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CASE REPORT

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

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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

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

WBC	3500	/μL
RBC	387	/μL
Hb	10.0	g/dL
Ht	31.3	%
Plt	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 (-)		

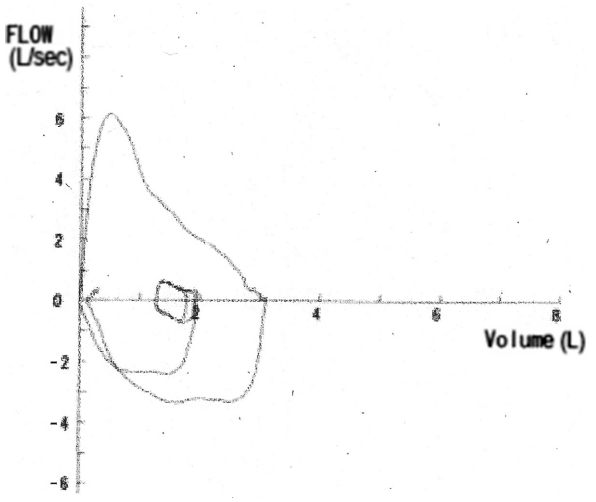

Figure 1. The flow volume curve was convex downward. Obstructive ventilatory impairment was not found.

anticholinergic agent. She had never undergone testing for airway hyper-reactivity, nor did she have any family history of asthma or atopy.

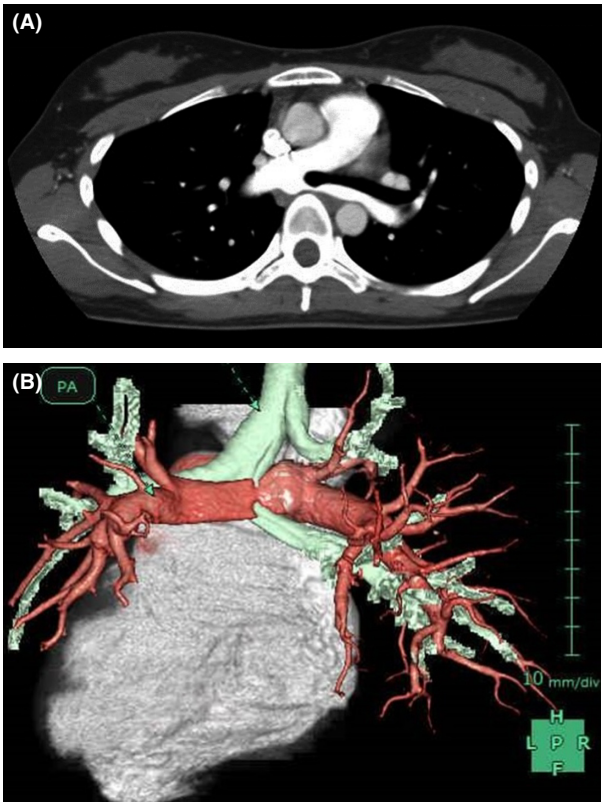

Figure 2. (A) Computed tomography (CT) image showing a pulmonary artery sling (PAS). The pressure exerted by the left pulmonary artery is the cause of left main bronchial stenosis. (B) A 3D-reconstructed CT image of the left PAS viewed from the posterior aspect.

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).

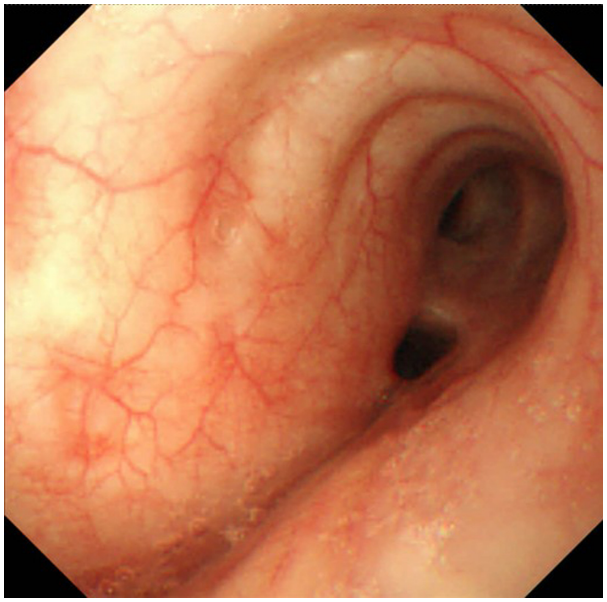

Figure 3. Bronchoscopy image showing left main bronchial stenosis caused by the pulmonary artery sling.

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.

Table 2. Previous pulmonary artery sling (PAS) cases diagnosed in adulthood.

Age	Sex	Year reported	Symptoms	Initial diagnosis	Reference
68	M	1993	No	Lung cancer	7
49	F	2008	Dyspnea	Absent right lung	8
62	F	2009	Traffic accident	PAS	9
42	M	2013	Pharyngeal discomfort	PAS	10
36	F	2013	Wheeze, bloody sputum	PAS	10
22	M	2013	Palpitations	PAS	11
33	F	2015	Repeated infection	Chronic bronchitis	12
25	F	2017	Wheeze, dyspnea	Asthma	Current case

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 underrecognized 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].

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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.

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