CASE REPORT

A case of a pulmonary artery sling misdiagnosed as refractory asthma for 20 years

Toshihide Inui¹, Hideyasu Yamada¹23, Norihito Hida³, Hideo Terashima², Takefumi Saito⁴ & Nobuyuki Hizawa³

¹Department of Respiratory Medicine, Hitachinaka General Hospital, Hitachi, Ltd., Hitachinaka-shi, Ibaraki, Japan
²Hitachinaka Medical Education and Research Centre, University of Tsukuba Hospital, Hitachinaka-shi, Ibaraki, Japan
³Department of Pulmonology, Graduate School of Comprehensive Human Sciences, University of Tsukuba, Tsukuba, Ibaraki, Japan
⁴Department of Respiratory Medicine, National Hospital Organization, Ibarakihigashi National Hospital, Tokai-mura, Naga-gun, Ibaraki, Japan

Correspondence
Hideyasu Yamada, Hitachinaka Medical Education and Research Centre, University of Tsukuba Hospital, 20-1 Ishikawa-cho, Hitachinaka-shi, Ibaraki 312-0057, Japan.
Tel: +81-29-354-5111; Fax: +81-29-354-6842;
E-mail: h.yamada@md.tsukuba.ac.jp

Funding Information
No sources of funding were declared for this study.

Received: 21 December 2016; Revised: 2 March 2017; Accepted: 14 March 2017

Clinical Case Reports 2017; 5(6): 863–866
doi: 10.1002/ccr3.960

Key Clinical Message
We report the case of a 25-year-old woman with a pulmonary artery sling who was misdiagnosed as having childhood-onset refractory asthma for approximately 20 years. The use of computed tomography may be useful for diagnosing this rare condition.

Keywords
Fractional exhaled nitric oxide, pulmonary artery sling, refractory asthma, tracheomalacia

Introduction
Isolated anomalies of the branch pulmonary arteries are rare and usually occur in the setting of complex congenital heart disease[1, 2]. These isolated anomalies are often not identified in the prenatal period. A pulmonary artery sling (PAS) is created by the anomalous origin of the left pulmonary artery from the posterior aspect of the right pulmonary artery [3]. The anomalous left pulmonary artery courses over the right main stem bronchus and then from the right to the left, moving posterior to the trachea or carina and anterior to the esophagus, to finally reach the hilum of the left lung. A PAS is a rare condition and often coexists with tracheal stenosis [4]. Although the symptoms, which include respiratory distress manifested by stridor, recurrent pneumonia, wheezing, and cyanosis, typically occur within the first month of life, lack of clinical experience can lead to wrong diagnoses and poor outcomes [1]. Here, we report the case of a 25-year-old woman with a PAS who was misdiagnosed as having childhood-onset refractory asthma for approximately 20 years.

Case Report
A 25-year-old woman who had been receiving inhaled corticosteroid treatment (fluticasone 500 μg/day) for asthma for approximately 20 years was referred to our hospital because of persistent dyspnea despite intensive asthma treatment. She had been treated with intravenous and oral steroid therapy for asthma attacks a couple of times a year, especially during the winter season. Her symptoms included convulsive dyspnea and wheezing heard during dyspnea attacks. The symptoms did not respond to treatment, which included inhaled corticosteroid, a long-acting beta-2 agonist, and an inhaled
anticholinergic agent. She had never undergone testing for airway hyper-reactivity, nor did she have any family history of asthma or atopy.

The white blood cell count was 3500/μL (3500–9000); IgE, 106 IU/mL (≦173) (Table 1); and fraction of exhaled nitric oxide, 16 ppm (≦37). Spirometry showed a vital capacity of 3.07 L (99%), forced expiratory volume in 1 sec (FEV1.0) of 2.73 L (88.6%), and FEV1.0% of 91.9%. The flow volume curve was convex downward, which was consistent with a diagnosis of asthma (Fig. 1). The airway reversibility was 60 mL and 2.2%. The Empey index was 7.41 (<8).

A chest computed tomography scan revealed a right tracheobronchial anomaly, left main bronchial stenosis, and the left pulmonary artery originating from the right pulmonary artery and encircling the distal trachea and right main stem bronchus as it coursed between the trachea and esophagus to reach the hilum of the left lung (Fig. 2). On the basis of the computed tomography (CT) findings, we diagnosed PAS; we considered that the PAS was causing her symptoms, including the wheezing and convulsive dyspnea. Cardiac ultrasonography failed to find any cardiac malformations. Bronchoscopic examination identified a right tracheobronchial anomaly and left main bronchial stenosis (Fig. 3).

Figure 1. The flow volume curve was convex downward. Obstructive ventilatory impairment was not found.

Table 1. The blood test results showed no appreciable abnormalities.

| Test   | Value   |
|--------|---------|
| WBC    | 3500/μL |
| RBC    | 387/μL  |
| Hb     | 10.0 g/dL |
| Ht     | 31.3%   |
| Pht    | 22.3 104/μL |
| Neu    | 62.1%   |
| Eos    | 2.0%    |
| Bas    | 0.6%    |
| Mon    | 6.0%    |
| Lym    | 29.3%   |
| TP     | 6.4 g/dL |
| Alb    | 4.1 g/dL |
| AST    | 12 U/L  |
| ALT    | 9 U/L   |
| LDH    | 116 U/L |
| γ-GTP  | 9 U/L   |
| T-Bil  | 0.5 mg/dL |
| AMY    | 52 U/L  |
| BUN    | 9.7 mg/dL |
| Cre    | 0.6 mg/dL |
| Na     | 140 mEq/L |
| K      | 3.8 mEq/L |
| Cl     | 109 mEq/L |
| CRP    | 0.04 mg/dL |
| TG     | 83 mg/dL |
| T-cho  | 174 mg/dL |
| IgE    | 106 IU/mL |
| BS     | 230 mg/dL |
| Hbs ag (-) |
| HCV ab (-) |
| RPR (-) |
| TP (-)  |

The white blood cell count was 3500/μL (3500–9000); IgE, 106 IU/mL (≦173) (Table 1); and fraction of exhaled nitric oxide, 16 ppm (≦37). Spirometry showed a vital capacity of 3.07 L (99%), forced expiratory volume in 1 sec (FEV1.0) of 2.73 L (88.6%), and FEV1.0% of 91.9%. The flow volume curve was convex downward, which was consistent with a diagnosis of asthma (Fig. 1). The airway reversibility was 60 mL and 2.2%. The Empey index was 7.41 (<8).

A chest computed tomography scan revealed a right tracheobronchial anomaly, left main bronchial stenosis, and the left pulmonary artery originating from the right pulmonary artery and encircling the distal trachea and right main stem bronchus as it coursed between the trachea and esophagus to reach the hilum of the left lung (Fig. 2).

On the basis of the computed tomography (CT) findings, we diagnosed PAS; we considered that the PAS was causing her symptoms, including the wheezing and convulsive dyspnea. Cardiac ultrasonography failed to find any cardiac malformations. Bronchoscopic examination identified a right tracheobronchial anomaly and left main bronchial stenosis (Fig. 3).
No abnormality was observed in the mucosa although the bronchoscopy did not proceed beyond the left secondary carina because of the stenosis. Her exercise capacity was as follows: (1) peak rate of oxygen consumption, 28.2 mL/min/kg (110% of normal); (2) minute ventilation–carbon dioxide production relationship, 40.5 (>34; slightly enhanced ventilation); (3) minute ventilation (L/min), 8.2 at rest and 61.8 at peak level, which was considered normal; and (4) dyspnea index (i.e., the ratio of minute ventilation at peak exercise to the maximal voluntary ventilation), 0.65, which indicated no movement limitation caused by ventilatory impairment. On the basis of these findings, we considered her condition to be not so severe as to require surgery. We discontinued all her asthma medications, and, thereafter, her symptoms did not worsen.

**Discussion**

We have here reported the case of a 25-year-old woman who was misdiagnosed as having long-standing, mild, persistent asthma characterized by dyspnea. She was initially treated with bronchodilators and inhaled corticosteroids, but without improvement. She underwent further evaluation with chest computed tomography and flexible bronchoscopy, which revealed focal tracheomalacia (TM) in the distal trachea secondary to chronic extrinsic compression due to a PAS. PAS is an under-recognized condition that presents with nonspecific symptoms such as dyspnea, cough, and recurrent infections. PAS usually presents within the first weeks to months of life [5, 6], and the discovery of PAS in adulthood is rare; only a few cases have been previously documented [7–12] (Table 2).

Diagnosis was delayed because there was no merger with congenital heart defects. To the best of our knowledge, this is the first reported case of PAS case presenting with asthma-like symptoms and treated as asthma over a long period. However, given that patients with this condition are potentially misdiagnosed as having more prevalent diseases such as asthma and chronic obstructive pulmonary disease, the differential diagnosis of refractory asthma should include focal TM in the distal trachea secondary to chronic extrinsic compression due to a PAS. The use of computed tomography may be useful for diagnosing this rare condition [13].

**Acknowledgments**

We thank Ms F. Miyamasu, Medical English Communications Center, University of Tsukuba, for proofreading the article.

**Conflict of Interest**

The authors have no conflict of interest to declare.

**Authorship**

TI, HY, and NH: involved in writing this article. NH, HT, and TS: gave advice on this article.

**References**

1. Tetter, J. T., E. M. Tetter, D. Y. Rafii, R. H. Anderson, and P. Bhatla. 2016. Fetal diagnosis of
abnormal origin of the left pulmonary artery.
Echocardiography 33:1258–1261.

2. Yu, J.-M., C.-P. Liao, S. Ge, Z.-C. Weng, M.-C. Hsiung, J.-K. Chang, et al. 2008. The prevalence and clinical impact of pulmonary artery sling on school-aged children: a large-scale screening study. Pediatr. Pulmonol. 43:656–661.

3. Hraska, V., J. Photiadis, C. Haun, E. Schindler, M. Schneider, P. Murin, et al. 2009. Pulmonary artery sling with tracheal stenosis. Multimed. Man. Cardiothorac. Surg. 2009:123. mmcts.2008.003343.

4. Chassagnon, G., B. Morel, E. Carpentier, H. Ducou Le Pointe, and D. Sirinelli. 2016. Tracheobronchial branching abnormalities: lobe-based classification scheme. Radiographics 36:358–373.

5. Zhan, Y., W. Ren, Y. Xiao, G. Song, and Q. Hu. 2013. Pulmonary arterial sling compressing the trachea presenting with recurrent stridor in an infant. J. Am. Coll. Cardiol. 62:1124.

6. Delacour, D., M. Demeyere, B. Dubourg, and J. N. Dacher 2017. Left pulmonary artery sling: a rare cause of congenital stridor. Diagn. Interv. Imaging. 98(1):85–87. doi:10.1016/j.diii.2016.06.012.

7. Procacci, C., E. Residori, M. Bertocco, P. Di Benedetto, I. A. Andreis, and N. D’Attoma 1993. Left pulmonary artery sling in the adult: case report and review of

the literature. Cardiovasc. Intervent. Radiol. 16: 388–391.

8. Espinosa, L., and P. Agarwal. 2008. Adult presentation of right lung agenesis and left pulmonary artery sling. Acta Radiol. 49:41–44.

9. Repanas, G., M. Papadopoulos, K. Anastasiadou, S. Galanis, G. Tzanaktsidis, and P. Palladas. 2009. Pulmonary artery sling. [Online]. URL: http://www.eurorad.org/case.php?id=7733; https://doi.org/DOI: 10.1594/EURORAD/CASE.7733

10. Komoto, K., S. Ha-Kawa, N. Tanigawa, and T. Kinoshita 2013. Two adult cases of pulmonary artery sling. Intern. Med. 52:1271–1272.

11. Kurian Maliel, M. D., M. Javed, M. D. Nasir, and D. O. Kevin Steel 2013. Pulmonary artery sling initially presenting with symptoms during exercise. J. Am. Osteopath. Coll. Radiol. 2:24–26.

12. Miyazaki, T., N. Yamasaki, T. Tsuchiya, K. Matsumoto, H. Hayashi, and K. Izumikawa 2015. Partial lung resection of supernumerary tracheal bronchus combined with pulmonary artery sling in an adult: report of a case. Gen. Thorac. Cardiovas. Surg. 63:173–176.

13. Lee, K. H., C. S. Yoon, K. O. Choe, et al. 2001. Use of imaging for assessing anatomical relationships of tracheobronchial anomalies associated with left pulmonary artery sling. Pediatr. Radiol. 31:269–278.