と考えられた。

E. 結論

Airway-centered Fibroelastosis に伴う肺高血圧症は肺病変の進行にともなう 3 群と思われたが今後症例数の蓄積が必要である。

F. 研究発表

1. 症例報告

Minomo S, Arai T, Tachibana K, Matsui H, Kasai T, Akira M, Inoue Y. Airway-centered Fibroelastosis Accompanied by Subpleural Lesions of Unknown Cause in a Young Man Who Later Developed Pulmonary Hypertension. Intern Med 2020;59:695–700.

Internal Medicine
The Japanese Society of Internal Medicine
doi: 10.2169/internalmedicine.3256-19
Intern Med 59: 695-700, 2020
http://internmed.jp

【 CASE REPORT 】

Airway-centered Fibroelastosis Accompanied by Subpleural Lesions of Unknown Cause in a Young Man Who Later Developed Pulmonary Hypertension

Shojiro Minomo[1], Toru Arai[1,2], Kazunobu Tachibana[1], Hideo Matsui[1], Takahiko Kasai[3], Masanori Akira[4] and Yoshikazu Inoue[1,2]

Abstract:
A 26-year-old man with a history of bronchial asthma was found to have high-density shadows along the bronchovascular bundle and in the subpleural area on computed tomography of the chest. Surgical lung biopsy specimens from the right S^5 showed fibroelastosis in the subpleural and central airway area with alveolar destruction. He was diagnosed with airway-centered fibroelastosis of unknown cause after multidisciplinary discussions. The patient developed pulmonary hypertension and died 6 years later. The patient was younger in comparison to patients in earlier reports and had more obvious subpleural fibroelastic lesions in the upper lobes than in previously described cases.

Key words: airway-centered fibroelastosis, bronchial asthma, bronchial abnormality, pulmonary hypertension, subpleural lesions, upper lobes

(Intern Med 59: 695-700, 2020)
(DOI: 10.2169/internalmedicine.3256-19)

Introduction

Airway-centered fibroelastosis was first proposed as a pathological entity with a specific clinical and imaging presentation in 2016 (1). The characteristics of the 5 reported cases are extensive airway-centered fibroelastosis of the upper lobes on histopathology, bronchial abnormality, and predominantly subpleural upper lobe consolidation on high-resolution computed tomography (CT) of the chest (1). The reported patients have all been middle-aged women who were previously diagnosed with bronchial asthma and obstructive or restrictive respiratory dysfunction (1). We herein report a rare case of airway-centered fibroelastosis in a young man with obvious parenchymal fibroelastosis in subpleural lesions of both upper lobes who developed pulmonary hypertension.

Case Report

A 26-year-old man presented to a local doctor with a complaint of cough in 2011. A chest radiograph showed an infiltrative shadow and loss of volume predominantly in both upper lung fields. CT of the chest showed high-density shadows mainly in the upper lung fields (Fig. 1a-c). These findings prompted a referral to our institution. The patient's past medical history included bronchial asthma since his teenage years, which was treated with an inhaled corticosteroid and a long-acting beta-agonist (salmeterol/fluticasone). He was a non-smoking clerical worker in a welding factory and had not inhaled mine dust. He used a down quilt and had no pets. His family history included a father with liver disease, a mother with hypertension and hyperlipidemia, and a younger brother with allergic rhinitis. His body mass index on the first admission to our hospital was 24.7. Wheeze was detected on chest auscultation and clubbed fingers were

[1]Department of Internal Medicine, National Hospital Organization Kinki-Chuo Chest Medical Center, Japan, [2]Clinical Research Center, National Hospital Organization Kinki-Chuo Chest Medical Center, Japan, [3]Department of Pathology, National Hospital Organization Kinki-Chuo Chest Medical Center, Japan and [4]Department of Radiology, National Hospital Organization Kinki-Chuo Chest Medical Center, Japan
Received: April 25, 2019; Accepted: July 31, 2019; Advance Publication by J-STAGE: October 7, 2019
Correspondence to Dr. Yoshikazu Inoue, inoue.yoshikazu.nt@mail.hosp.go.jp