

[CASE REPORT]

An Adult Case of Pulmonary Artery Sling Accompanied by Tracheobronchomalacia

Sahoko Imoto¹, Ryosuke Satomi¹, Mayuko Watase¹, Matsuo So¹, Hiroaki Murakami¹, Sakiko Hosoo¹, Iio Miwa¹, Kazuyuki Fujimoto¹, Shigenari Nukaga¹, Kazuma Yagi¹, Sota Oguro², Takahiko Oyama¹, Ryoichi Kato¹ and Yoshitaka Oyamada¹

Abstract:

Pulmonary artery (PA) sling is a congenital disease in which the left PA abnormally arises from the right PA and is usually diagnosed during the infantile period. We present an adult case of PA sling accompanied by tracheobronchomalacia found in a 49-year-old woman with a history of recurrent pneumonia. Computed tomography of the chest showed that the left lung was nourished by two aberrant PAs. Bronchoscopy demonstrated achondroplasia of the trachea and the right bronchus, which we speculate to have resulted in their stenosis. The recurrent pneumonia was attributable to these tracheobronchial structural abnormalities; we therefore stress the importance of focusing on the anatomic abnormalities in such cases.

Key words: pulmonary artery sling, recurrent lung infections, tracheobronchomalacia

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Introduction

Pulmonary artery (PA) sling is a congenital malformation defined as anomalous origin of the left PA from the right PA. This aberrant left PA crosses above and over the right main-stem bronchus or at the point at which it passes between the distal trachea and esophagus (1). In most cases, progressive respiratory symptoms (e.g., stridor, wheezing, and cyanosis) due to airway stenosis by the surrounding aberrant left PA or by the accompanying tracheobronchomalacia develop during the few first weeks or months of life. Ventilatory support may be necessary if the airway stenosis is very severe (2). PA sling is rarely reported in adulthood (3-10).

Case Report

A 49-year-old woman with a history of recurrent pulmonary infections was found to have two cavities with fluid collection in the right upper lung field on chest radiography

(Fig. 1). She had contracted pneumonia at 32 years of age and developed a lung abscess at 46 years of age, both of which required medical treatment in a hospital. She had a persistent cough with sputum that was being treated on an outpatient basis and had come for a follow-up. She had no other symptoms, and a physical examination revealed no significant findings. She was diagnosed with an indolent infection in the cavities.

Chest computed tomography (CT) (Fig. 2) demonstrated two cavities with thickened walls in the right lung that could be attributed to the previous pulmonary infections. Coincidentally, the left lung was found to be nourished by two aberrant PAs on CT (Fig. 2B, C). The superior PA originated from the pulmonary trunk (Fig. 2B), while the inferior PA arose from the right main PA (Fig. 2C). The red arrow in Fig. 2C indicates the same blood vessel as indicated by the red arrow in Fig. 2F; the inferior PA crosses the right main bronchus and distal trachea dorsally and the esophagus ventrally. The trachea was bifurcated at a more caudal level than usual, and the right main bronchus showed stenosis (Fig. 2D). These CT findings were consistent with a diagno-

¹Department of Respiratory Medicine, National Hospital Organization Tokyo Medical Center, Japan and ²Department of Radiology, National Hospital Organization Tokyo Medical Center, Japan

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Correspondence to Dr. Ryosuke Satomi, r_satomi@hotmail.com



Figure 1. Chest X-ray shows two cavities in the right upper lung field. One cavity contains a small amount of fluid. The mediastinum is shifted to the right.

sis of PA sling. The right lung was hypoplastic, along with incomplete lobation.

To identify possible pathogens causing the infection in the cavity, we performed bronchoscopy (Fig. 3), which revealed a diverticulum on the right wall of the distal trachea at the level at which the orifice of the right main bronchus normally exists. The right main bronchus was stenotic because of achondroplasia, resulting in its collapse during expiration. It was difficult to insert a 6-mm-diameter bronchoscope into the right main bronchus. Therefore, a thinner scope with a diameter of 4 mm was required to obtain microbiologic specimens from the cavity. In addition, we performed a lung biopsy, scraping, and washing after visualizing the wall of the cavity by transtracheobronchial endoscopic ultrasonography. The left bronchi were normal. Based on these findings, we diagnosed the case as an adult case of PA sling.

A pulmonary function test revealed a well-maintained vi-

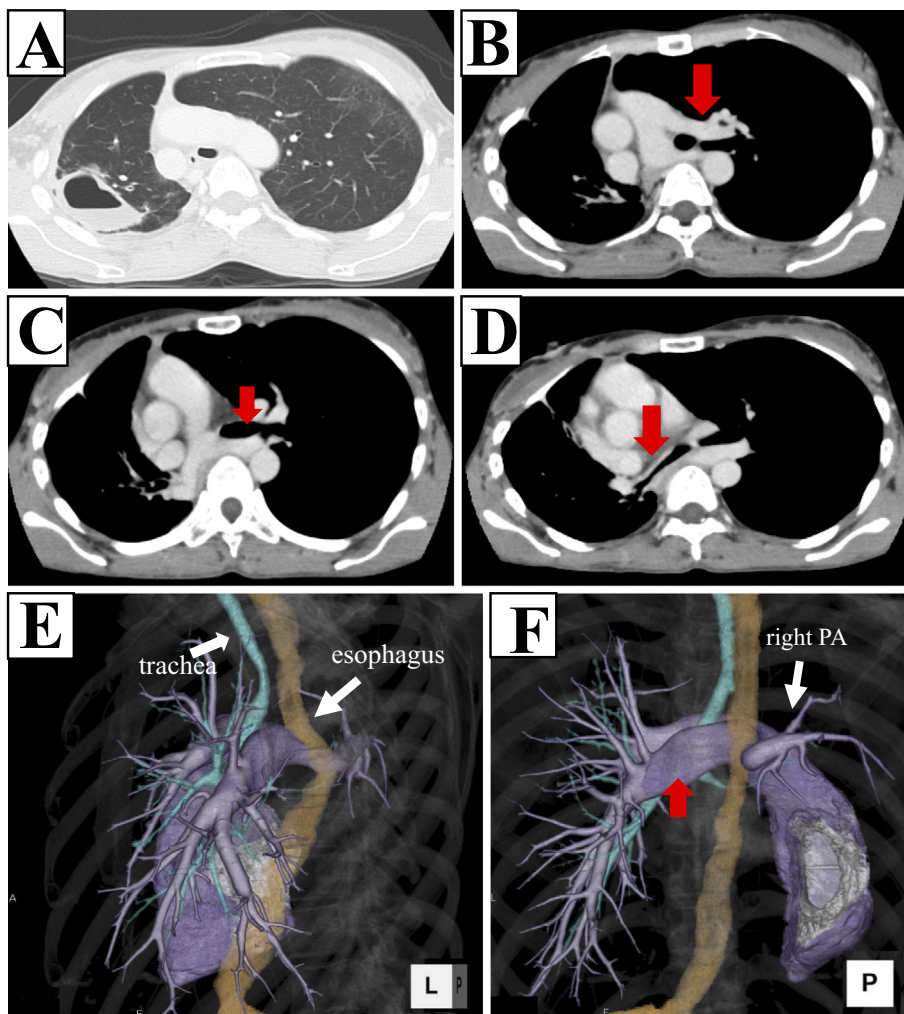


Figure 2. Chest computed tomography show a thickened-wall cavity with fluid collection in the right lung (A) and two aberrant left PAs: the superior PA originating from the pulmonary trunk (B: arrow) and the inferior PA arising from the right PA (C: arrow). The right bronchus is stenotic (D: arrow). Three-dimensional computed tomography shows that the left inferior PA runs between the trachea and the esophagus to the left lung. The right PA is hypoplastic. The letters in the boxes at the lower right corner of each image indicate the direction being observed: 'L' indicates left and 'P' indicates posterior (E, F).

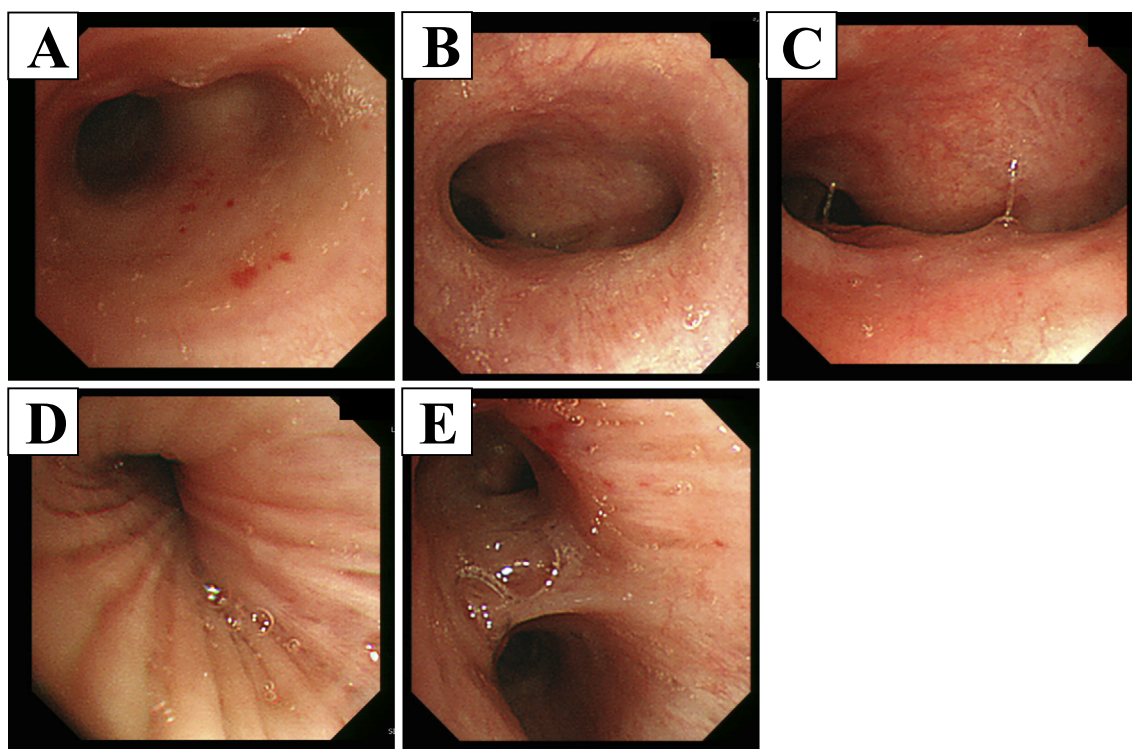


Figure 3. Bronchoscopy shows a diverticulum on the right wall of the distal trachea at a level where the orifice of the right main bronchus normally exists (A). The distal trachea collapses during expiration (B: inspiration, C: expiration); achondroplasia of the right main bronchus is noted (D). The left bronchi are normal (E).

tal capacity (81.4% of predicted) and forced expiratory volume in 1 second ($FEV_{1.0}$)/forced vital capacity (73.7%). The $FEV_{1.0}$ and peak expiratory flow rate were low (68.7% and 52.8% of the predicted values, respectively). Cardiac ultrasonography showed no abnormality. We suspected the possible causes of the cavities to be mycobacterial infection, fungal infection, or malignancy. However, no pathogens or malignant cells were identified in the obtained specimens. The biopsied tissue showed chronic inflammation of the bronchioles and alveolus. Therefore, instead of specific antibacterial agents, low-dose macrolide therapy was initiated, with the aim of facilitating bronchial clearance and anti-inflammation action.

Discussion

PA sling was first reported in 1897 by Glaevecke and Doehle (11). It is a rare vascular anomaly in which the left PA arises from the right PA. The abnormal formation of the left PA from the right sixth vascular arch (rather than the left) results in the aberrant left PA arising from the posterior aspect of the right PA. A strong embryologic association exists between PA sling, the development of tracheobronchial anomalies, and hypoplasia of the right lung (12, 13). In typical cases, the left main pulmonary artery arises from the right main pulmonary artery. However, as in the present case, an anomalous origin of the left inferior pulmonary artery from the right pulmonary artery is termed partial PA

sling (14). In most cases, the diagnosis is made during the infantile period. Typically, these patients present with severe respiratory distress because of airway stenosis due to the aberrant left PA or accompanying tracheobronchomalacia. Over 50% of cases of PA sling, including infantile cases, have associated tracheobronchial anomalies, such as tracheomalacia, stenosis, webs, or complete tracheal rings (4). Furthermore, congenital heart disorders are observed in approximately 10% to 15% of cases of PA sling (2). However, the present case did not have any associated heart disorders.

Presentation of PA sling in adults is rare (3-10). Unlike infantile cases, most adult cases are asymptomatic or mildly symptomatic (15). To our knowledge, there are only three documented adult cases of PA sling accompanied by tracheobronchial anomalies, similar to the present case (3, 4, 6). All patients were women who presented with symptoms of recurrent pneumonia or wheezing (Table). None of them had tracheobronchial anomalies as seen in the present case.

We speculate that, in the present case, bronchomalacia decreased the bronchial clearance and eventually contributed to the frequent pulmonary infections. Only three of the adult cases of PA sling reported in the previous literature required surgical treatment (4-6). A reported alternative approach for the treatment of this condition is stent therapy; however, this approach has been shown to be less effective than surgical treatment. As Table shows, surgery was performed in two cases, and improved symptoms were noted in both. For the

Table. Patients who have been Diagnosed with PA Sling Accompanied by Tracheobronchomalacia. Only Four Cases have been Reported to Date.

Age	Sex	Symptoms	Tracheobronchial anomalies	Treatment	Outcome	Reference
25	F	Wheezes, dyspnea	Tracheomalacia	Discontinuation asthma medications	Not worse	3
29	F	Wheezes, dyspnea	Tracheomalacia	Surgical resection of the vascular sling	Improvement	4
33	F	Repeated pneumonia	Bronchial stenosis	Pulmonary resection for the infected lesion	Improvement	6
49	F	Repeated pneumonia	Bronchial stenosis, Tracheomalacia	Low dose macrolide therapy	Not recurrence	Current case

present case, surgical treatment will be considered as an option if the patient presents with repeated pulmonary infections in the future.

Conclusion

We herein report a case of an adult patient with PA sling accompanied by tracheobronchomalacia. In patients who have repeated pulmonary infections at a relatively young age, the possibility of an airway abnormality should be considered. PA sling should be considered in the differential diagnosis of similar cases.

The authors state that they have no Conflict of Interest (COI).

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