Bronchial Atresia with Collapse of the Right Upper Lobe

-A Case Report-

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=Abstract=

우상엽의 허탈을 동반한 기관지 폐쇄증

-1례 보고-

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Congenital bronchial atresia is one of the rare bronchopulmonary anomalies which is thought to be caused by a vascular insult of uncertain timing during fefal development. It is defined as an anomaly which does not have communication between a segmental or lobar bronchus and the main airway. Because of the collateral ventilation, almost all of these cases show hyperlucency of the involved segment or lobe in chest roentgenogram. We report an extremely rare case of congenital bronchial atresia with collapse of the right upper lobe which was treated by surgical resection.

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Key words : 1. Bronchial atresia
2. Bronchial disease

CASE

A 27-year-old man was admitted to our hospital in May 1995 due to an abnormal haziness in the right upper lung field on chest roentgenogram found during an examination in our outpatient department. He was complaining of mild fever, productive coughing and pain in his right upper chest for the five days prior to his visit. For the past eight months, the patient has been suffering from hyperthyroidism and is being treated with propylthiouracil at another hospital, but no roentgenogram was given during his treatment period. Physical examination at our hospital revealed an exophthalmos, mild fever, and moist rale in the right upper lung field.

A chest computed tomogram revealed a heterogeneous soft tissue mass density around the right upper lobar bronchus and the collapse of the right upper lobe without any evidence of lymphadenopathy(Fig 1). In bronchoscopy, the opening of the right upper lobar bronchus was not evident(Fig 2). There was a

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Fig. 1. Preoperative chest roentgenogram showing abnormal hazziness in the right upper lung field

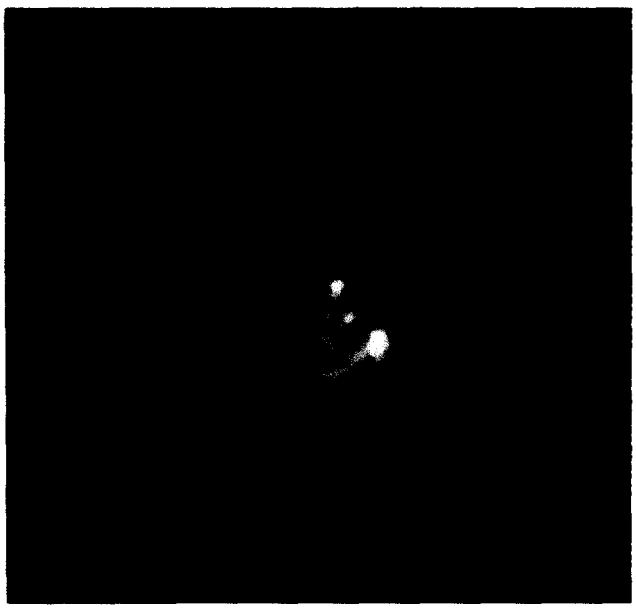

Fig. 2. Bronchoscopic finding. There is no bronchial opening to the right upper lobe.(arrow).

mild mucosal change in the region of the bronchial opening, but the results of the biopsy were normal. In the follow up chest

Fig. 3. The forceps indicate the remained right upper bonchial stump. The stump was totally occluded without any connection to the main bronchus.

roentgenogram, one week later, the radioopacity of the right upper lung had increased and the collapse had progressed.

The patient underwent thoracotomy under a one-lung ventilation. There were mild adhesions in the apex, a severe collapse of the upper lobe, and the interlobar fissures were complete. The lobar artery and vein were in normal positions but hypoplastic, and the diameter of the right upper lobar bronchus was about seven mm. When the bronchus was divided near the right main bronchus, the opening of the stump to the main bronchus was totally occluded(Fig 3) and the distal part of the bronchus was filled with yellowish, creamy mucus. When we tried to dissect the bronchial stump further to investigate the connection to the main bronchus, the remained stump was easily separated from the main bronchus. Examination of the resected specimen revealed the organizing pneumonia. The patient was discharged on the 14th postoperative day without any complications, and remains asymptomatic.

COMMENT

Congenital bronchial atresia is a rare anomaly characterized by a bronchocele caused by a mucus-filled, blindly terminating segmental or lobar bronchus, with hyperinflation of the obstructed segment of the lung. The first case of this disease entity was reported in 1953 by Ramsay and, in 1963, Simon and Reid established the criteria for diagnosis while describing their three cases at the segmental level[1]. Most recently, Mori and associates reviewed the English and Japanese literature and reported on a total of 101 cases[2].

There are two theories for probable causes of bronchial atresia[3]. The first theory proposes that a single vascular insult occurrs during the early phase(around the fifth week of gestation) of lung budding[4]. A nest of proliferating cells loses

connection with the distal tip of the developing bud and continues to branch independently. Consequently, normal branching distal to atresia is maintained without actual connection to the central airway[3]. The second theory suggests that the insult of the blood supply occurs after completion of bronchial branching, resulting in infarction and repair which occlude the lumen of the bronchus. This vascular insult would occur after the 15th week[1,5,6].

Aeration of the involved segment or lobe is maintained by collateral ventilations: 1) interalveolar pores of Kohn; 2) bronchoalveolar channels of Lambert; 3) interbronchiolar channels[7,8]. Hyperinflation of the lung occurs because each of the collateral pathways favors air movement into the obstructed segment by a check-valve mechanism. In lobar atresia, incomplete fissures between lobes ensure that the same pathways remain operative. But, although extremely rare, it is possible that trans-segmental interalveolar passage of air extends only to the boundaries of a lobe, so that occlusion of a lobar bronchus will always result in atelectasis if interlobar septa are intact[8], as in our case.

Although computed tomography and magnetic resonance imaging usually make an accurate diagnosis of bronchial atresia possible, it was difficult to correctly diagnose in our case because the lung had collapsed. It was only possible to make an accurate diagnosis by using bronchoscopy. Surgical treatment is not often indicated for asymptomatic patients. Moreover, although surgical intervention is required for patients with specific indications such as encroachment of normal lung tissue or infection as in our case, resection should be limited as much as possible to preserve the normal lung tissue[2,7].

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=국문초록=

선천성 기관지 폐쇄증은 발생학적으로 태아의 어떤 시기에 기관지 계통에 대한 혈관이 차단됨으로 인해 유발된다고 여겨지는 아주 드문 폐-기관지 질환의 하나로, 이 질환은 주기관지와 폐구역 또는 폐엽 사이에 기도의 연결이 되어 있지 않은 이상증으로 정의된다. 그간 보고된 모든 경우의 기관지 폐쇄증은 측부 순환이 존재함으로 인해, 폐쇄된 폐구역이나 폐엽이 흉부 단순 촬영상 과투과 음영을 보였다고 하였으나, 본원에서는 그간 이론적으로만 가능하다고 했던 폐엽의 허탈을 동반한 기관지 폐쇄증을 수술 치험하였기에 보고하는 바이다.

공 지 사 항

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