□ Case Report □

Anomalous systemic arterial supply to lung without sequestration in an infant who has congenital heart disease : a case report

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Systemic arterial supply from the aorta to the lung is a rare congenital anomaly within the spectrum of bronchopulmonary sequestration according to Pryce's terminology. We describe our experience of this anomaly in an infant with congenital cardiac disease confirmed by multidetector CT scan. We found a systemic arterial supply from the aorta to the right lower lobe of lung without right lower lobar pulmonary artery and bronchopulmonary sequestration. This combination of congenital anomaly is most rare form. **(Korean J Pediatr 2006;49:895-897)**

Key Words : Bronchopulmonary sequestration, Atrial septal defect, Pulmonary artery

Introduction

Aberrant systemic arterialization to the lung without sequestration is the rarest form of congenital anomaly[1-4]. In this rare abnormality, arterial supply of one or more of the basal segments of the lower lobe comes from an aberrant artery arising from aorta. In adult, this anomaly requires surgery because of frequent hemoptysis caused by localized pulmonary hypertension.

In this case report, we noted that the anomalous systemic arterial supply to the right lung base without sequestration in an infant with congenital heart disease during the time of follow ups because of her recurrent lower respiratory tract infections.

Case Report

7 months old female infant came to our hospital due to 2 days of fever and cough. When she was 3 months of age, she was treated for her pneumonia and acute gastroenteritis and when she was 4 months of age, she was also treated for her CMV pneumonia. At that time she was also diagnosed having a congenital heart disease(Atrial septal defect, secundum).

On physical examination, she was acute ill looked and her body temperature was 37.8℃, pulse rate was 156/min, respiratory rate was 36/min, blood pressure was 112/60 mmHg. Coarse breathing sound with rales on right lung field and regular heart beat with systolic murmur (grade 2-3/6) on left upper sternal border were noted but no chest wall retraction.

Chest X-ray showed pneumonic infiltrations on both lung fields and cardio-thoracic ratio was 52.1% (Fig. 1).

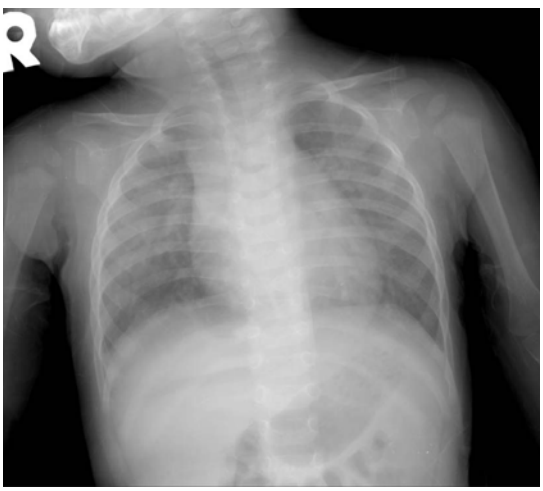

Fig. 1. Chest PA shows both lung infiltration at the present of admission.

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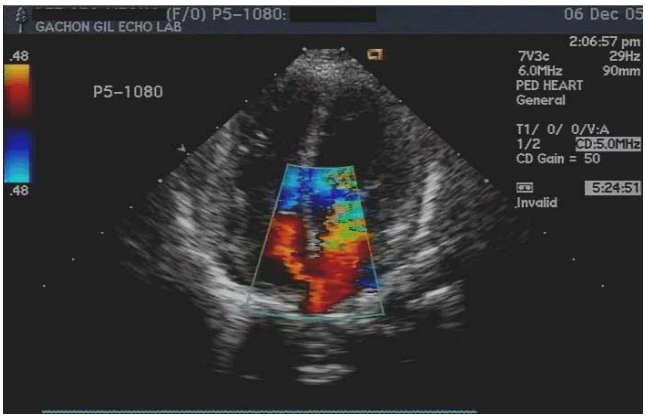

Fig. 2. Echocardiography (apical 4 chamber view) shows atrial septal defect.

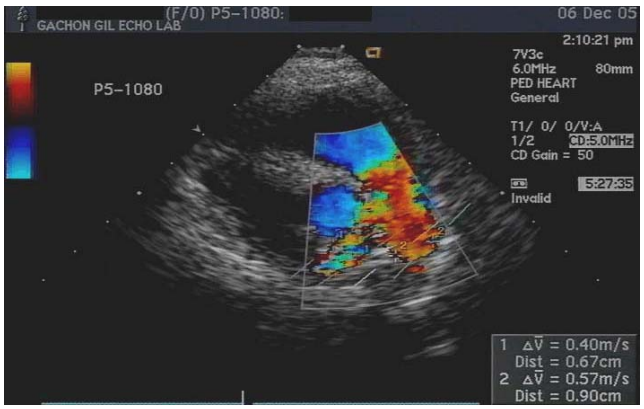

Fig. 3. Echocardiography (parasternal short axis view) shows hypoplastic right pulmonary artery.

On her echocardiography findings showed atrial septal defect (Fig. 2) without a significant interval change (size 6-7 mm) and right pulmonary artery size was smaller than left pulmonary artery (Fig. 3).

To evaluate the cause of recurrent lower respiratory tract infections without any evidence of heart failure, CT angiography was done. In the CT scanning, right lower lung was supplied by an aberrant systemic artery arise from celiac trunk of aorta without any evidence of pulmonary sequestration and a right lower lobar pulmonary artery was not noted (Fig. 4, 5).

Discussion

The systemic arterial supply to the lungs, especially to the basal segment of the lower lobe without pulmonary sequestration has been categorized as type I according to Pryce's nomenclature[5]. However, in some rare cases, pulmonary artery supplies to the basal segment were not noted either[6]. In our case, there was an atypical (hypoplastic) right pulmonary artery without lower lobar branch of it.

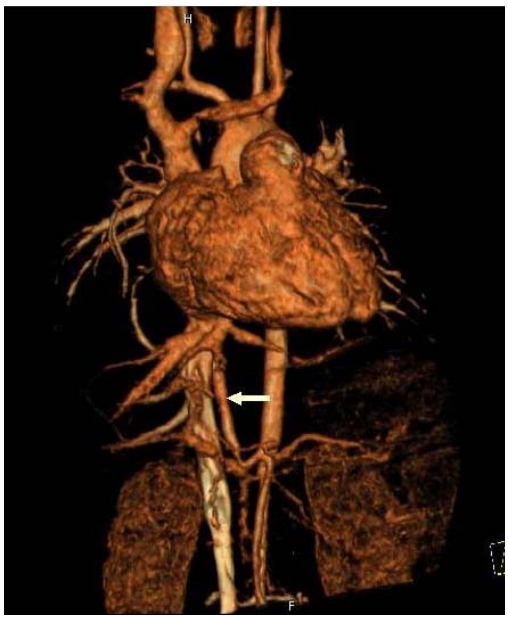

Fig. 4. 3-D reconstriction of multidetector CT scan shows aberrant artery originated from abdominal aorta (arrow).

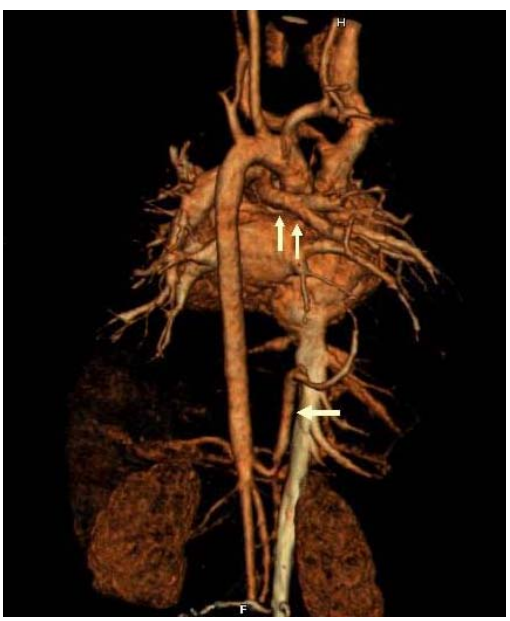

Fig. 5. The backside of 3-D reconstruction of multidetedtor CT scan shows aberrant artery (arrow), absence of right lower pulmonary artery (double arrow).

The etiology of the systemic arterial supply to the basal segment of the lung without a pulmonary artery supply has been still contentious[7]. Although the exact cause of this lesion is unknown, persistence of an embryonic con-

nection between aorta and pulmonary parenchyma might lead to this anomaly[8]. The basal segments of the left lower lobe are involved most frequently[9, 10]. A similar acquired lesion of pseudosequestration has been described[11]. The systemic arterial supplies of the pseudosequestration are pleural due to hypervascularization of systemic arteries supplying to lung as well as chest wall. This condition may lead recurrent pulmonary infections. In our case, there were also recurrent pulmonary infections and normal bronchial trees. but we believe that the lesion of our case may be congenital because of her age.

Most of adult patients are asymptomatic or may have recurrent hemoptysis, however most common clinical manifestation among pediatric patients is a cardiac murmur[9]. In our case, the patient also had cardiac murmur, but its origin was not clear, because this patient also had a cardiac anomaly-atrial septal defect and a hypoplastic right pulmonary artery.

The anastomosis procedure to preserve normal circulation of lung would not so effective treatment[7]. It is not clear whether an aberrant systemic artery could exchange blood gas in alveolar-capillary region, because its arterial walls might be thickened and sclerosing change by systemic artery pressure.

The 64 sliced multidetector CT is very useful for diagnosis of various cadiovascular problems[12]. In this case, we could not find the exact reason of her recurrent pneumonia only by chest X-ray and echocardiography. So for better evaluation of unexplained combination of cardiac and pulmonary problem, multidetector CT angiography would be a good choice.

We report a case of anomalous systemic arterial supply to lung without sequestration in an infant patient who had recurrent pneumonia infections, not matched well with her present cardiac condition and this case was confirmed by the multidetector CT scanning evaluation.

한 글 요 약

선천성 심장질환을 가진 영아에서 발견된 폐 격리증을 동반하지 않은 폐의 이상 체 동맥 기시 1례

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폐 격리증이 없는 정상적인 폐에 체 혈관이 이상 기시하는 것은 매우 드문 질환이다. 저자들은 반복적인 하기도 감염을 보이는 영아에서 선천성 심장질환의 상태가 본 하기도 감염을 나타낼 정도로 심하지 않음을 이상히 여겨 컴퓨터 단층 촬영을 시행하였다. 그 결과 폐 격리증이 없는 폐의 이상 체 동맥 기시가 우측 폐하 엽에 존재함과 동시에 우측 하엽 폐동맥이 없는 사실을 확인하게 되었다. 어린 나이에 이 같은 선천성 질환의 조합은 이전에 보고 된 바가 없는 매우 드문 질환으로 생각된다.

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