We present a case of congenital cystic adenomatoid malformation (CCAM) in a 25-year-old male who was presented with chronic cough. Chest radiography revealed an abnormal mass-like shadow in the right lower pulmonary zone. A contrast enhanced computed tomography showed an 11 cm solid, cystic mixed mass on the right lower lobe. A right lower lobectomy was performed by video-assisted thoracoscopic surgery without complications. The gross specimen showed a massive cavitation with multiloculated cysts of varying size, consistent with CCAM, along with noticeable granulomatous inflammation. Non-tuberculosis mycobacteria were isolated from a bronchial wash specimen, and the resected tissue homogenates were positive for *Mycobacterium avium-intracellulare* complex by polymerase chain reaction.

Key Words: Cystic Adenomatoid Malformation of Lung, Congenital; Nontuberculous Mycobacteria; *Mycobacterium avium* Complex
showed an abnormal mass-like shadow in the right lower lung (Figure 1A). Contrast-enhanced computed tomography (CT) revealed an 11 cm solid, cystic mixed mass in the right lower lobe without a feeding vessel from the descending aorta (Figure 1B). Bronchoscopy disclosed purulent secretion from the posterior basal segmental bronchus of the right lower lobe. A pulmonary function test was normal.

Subsequently, the patient underwent a right lower lobectomy by video-assisted thoracoscopic surgery. A macroscopic examination showed a massive cavitation with multiloculated cysts of varying size within the lesion and pleural fibrosis (Figure 2). Microscopically, the specimen consisted of multiple cysts (Figure 3A) which were lined by cuboidal-to-ciliated pseudostratified columnar epithelium, and some cysts contained extensive secondary inflammation (Figure 3B). These histological features were consistent with typical type I CCAM. In addition, a microscopic examination revealed a noticeable caseous granulomatous inflammation in the cyst walls (Figure 3A). The necrotic center was surrounded by epithelioid histiocytes and then by a rim of lymphocytes (Figure 3C). Forty colony forming units of non-tuberculous mycobacterium (NTM) were isolated from a bronchial wash specimen. Furthermore, resected lung tissue homogenates were positive for MAC by real-time polymerase chain reaction (Figure 3D). The post-operative course was uneventful without anti-mycobacterial treatment. Two years after surgery, chest radiography showed a well-expanded right lung without lesions.

Discussion

CCAM is a rare developmental, non-hereditary, hamartomatous abnormality of the lower respiratory tract first described in 1949. It occurs sporadically and is not related to genetic predisposition, gender predilection, or maternal factors such as race, age, or environmental exposures. Stocker et al. suggested a new name, congenital pulmonary airway malformation, and classified CCAM into five types based on the site of the defect in the tracheobronchial tree. Type I CCAM is the most common subtype and consists of a single or multiple large epithelial-lined cysts. The walls of the cysts con-
Figure 3. (A) The lesion consists of multiple cysts with caseous granulomas and inflamed granulation tissue (H&E stain, ×40). (B) The cysts are lined by cuboidal-to-ciliated pseudostratified columnar epithelium (H&E stain, ×100). (C) A marked caseous granuloma is shown, and the necrotic center is surrounded by epithelioid histiocytes (H&E stain, ×100). (D) Electrophoresis of the polymerase chain reaction product. M: size marker; PC 1: positive control (Kit-positive sample); PC 2: positive control (Mycobacterium avium-intracellulare complex); NC: negative control; Pt: patient sample.

The pathogenesis of CCAM is uncertain but may include an imbalance between cell proliferation and apoptosis during organogenesis. Affected patients may present with symptoms, including cough, dyspnea, hemoptysis, and respiratory distress, or remain asymptomatic. The typical manifestations include progressive respiratory distress in the newborn and recurrent pulmonary infections in older children.

The majority of cases are identified by prenatal ultrasound examinations. Additional prenatal magnetic resonance imaging may help to distinguish CCAM from other congenital lesions. A postnatal diagnosis can usually be made using plain radiography. Chest CT scans may be helpful for confirming the diagnosis in cases that are confusing. The differential diagnosis of CCAM includes bronchopulmonary sequestration, a congenital diaphragmatic hernia, a bronchogenic cyst, congenital lobar emphysema, and pneumatoceles. The most frequent complication of CCAM is recurrent or persistent pulmonary infection. Other complications include hemopto-nemorrhage, hemoptysis, and chronic cough. CCAM has been associated with the development of malignancies, such as bronchioloalveolar carcinoma, adenocarcinoma in aged, and pleuropulmonary blastoma. Surgical resection of the affected part of the lung is the treatment of choice, and this prevents recurrent infections and malignant transformation.

Pulmonary NTM infection usually occurs in patients with destroyed lungs, including those with chronic obstructive lung disease, pulmonary tuberculosis, bronchiectasis, or pneumonoconiosis. While NTM infection is known to occur commonly in old age, NTM infection occurs in destroyed lung caused by CCAM despite young age in our patient.

A few case reports have detailed NTM infection in congenital lung lesions such as bronchogenic cysts and
pulmonary sequestration, and surgical resections were performed in all cases with or without anti-mycobacterial medication. However, NTM has never been described in the etiology of an infected CCAM. To our knowledge, our patient is the first reported case of a CCAM combined with MAC disease. Surgical resection is the preferable treatment for the exact diagnosis and immediate removal of the infectious focus, thus preventing complications related to the infection or the malformation itself.

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