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A Case of Adult Congenital Cystic Adenomatoid Malformation of the Lung with Atypical Adenomatous Hyperplasia

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1. Introduction

Congenital cystic adenomatoid malformation of the lung is a rare disease that shows multiple cystic lesions in pulmonary tissues in the development process. It was first described by Chin et al.¹ in 1949 and its incidence is known to be 1:25,000 to 1:35,000². With the development of prenatal diagnosis, this disease can be diagnosed in 60% and detected within 2 years because of such symptoms as respiratory distress by compression of surrounding lung tissues immediately after birth and repeated respiratory infections in infancy. Among adults, it is detected accidentally on X-ray or by such symptoms as pneumonia, pneumothorax, and hemoptysis. In Korea, the first case in a 28 year-old woman was reported by Geun-Heung Ki et al.³ in 1989. Since then, about 25 adult cases were reported until 2006.

Atypical adenomatous hyperplasia is pathologically defined as the proliferation of atypical cuboidal or columnar cepithelial cells which are typically 5 mm or smaller in size along the alveolus or respiratory bronchiole^{4,5}. In 1999, the WHO classified this disease as a precancerous lesion together with squamous dysplasia and diffuse idiopathic pulmonary neuroendocrine cell hyperplasia⁵. It is known to be found in about 12% of lung cancers and highly correlated with glandular cancer⁶.

2. Case

Patient: O-Back Kim, Female, 37

Current case history: The patient had been treated in a private clinic due to purulent sputum and cough and hemoptysis which had started two weeks before transfer to our hospital.

Past case history: The patient had been treated for hyperthyroidism from 1999 to 2003.

Examination findings: Vital signs were stable. Chest examination detected reduced bilateral breathing sounds only.

Laboratory findings: The peripheral blood examination found 13.2 g/dL of hemoglobin, 39.5 % of hematocrit, 5210 /mm³ of white blood cells (neutrophils 43%, lymphocytes 44.5%, monocytes 9.2%), and 255,000 /mm³ of platelets. The results of serum biochemical examination, hepatitis marker test, urinalysis, mycoplasma antigen test, and pulmonary function test were normal. The thyroid function test found minor hypothyroidism with 8.72 nIU/ml of thyroid stimulating hormone (TSH), 0.894 ng/dl and 1.19ng/ml of free T4 and T3 respectively. The results of the sputum culture and bronchial washing cytodiagnosis were also negative. The results of arterial blood gas analysis which was conducted in atmosphere were 7.407 of pH, 41.2 mmHg of carbon dioxide tension, 82.6 mmHg of oxygen tension, 25.4 mEq/L of bicarbonates, and 96.2% of oxygen saturation. The tumor marker test result was also normal with 1.49 ng/mL of CEA and 1.57 ng/mL of cyfra 21-1.

Chest radiology: The chest X-ray showed a 2.5cm cavitating lesion in the left upper zone. (Figure 1).

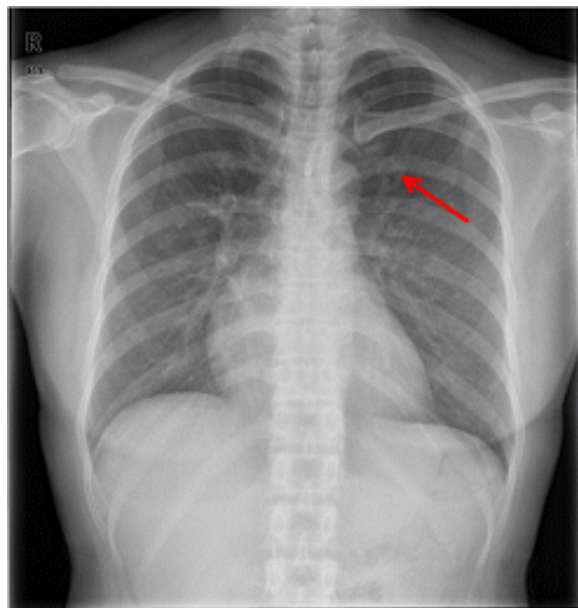


Fig. 1. Chest PA shows cavitary lesion in the left upper zone

On the second day after hospitalization, the chest CT showed a diffuse ground glass appearance and cystic lesions that spread in a branching pattern were found in the left lung (Figure 2).

Clinical progress: The patient received symptomatic therapy as there were no features of infection. The bronchial endoscopy did not reveal any lesions. The bronchoalveolar lavage fluid test found 82% of macrophages, 8% of neutrophils, 0.5% of eosinophils, and 9.5% of lymphocytes. Endobronchial lung biopsy was performed at the left lower lobe and only chronic inflammatory changes were noted. On the 9th day after hospitalization, a wedge resection of the top section of the left upper lobe was performed. On the 20th day after hospitalization, the patient was discharged with no complications.

Biopsy: The microscopic findings of the resected lung showed cystic lesions of various sizes. The cells covering them were diverse, including ciliated pseudostratified columnar cells, monolayer columnar cell, and cuboid cells, which corresponded to type 2 of the congenital cystic adenomatoid malformation(Figure 3).

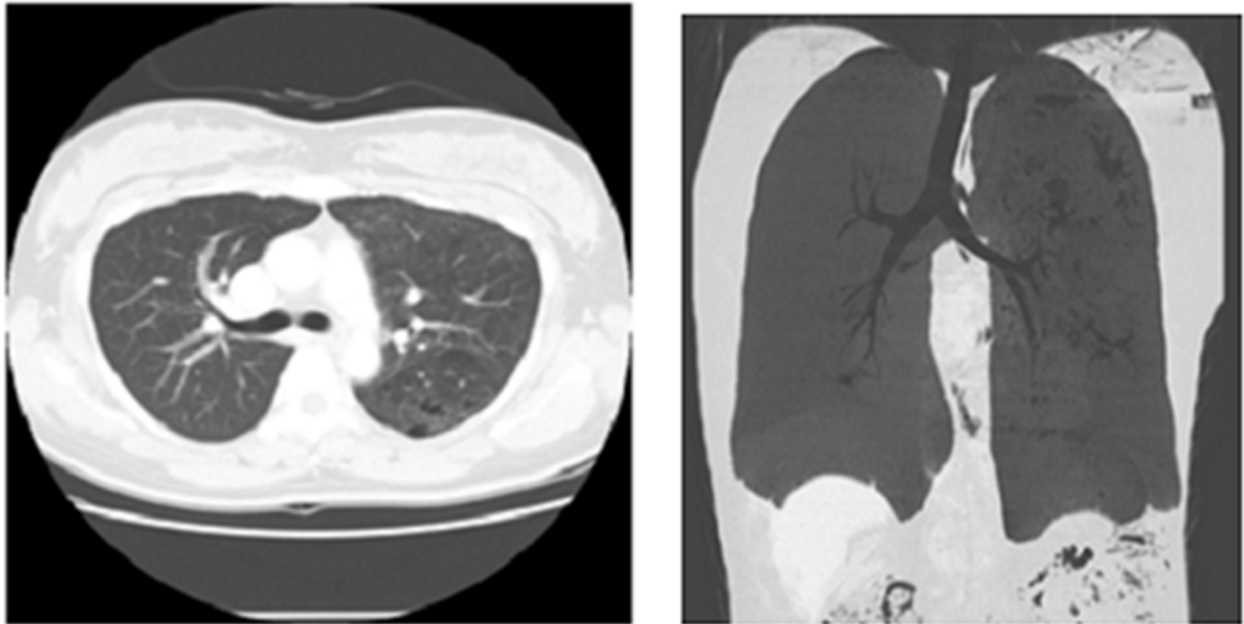


Fig. 2. Chest CT shows diffuse ground glass opacity and multiple branching cystic lesion in left lung

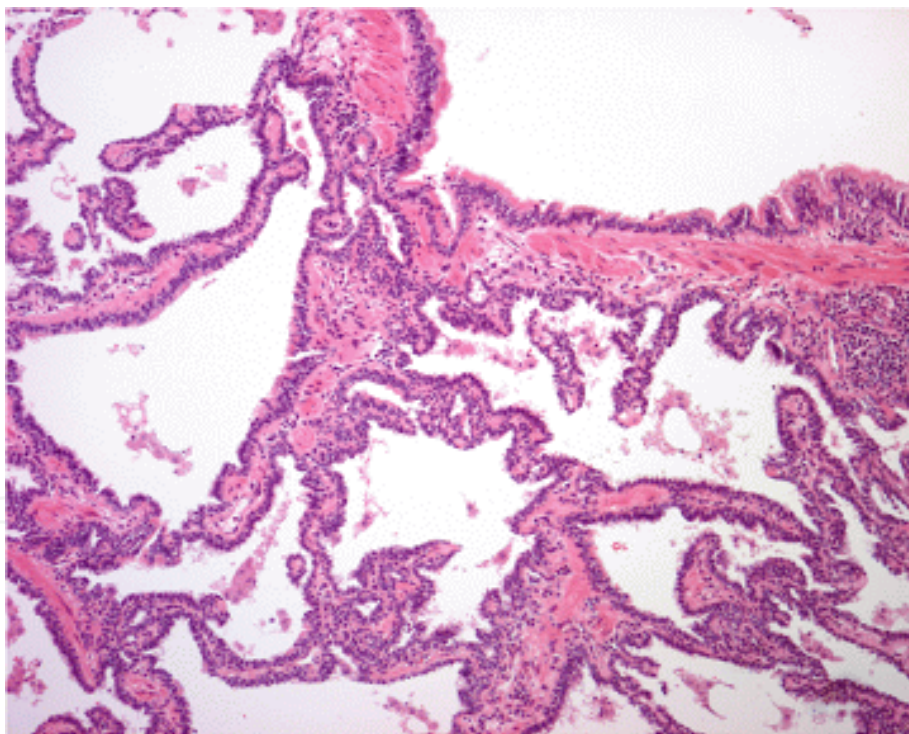


Fig. 3. The lining cells are pseudostratified ciliated columnar cells to columnar and cuboidal cells like bronchiolar epithelial cells (H&E, x100).

As there were minor atypical nuclei locally and linear structures filled with mucus in the cytoplasm, it looked similar to the mucoid bronchioloalveolar carcinoma, but because their sizes were all smaller than 5mm, they were judged to be atypical adenomatous proliferation (Figure 4).

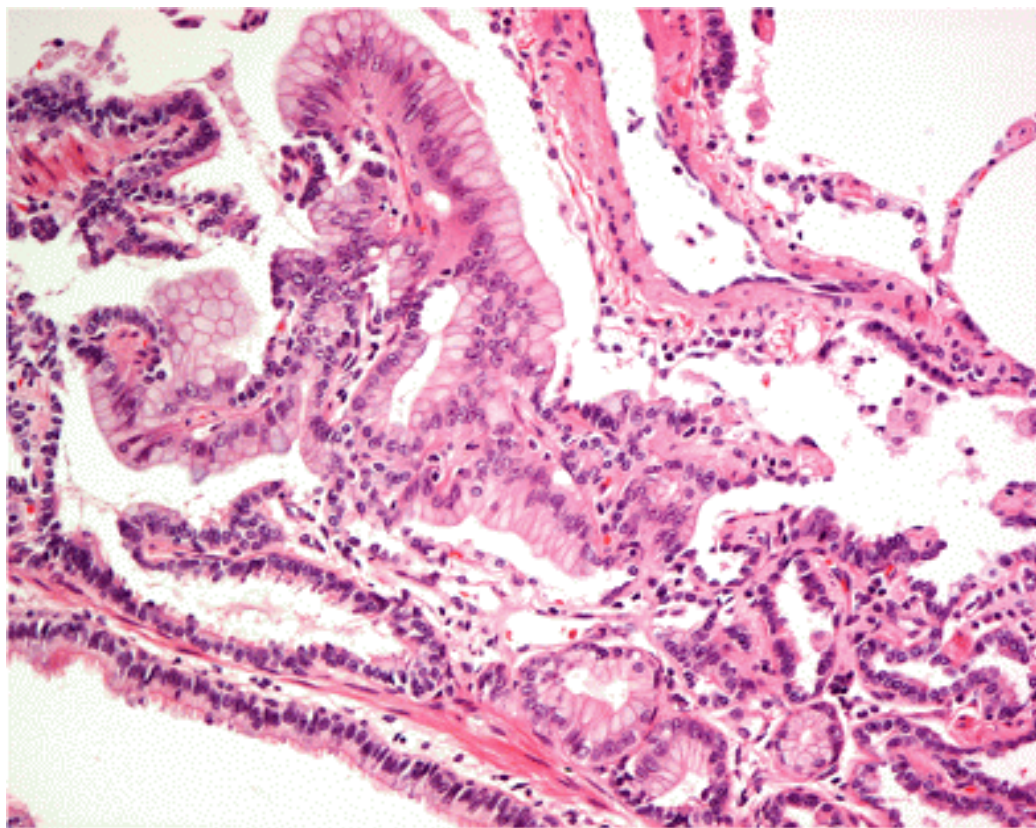


Fig. 4. Focal mucinous epithelial lesion with mild atypism is revealed in the intervening parenchyma (H&E, x200).

Treatment and progress: After discharge, the patient was transferred to another hospital at her request; As it was checked, she was being followed up with chest CT every 3 months with no specific treatment.

3. Discussion

The congenital cystic adenomatoid malformation of the lung is very rare and its incidence is known to be 1:25,000 - 1:35,000². The cause is not known, but two hypotheses have been suggested: cessation in the development of the lung tissues and no development of aveoli during the development process of the respiratory system^{7,8}. The time when the malformation occurs is estimated to be between 5 and 6 weeks before the lobe divides and prechondrial tissues are formed in the bronchus. It has been reported that it causes stillborn and premature infants. Immediately after birth, respiratory distress is frequent in most cases, and as the baby grows, repeated infections and pneumothorax are more frequent than respiratory distress. Accompanying malformations include kidney growth failure, diaphragmatic hernia, jejunal atresia, and colon growth failure¹. A histological characteristic is the arrangement of polyp-shaped cysts of various sizes in the bronchial epithelium or simple columnar epithelium with no cartilaginous tissues or inflammatory reaction. Stoker et al. classified them based on size and pathological findings into type I (only a few large thick walled cyst), type II (numerous, evenly spaced cyst, less than 1cm), and type III (less numerous, firm and bulky masses) in 1977. In 1994, they re-classified them into 5 types based on bronchial invasion: type 0 (bronchial), type I (bronchial/bronchiolar), type II

(bronchiolar), type III (bronchiolar/alveolar), and type IV (peripheral). In 1994, they reported that type I was the most frequent at 50 - 70% and type III showed the worst prognosis. Radiographic diagnoses include chest X-ray test, CT, and prenatal ultrasonography. Among them, chest CT can observe lesions that contain cysts with multiple large and small thin walls. It must be differentiated from pneumonia accompanied by pneumatosis, pulmonary sequestration, congenital lobar emphysema, and bronchiectasis. Definite diagnosis is only possible by pathological tests. Some claim that it is associated with malignant tumors of the lung. There was a report of pulmonary rhabdomyosarcoma in 22-month old boy, and several papers reported the accompaniment of mucoid bronchioloalveolar carcinoma in adults and children^{3,9}. Hence, Ioachimescu et al.⁹ recommended surgical removal even if there were no symptoms, because it may become malignant.

Pathological findings in this patient were accompanied by atypical adenomatous hyperplasia. Atypical adenomatous hyperplasia is pathologically defined as the proliferation of atypical cuboidal epithelial cells or columnar epithelial cells along the alveolus or respiratory bronchiole⁴. Their sizes are mostly 5 mm or smaller, although a size of 19mm has been reported. As it is difficult to differentiate from bronchioloalveolar carcinoma, one researcher suggested 5 mm as the reference size for differentiation. It occurs in up to 5% of normal people and is usually asymptomatic. It is known to develop in 2.9% of total population and increases to 10 - 23.2% in lung cancer^{11,12,13,14,15}. The recent diagnostic rate is increasing due to low dose chest CT as a lung cancer screening test. Although there are no specific CT findings, the most frequent finding are nodules with a good boundary accompanied by a ground glass appearance¹⁰. In 1999, the WHO classified this disease as a precancerous lesion together with squamous dysplasia and diffuse idiopathic pulmonary neuroendocrine cell hyperplasia⁵. Chapman et al.⁶ analyzed the pathological findings of 554 patients with primary lung cancer and found that atypical adenomatous hyperplasia was accompanied in 67 cases (12.1%), and that the percentage of glandular cancer was the highest (glandular cancer 23.2%, giant cell undifferentiated cancer 12.5%, epithelial carcinoma 2.2%). There are many different opinions among pathologists and no established views on the differentiation level of atypical adenomatous hyperplasia, classification based on this, and its relationship with adenocarcinoma. However, there are some reports related to morphological changes of the nuclei, expression of Ki-67 and p53, and K-ras mutation, which are expected to be helpful for better investigation of the characteristics of atypical adenomatous hyperplasia as a precancerous lesion^{5,10}. As there are no principles in therapy yet, careful follow-up is needed.

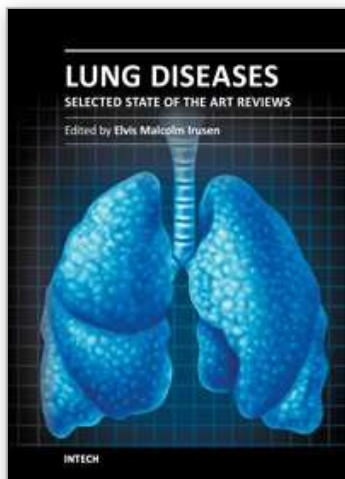
Although there is controversy about the treatment of this patient, the authors believe that pneumonectomy of the left lung will be necessary because the patient has both congenital cystic adenomatoid malformation which can be accompanied by bronchioloalveolar carcinoma and atypical adenomatous hyperplasia which is a precancerous lesion.

After a literature review, it is believed that this is the first case report of congenital cystic adenomatoid malformation accompanied by atypical adenomatous hyperplasia.

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