Korean Journal of Pediatrics Vol. 51, No. 8, 2008
DOI : 10.3345/kjp.2008.51.8.892
Case report

A case of acute respiratory distress syndrome associated with congenital H-type tracheoesophageal fistula and gastroesophageal reflux

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= Abstract =

H-type tracheoesophageal fistula (TEF) is extremely rare in infants and children, and clinical manifestations of this condition are diverse based on its severity. Some cases of congenital TEF diagnosed in adulthood have been reported, which indicate the difficulty of early diagnosis of this disease. Gastroesophageal reflux (GER) may induce chronic aspiration, pulmonary aspiration, apparent life-threatening events, and failure to thrive. We report a 5-month- old boy whose recurrent pneumonia and wheezing did not improve under usual treatment and led to acute respiratory distress syndrome. He was found to have severe GER on the second-trial of the esophagogram and was eventually revealed to have congenital H-type TEF upon repeated evaluation. **(Korean J Pediatr 2008;51:892-895)**

Key Words : Acute respiratory distress syndrome, H-type tracheoesophageal fistula, Gastroesophageal reflux

Introduction

H-type tracheoesophageal fistula (TEF) or TEF without atresia (type H or N) is extremely rare in infants and accounts for approximately 4% to 5% of all congenital tracheoesophageal malformations[1]. The clinical features are variable according to its severity. The most common features are recurrent respiratory symptoms, coughing and aspiration during feeding, abdominal distension, repeated cyanosis, and hypersalivation. The early diagnosis of this disorder is difficult and some rare cases may remain undiagnosed until adulthood[2-5]. These undetected fistulas may be presented as chronic lung disease because repeated aspiration can lead to recurrent infection and bronchiectasis[4].

Gastroesophageal reflux (GER) is common in infants and children, and most children will outgrow their symptoms by 12 to 18 months of age. But severe GER may induce complications such as reflux esophagitis, Barrett's esophagus, esophageal stricture, pulmonary complications, apnea, failure to thrive. Pulmonary aspiration, apparent life-threatening events and failure to thrive are well known complication of severe GER, and are current indications for fundoplication[6].

Here, we report a 5-month-old male infant whose recurrent pneumonia with wheezing did not respond to usual treatment of pneumonia and asthma, and eventually lead to acute respiratory distress syndrome (ARDS). His H-type TEF and GER was confirmed by repeated barium-esophagogram.

Case report

A 5-month-old boy was referred to our hospital for an exacerbation of pneumonia with recurrent wheezing, which did not respond to usual treatment for asthma. He was born by Cesarean delivery due to cephalopelvic disproportion at 38 weeks at a local obstetric clinic, and weighed about 2,800 g. His mother was healthy with no history of any drugs except iron replacements. No specific problems were noted during pregnancy. He developed respiratory difficulty on the first day after birth and was diagnosed to have pneumonia in both lung fields. He was transferred to tertiary class hospital, and admitted for one month. On suspicion of his having recurrent aspiration, he went through some examinations including brain neurosonography, echocardiography, barium esophagogram, but no abnormal results were found. He again had pneumonia a month later, and he was discharged after 10-day symptomatic treatment.

Received : 8 February 2008, Revised : 21 May 2008, Accepted : 20 June 2008
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This study was supported by research funds from Dong-A University

He was admitted to a local hospital for pneumonia and received ampicillin and chlarythromycin antibiotic therapy for one month before he was transferred to our hospital. He had dyspnea accompanied by persistent wheezing. Though he was treated with short-acting β-agonist, intravenous methyl-prednisolone and continuous infusion of aminophylline, his wheezing did not improve at all.

On admission, his respiratory rate was 40-50 breath/min, and his oxygen saturation was 98% without oxygen supplement. Other vital signs were within normal limits (blood pressure 100/60 mmHg, heart rate 130/min, body temperature 37.0℃). On auscultation, rales with mild wheezing were found on both lung fields, but they gradually improved by asthma treatment. His respiratory difficulty was almost resolved on the third day of admission, but on the forth day respiratory difficulty and fever were developed again. After milk feeding, chest X-ray showed increased extent of bilateral ill-defined air space nodules and ground glass opacity, associated with underlying focal consolidations prominently defined in the right upper lung field (Fig. 1). He was diagnosed with ARDS, and received ventilator care for one month, and was weaned successfully. During treatment, we found that percutaneous oxygen saturation and heart rates were decreased with medications given through nasogastric tube. After extubation, we checked barium esophagography, which showed severe GER (Fig. 2).

His choking symptoms and respiratory difficulty didn't improve even though he was treated with prokinetics, H_2-blockers and proton pump inhibitors. According to the recently reported indications[6], he went on Nissen fundoplication. The operation was successful, and there was no

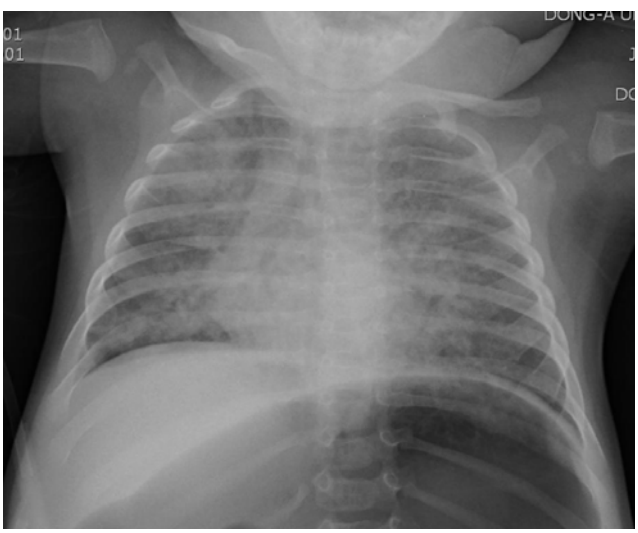

Fig. 1. Chest X-ray shows increased extent of bilateral ill-defined air space nodules and ground glass opacity, associated with underlying focal consolidations prominently defined in the right upper lung field, which is compatible to ARDS.

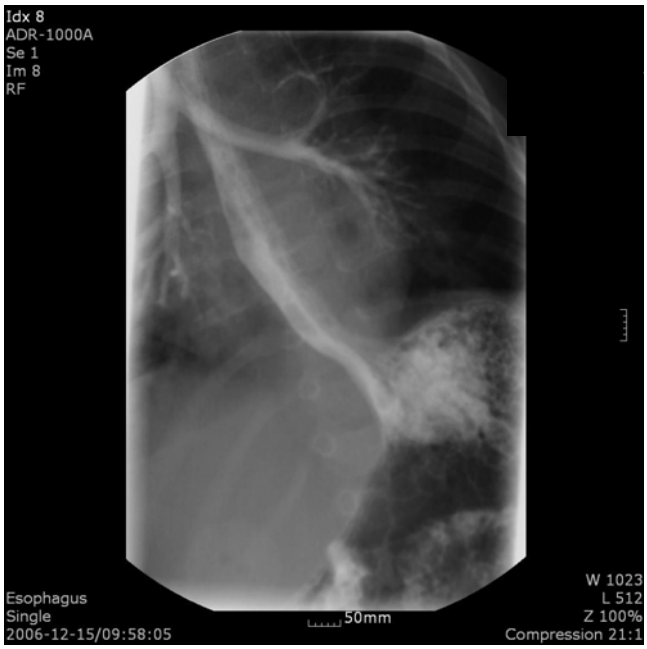

Fig. 2. Severe GER was found on the second trial of esophagogram. Barium bronchogram shows that reflux extends to bronchiole levels. At the time of this test, no other abnormality was seen.

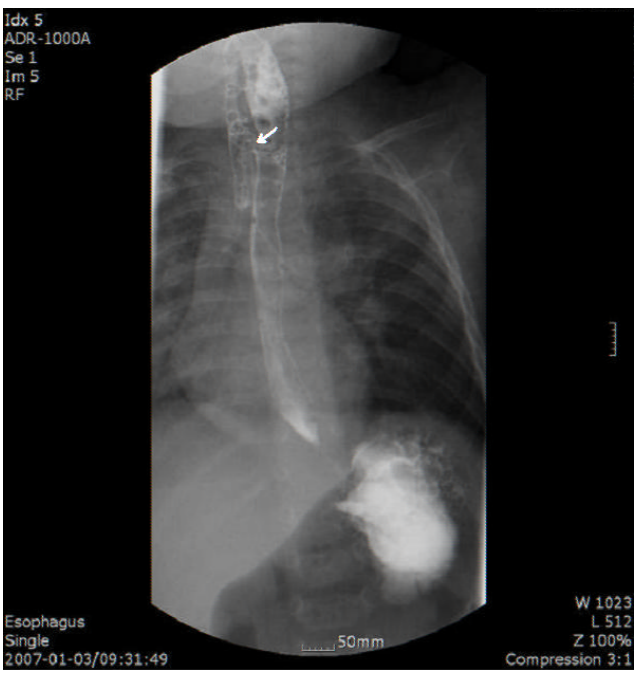

Fig. 3. H-type TEF was found on the third trial of esophagogram.

complication like hiatal hernia. But after the operation, he still showed symptoms of continuous coughing and choking. He went through barium esophagogram again, which revealed H-type TEF (Fig. 3). After TEF repair operation, his respiratory symptoms were completely disappeared, and there was no more recurrent respiratory infection. Now his growth became normal.

Discussion

Congenital TEF without esophageal atresia (EA) (H-type) is very rare esophageal anomaly, which is also hard to diagnosis. Neonates with H-type TEF usually have choking episodes and cyanotic spells[7]. In some cases, infants may show its symptoms when they are older with recurrent or multiple pneumonias[8].

Because of its difficulty to detect, congenital TEF should be considered in the diagnosis of infants and young adults with respiratory distress or recurrent pneumonia. Patients who are suspected of having an TEF should check plain x-ray film, which may show evidence of aspiration and gastric dilatation. The diagnosis can be established by contrast study like esophagogram, which shows the fistula in 80% of cases[9]. Tc-99m sulfur colloid scintigraphic technique has been used to confirm the diagnosis non-invasively[10]. Bronchoscopy and esophagoscopy can confirm the diagnosis by demonstrating the fistula opening[11]. Computed tomography may be helpful in identifying the TEF[12]. Prompt surgery of TEF is the optimum treatment. Some complications after TEF repair have been reported like post-surgery RDS, post-surgery GER, stricture and stenosis, tracheomalacia, recurrent TEF, pulmonary hemorrhage, and growth retardation and esophageal motility disorders, which may be life-threatening[12-17]. Recent studies show good results after endoscopic repair[18].

For the recent strategy for GER, many authors concerns about fundoplication associated complications in children[20]. Increased hospitalization due to pneumonia, respiratory distress, and failure to thrive are the major complications of fundoplication. There was some great advances in medical treatment, so many reports suggest that the fundoplication should be performed only in case with well documented GER and have failed medical management[6] like our patient. Our patient's result was good even though his TEF was found quite late. Follow up was made about 2 years, and he didn't show any other complication after 2 major operation.

In this case, the patient had both GER and congenital TEF. Either can contribute to severe aspiration and lung damage, which eventually lead to ARDS. We don't know which factors had more influenced on the patient whose condition grew more serious to ARDS; maybe GER was the factor that lead the cascade and the congenital TEF contributed a little, or maybe the opposite theory would be right; or both contributed equally. Though there are many reports of GER as a complication of TEF repair operation[15], case of both GER and congenital TEF is rare. There is a report that found a 10% incidence of GER in a series of 30 fistulae[19].

This case presents the worst result of recurrent aspiration made by congenital H-type TEF and severe GER, which were detected after several times of study.

한 글 요 약

급성호흡곤란증후군을 초래한 위식도역류와 H-형태의 선천기관식도루 1예

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최희원 · 김묘징 · 정진아

H-형태의 식도기관루 또는 선천 식도 폐쇄 없는 식도 기관루는 소아에서 매우 드문 형태이며, 그 임상 양상도 질환의 중등도에 따라 다양하게 나타난다. 몇몇의 성인에서 발견된 선천 식도기관루는 이 질환이 조기 진단되기 어려움을 반영한다고 볼 수 있다. 위식도역류는 만성적인 흡인을 일으킬 수 있으며 폐흡인, 생명이 위험할 정도의 호흡기증상 또는 성장장애 등이 흔히 일으킬 수 있는 중한 합병증이다. 저자들은 통상의 치료에 반응을 보이지 않는 반복적인 폐렴과 천명을 동반하고 급성호흡곤란증후군으로까지 진행하였으며 반복된 식도조영촬영에서 심한 위식도역류와 H-형태의 식도기관루가 발견되었던 5개월 남아의 증례를 경험하였기에 이를 보고하는 바이다.

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