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Association of Communicating Extralobar Bronchopulmonary Sequestration with Intralobar Sequestration in a Patient Who had Anal Atresia**

İbrahim Karaman[1*], Selma Uysal Ramadan[2], Ayşegül Zenciroğlu[3], Özlem Balcı[4], Ayşe Karaman[5]

[1,4,5;] University of Health Sciences Turkey, Ankara Dr Sami Ulus Maternity and Children Health and Research Application Center, Departments of Pediatric Surgery, Ankara, Turkey
[2;] University of Health Sciences Turkey, Ankara Keçiören Training and Research Hospital, Department of Radiology, Ankara, Turkey
[3;] University of Health Sciences Turkey, Ankara Dr Sami Ulus Maternity and Children Health and Research Application Center, Departments of Neonatology, Ankara, Turkey

ORCID; 0000-0001-7490-4158, 0000-0002-3653-7892, 0000-0002-3488-4962, 0000-0002-4926-5451, 0000-0001-6860-1222

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Abstract; Bronchopulmonary sequestration is an uncommon anomaly in children. It is characterized by nonfunctioning pulmonary tissue that is not connected to the normal tracheobronchial tree, and its blood supply derived from a systemic arterial source. A 2900-gram male term baby was referred to our clinic with the diagnosis of anal atresia. A diverting sigmoidostomy was performed. He was started to oral feeding on the second postoperative day, but patient's general condition was impaired and right lower lobe pneumonia was documented on chest X-ray. A thoracic CT scan demonstrated multiple cysts and pneumonic consolidation, which indicated abscess at right lower lobe. The thoracic CT scan also showed bronchus like structure, which was entering to the esophagus. An esophagogram showed a fistula from distal esophagus to the right lower lobe of the lung. At thoracotomy, a connection between the right lower lobe of the lung and distal of the esophagus by its bronchus was noted. An anomalous artery was found, originating from thoracic aorta. The fistula was divided from esophagus and right lower lobectomy was performed. Also basal region of the right middle lobe were not inflated with ventilation and congested with a clear demarcation from normal appearing lung. This situation was evaluated as an intralobar sequestration and resection was performed. We present here association of communicating extralobar sequestration with intralobar sequestration in a patient who had anal atresia. To the best of our knowledge this combination has never been reported before.

INTRODUCTION

Bronchopulmonary sequestration (BPS) is an uncommon anomaly in children. It is characterized by nonfunctioning pulmonary tissue that is not connected to the normal tracheobronchial tree, and its blood supply derived from a systemic arterial source [1].

Bronchopulmonary sequestrations are subdivided into extralobar (eBPS) and intralobar (iBPS) types. eBPS is completely separate from the normal lung and invested by its own visceral pleura. eBPS is more common on the left side and most of them are found in the left costophrenic angle. iBPS is incorporated within the normal surrounding lung and there isn't any anatomic plane of clevage between the sequestered and the normal pulmonary parenchyma and most of them are found at the posterior basal segments of the lower lobes [1,2,3].

The coexistence of eBPS and iBPS is extremely rare [2]. We present here association of communicating eBPS with iBPS in a patient who had anal atresia. To our knowledge this combination has never been reported before.

Case report

A 2900-gram male term baby, born through spontaneous vaginal delivery to a mother of 22 years of age in the first pregnancy was referred to our clinic with the diagnosis of anal atresia. Anal atresia and right nonpalpable testis were detected on physical examination. Meconium was observed at external urinary meatus and invertogram showed high type anorectal malformation. The abdominal ultrasound and echocardiogram were normal.

A diverting colostomy was performed. In the follow up period of the patient, oral feeding related recurrent pulmonary infections was developed. A chest radiograph showed necrotizing pneumonia on the right lower lobe. A thoracic CT scan demonstrated multiple cysts and pneumonic consolidation, which indicated abscess at right lower lobe. The thoracic CT scan also showed bronchus like structure, which was entering to the eosophagus. An eosophagogram showed a fistula from distal eosophagus to the right lower lobe of the lung (Figure 1).

*Corresponding author: Ibrahim Karaman, E-mail; ibrahimkaraman@hotmail.com, http://dx.doi.org/10.29228/jamp.44262
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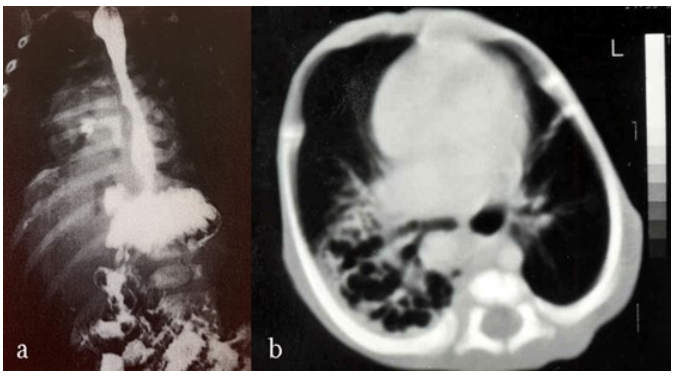

Figure 1: (a) An upper gastrointestinal contrast study showing an esophageal bronchus, (b) A thoracic CT scan demonstrated multiple cysts and pneumonic consolidation, which suggested that abscess. The thoracic CT scan also showed bronchus like structure was entering the esophagus.

He was operated when he was 27 days old. At thoracotomy, the right lower lobe of lung was sequestrated and connected to the distal esophagus by its bronchus. Esophageal bronchus of the sequestrated right lower lobe was divided into two branches in the lobe, as seen in the thorax CT. An anomalous artery was found, originating from thoracic aorta. The fistula was divided from esophagus and right lower lobectomy was performed. Microscopic examination of the eBPS revealed that pulmonary parenchyma was normal except the multiple spaces, which were filled by purulent material. Also basal region of the right middle lobe were not inflated with ventilation and congested with a clear demarcation from normal appearing lung (Figure 2). A large vessel feeding to the mass was detected but its origin could not be identified. This situation was evaluated as an iBPS and resection was performed. In the pathologic examination, normal pulmonary parenchyma, which had no communication with tracheobronchial tree, was demonstrated. The patient died on postoperative seventh day due to Klebsiella sepsis.

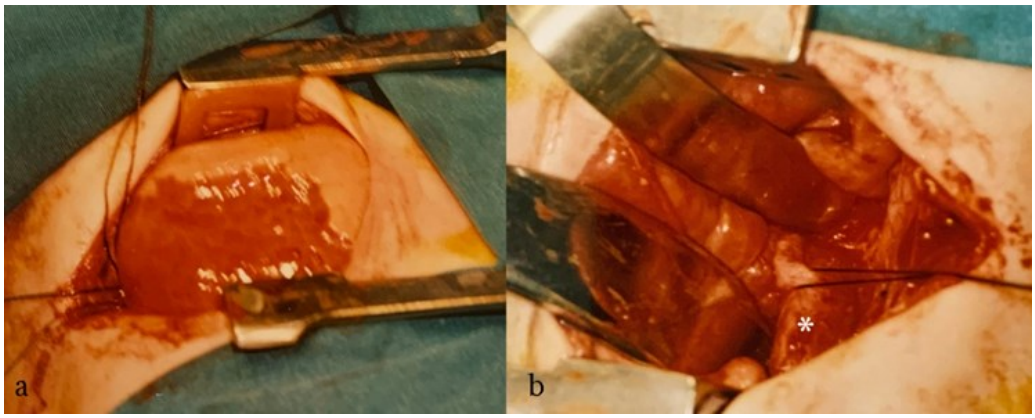

Figure 2: (a) Basal region of the right middle lobe were congested with a clear demarcation from normal appearing lung. (b) Intraoperative view of esophageal bronchus (suspended with silk suture), which provides the link between bronchopulmonary sequestration and esophagus (*).

DISCUSSION

eBPS occur on the left costophrenic sulcus from 80 to 90 % of the patients and eighty per cent of affected patients are boys [3]. The sequestrated lung tissue can also present at intradiaphragmatic or subdiaphragmatic location [4]. A large number of anomalies associated with eBPS have been reported. Additional anomalies such as diaphragmatic hernia, pectus excavatum, congenital heart disease, esophageal atresia, foregut duplications and vertebral anomalies were reported from 15 to 60 % of patients [1,3]. Most iBPS are found within the basal segments of lower lobe, with two third occur at the left side. Associated anomalies are rarely reported with iBPS [1].

There are several theories about the development of the sequestration. The most widely accepted theory is that an accessory lung bud arises from foregut is separate from the normal lung and becomes a pulmonary sequestration and this accessory lung bud keeps its systemic blood supply derived from the surrounding thoracic mesenchymal tissue. If sequestrated lung bud forms before the development of pleura, it becomes an iBPS. If the accessory bud forms after the development of pleura, it becomes invested by its own pleura and an eBPS is occur [5].

But, there are some theories about iBPS, which indicates that iBPS is an acquired one. According to this theory, bronchial obstruction caused by aspiration or inflammation is leads to formation of iBPS. It was claimed that chronic infection occluding the pulmonary artery and this situation is leading to neovascularization from the systemic circulation. However, as in our case, neonatal cases suggest congenital origin [6].

Both iBPS and eBPS may communicate with the eosophagus or the stomach and this situation is called by communicating bronchopulmonary foregut malformation [4]. This communication may lead infection of the sequestrated lung, which causes symptoms early period as our patient.

The association of iBPS with eBPS is extremely rare. We presented here association of communicating eBPS with iBPS and anal atresia. To the best of our knowledge this combination has never been reported.

Conflict of interest

There is no conflict of interest among the authors of the article.

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