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Diagnosis of Unilateral Pulmonary Arterial Agenesis using Scintiangiography*

— A case report —

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〈국문초록〉

섬광심혈관조영술을 이용한 일측성 폐동맥형성 부전증의 진단

— 1예 보고 —

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일측성 폐동맥 형성부전은 대개 단독으로 오는 기형이지만 때로는 다른 선천성 심질환을 동반한다.

이는 증상없이 우연히 발견되기도 하고 반복되는 폐염증상을 나타내기도 한다.

단순흉부사진에서 환측 폐의 용적이 적어보일 때는 일단 감별진단으로 폐동맥의 형성부전을 생각하게 된다.

저자들은 반복되는 폐염으로 내원한 7세 소년에서 섬광심혈관조영술을 시행하여 선천성 우폐동맥 형성부전증을 진단하였기에 문헌고찰과 함께 보고하는 바이다.

A 7-year-old boy presented with productive cough and mild dyspnea. He had a history of intlermittant attacks of bronchopneumonia, but otherwise he was healthy. He had a small right lung on chest roentgenogram. The diagnosis of developmental hypoplasia of the right lung due to congenital absence of the right pulmonary artery was made by radionuclide angiocardiography and subsequently confirmed by cardiac catheterization and angiography.

Unilateral absence of a pulmonary artery is usually an isolated anomaly and sometimes associated with other cardiac anomaies.[1,2] Patients are often asymptomatic or present with recurrent lung infections. Chest roentgenograms might suggest hypoplasia or aplasia of a pulmonary artery, demonstrating a small lung. Perfusion lung scan may be used for diagnosis, but it has limited value in evaluating pulmonary arterial flow. Thus we chose radionuclide angiocardiography and obtained suquential dynamic images of pulmonary circulation. On radionuclide angiography, complete absence of the right pulmonary

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artery was demonstrated and confirmed the diagnosis. Those radionuclide angiographic images were very hepful in subsequent cardiac catheterization.

Case Report

A 7-year-old boy was admitted with a one month history of productive cough accompanying mild dyspnea. The patient was a healthy, rather thin looking boy. He was delivered normally and had no special problems, except for intermittent coughing, until admission. No family history of tuberculosis or asthma was found. BCG and all other vaccinations were done at appropriate times. Physical examination revealed coarse breath sounds with rhonchi and rales over the right lung. No cardiac murmurs were heard. The chest roentgenogram on admission showed mediastinal and tracheal deviation to the right. The right lung volume was small but was not hyperluclent. The right paratracheal area was widened and there was decreased aeration of the right upper lobe (Fig. 1). A diagnosis of bronchopneumonia was suggested and the decreased aeration in the right upper lobe was interpretated as atelectasis of upper lobe caused by either mucus plug resulting in recurrent pnbeumonia or compression of bronchus by tuberculous lymphadenitis. After symptomatic treatment of pneumonia, the patient's symptoms were relieved. Follow up chest roentgenogram showed no change compared with the initial roentgenogram. The possibility of nonpulmonary cause of the hypoplastic lung was then suspected and radionuclide angiocardiography was suggested before catheterization angiography because it was much simpler examination. The technique of radionuclide angiography was bolus injection of 10mci Tc-99m-tagged HSA (human serum albumin). A twenty-one gauge butterfly needle attached to an extension tube and a three-way stop cock was fixed in a antecubital vein. A small volume of radionuclide injection was

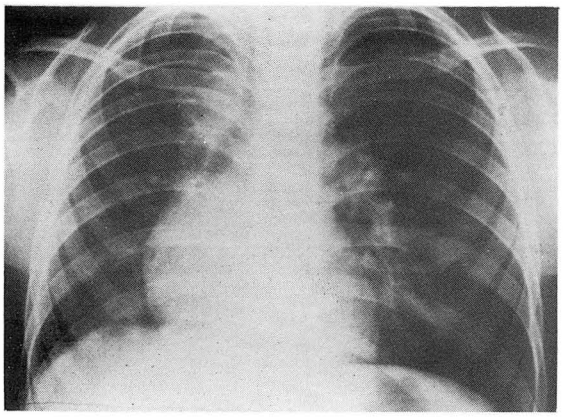

Fig. 1. Chest roentgenogram showing mediastinal shift to the right and a small right lung. Right hilar vessels are obscured by the heart. Some decreased aeration is seen in the right upper lobe.

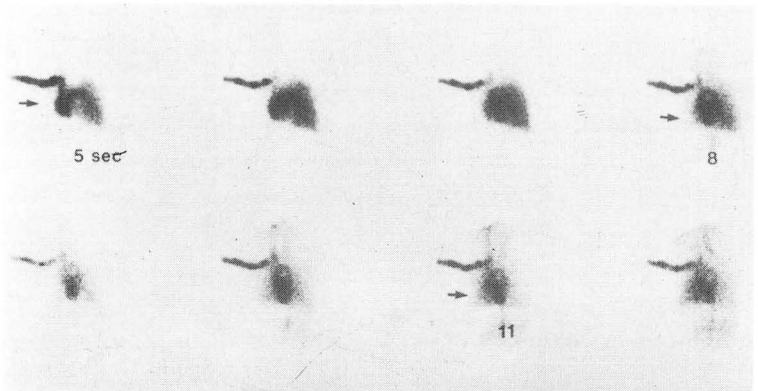

Fig. 2. Radionuclide angiocardiogram. 5 sec after radionuclide injection in antecubital vein, SVC, RA, RV, left main pulmonary artery and the activity in the left lung clearly seen. No activity is present in the right lung (arrow, left upper row). 8 sec image showing LV and aorta, and some faint activity in right lower lung (arrow, right upper low). The right lung activity become distinct in sequential images (arrow, lower low).

followed by 5 ml of normal saline flush to facilitate the bolus injection. The images were taken at one second intervals for 24 seconds. Five seconds after injection, the antecubital vein, superior vena cava, right ventricle, left main pulmonay artery and the left lung were clearly visualized, but no activity was seen in the right lung (Fig. 2). Subsequent sequential images showed then faint activity in the right lower lung progressively increasing in activity. The delayed uptake in the right lung was interpretated as collateral supply from the aorta. Cardiac catheterization was done and demonstrated complete absence of the right main pulmonary artery and marked dilation of the left pulmonary artery (Fig. 3). On the late sequential levocardiogram phase, dilated right internal mammary artery and mesh-like collateral vessels from the thoracic aorta supplied the right lung (Fig. 4). These corresponded to the delayed appearance of activity in the right lung on radionuclide angiocardiography.

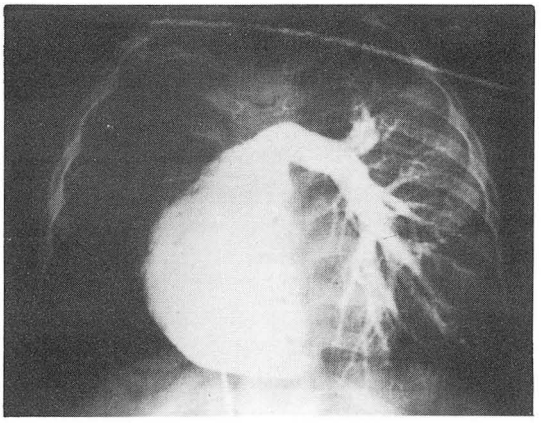

Fig. 3. Right ventricle cardiac catheterization. Complete absence of the right pulmonary artery and dilated the left pulmonary artery.

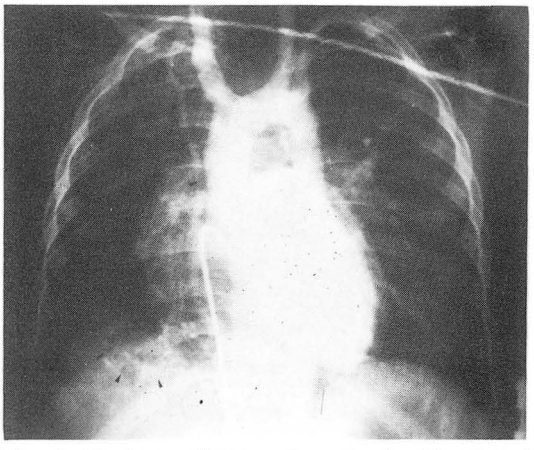

Fig. 4. The levocardiogram phase showing dilatation of the right internal mammary artery (upper arrow heads) and mesh-like abundant collaterals from descending aorta (lower arrow heads).

Discussion

Unilateral absence of pulmonary artery may occur as an isolated or in combination with other cardiac anomalies (usually tetralogy of Fallot) or vascular abnormalities.[1,2] Since the single hypoplastic or aplastic pulmonary artery often has no specific associated symptoms, the diagnosis depends on the suspicion aroused by conventional chest roentgenogram. The chest roentgenograms show a small lung with mediastinal shift to the affected side, decreased vascularity, and a spidery appearance of vessels.[3] However, these findings are identical to collapse of the one lung lobe on a hypoplastic thorax and an erroneous diagnosis coud be made. In our case, initially the diagnosis was the right upper lobe atelectasis due to either mucus plugging following repeated pneumonia or compression of bronchi by tuberculous nature. Persistent small lung volume without other clinical problems had raised the suspicion of non-pulmonary cause of hypoplastic lung. For diagnosing pulmonary vascular malformations, cardiac catheterization and angiography should be done. It is an invasive and complicated method in the pediatric age group. Perfusion lung scan commonly is used for detection of pulmonary arterial flow disturbances.[4] However, lung perfusion scan revealed nonspecific absent or decreased uptake defects which could not differentiate absence of pulmonary artery from bronchial obstructions producing decreased pulmonary arterial flow. For this reason, radionuclide angiography is very useful for diagnosis of abnormalities of pulmonary arterial flow pathways. Normal radionuclide angiographic circulation times from superior vena cava to right ventricle is 1.5 sec, to lung 3 sec, to left ventricle 6 sec, and to aorta 7 sec. The time course of activity depends on the age of the patient and adequacy

of bolus injection.[5] Both lungs should have simultaneous isotope accumulation with equal activity distribution. In aplasia of the right pulmonary artery, the sequential image showed good visualization of the SVC, right strium, right ventricle, left pulmonary artery and simulaneous activity in the left lung. But no activity in the right lung was found. This is different from the perfusion lung scan which showed uptake defect, however, a static image is not like the dynamic one seen in radionuclide angiocardiography. Moreover, the other useful information obtained in radionuclide angiocardiography was delayed appearance of activity in the involved lung in later sequential images which proves the existence of systemic collateral supply of the diseased lung from aorta. This finding was supported by the cardiac catheterization angiography.

In conclusion, when there is a solitary small lung in a patient with recurrent pneumonia, developmental anomaly of pulmonary artery should be considered and radionuclide angiography should be done first. If the finding is confirmed no further ivestigation is needed.

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