

Left lung hypoplasia and arrhythmia: a rare case

Hipoplazja lewego płuca i niemiary – rzadki przypadek

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Abstract

Pulmonary hypoplasia is a rare congenital disorder. Respiratory and circulatory systems sustain a closely correlated function, and if the function of one is impaired, the other will inescapably be affected. We presented a unilateral lung hypoplasia case that manifested a cardiological disorder (ventricular arrhythmia) rather than a respiratory-related symptom. We highly recommend that such patients should be followed-up by a cardiologist beside a pulmonologist.

Key words: lung hypoplasia, arrhythmia, echocardiography, congenital lung disorder

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Introduction

Pulmonary hypoplasia, which is the result of insult to the embryo during the 4th to 5th week of intrauterine life, is a rare congenital disorder. Incidence of this entity ranges from 1–2/10,000 live births [1]. Both genders are affected almost equally. Usually, it is unilateral and is characterized by a decrease in the number or size of the airways, vessels, and alveoli resulting in a small fibrotic and nonfunctioning lung. Also, pulmonary artery and vein atresia could accompany, sometimes [2]. Computed tomography (CT) is a favourable tool for diagnosis [3].

Respiratory and circulatory systems sustain a closely correlated function. If one develops a pathology, the other will inevitably be affected. Here we describe a unilateral lung hypoplasia case that showed ventricular arrhythmia.

Case presentation

A 49-year-old female patient, who had no known history of chronic disease, presented to the cardiology

outpatient clinic with an occasional palpitation. Her initial vital findings were as follows; blood pressure, 122/76 mm Hg; heart rate, 85/minute; body temperature, 36.5 °C; saturation, 98%. On physical examination, left-sided lung did not attend to inspiration, auscultation sounds of his left lung could not have been detected, and obvious left-sided dullness was located on percussion. 12-lead electrocardiogram (ECG) showed sinus rhythm and two ventricular extra beats with a rate of 85 beats/minute (Figure 1). According to laboratory parameters, biochemistry, haematology and thyroid function tests were within the normal range but total cholesterol was 250 mg/dL (reference range < 200 mg/dL). Transthoracic echocardiography (TTE) revealed dilatation in right heart chambers (right ventricle; 4.6 cm, right atrium; 4.5 cm), a thin and aneurysmatic interatrial septum, elevated pulmonary arterial systolic pressure (40 mm Hg), and that the anatomical position of the heart was shifted to the left (Figure 2A, B, C). A contrast-enhanced thorax CT angiography was performed to reveal detailed anatomy and clarify the diagnosis. The CT demonstrated

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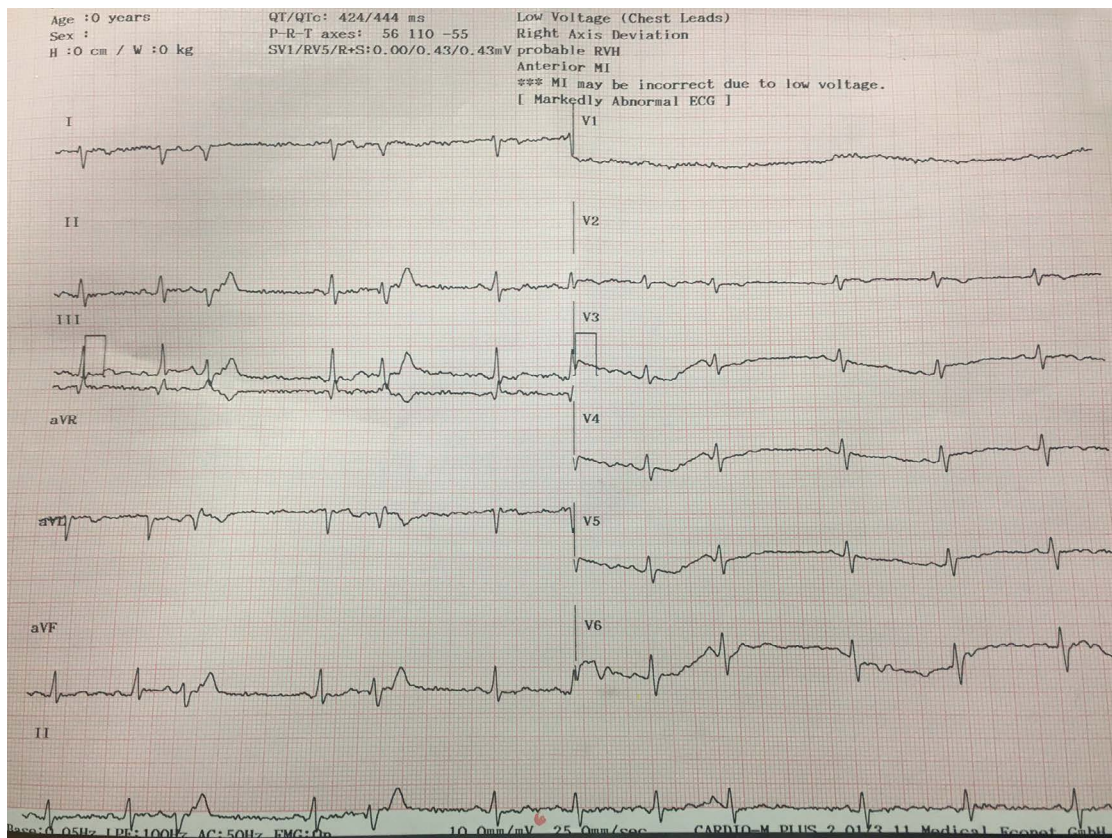


Figure 1. Electrocardiogram: Sinus rhythm, a rate of 85 beats/min and ventricular extra beats

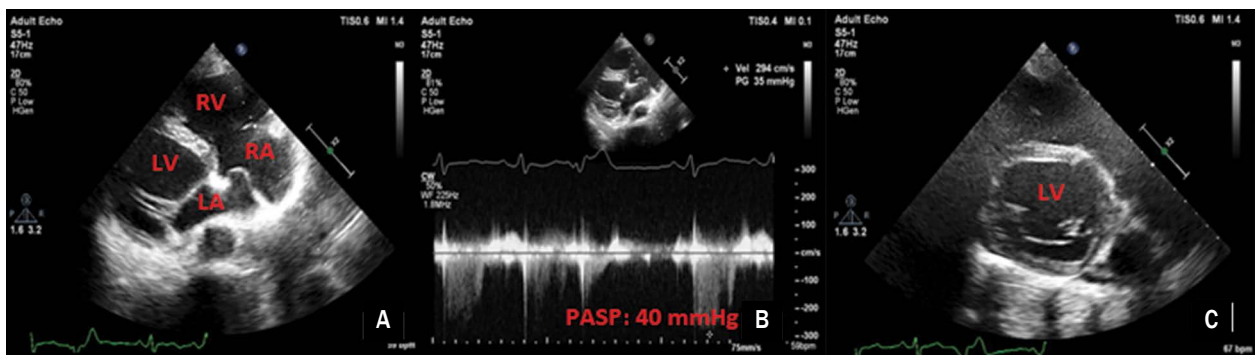


Figure 2. Transthoracic echocardiography; parasternal long axis: **A**. Right heart chambers were dilated. The interatrial septum was thin and aneurysmatic; **B**. Pulmonary arterial systolic pressure (PASP): 40 mm Hg; **C**. The left pulmonary vein and arteries could not have been seen; LA – left atrium; LV – left ventricle; RA – right atrium; RV – right ventricle

a hypoplastic left lung and a shifted mediastinum to the left side. Additionally, hypertrophy and emphysema were observed in the right lung, and the right lung upper lobe extended to the left lung side (Figure 3A, B, C). A 24-hour ambulatory rhythm Holter showed a total of 15,000 extra ventricular beats. A 50 mg metoprolol pill once a day was started and the patient was referred to the chest disease clinic for follow-up.

Discussion

Lung malformations, which are the results of insult to the embryo during the 4th to 5th week of intrauterine life, are very rare to occur with varying degrees of severity and with an incidence as low as 1–2/10,000–12,000 births [2, 3]. Boyden categorized them as pulmonary agenesis, aplasia, and hypoplasia[4]. Hypoplasia, which

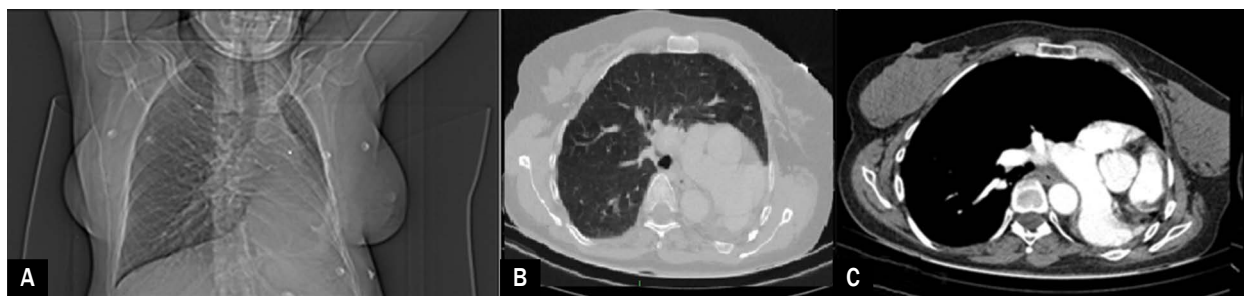


Figure 3. Contrast-enhanced thorax computed tomography: **A.** The patient's left lung was hypoplastic, mediastinum shifted to the left; **B.** Hypertrophy findings and areas of emphysema were observed in the right lung; **C.** Upper lobe of right lung extended and located to the upper part of the left-sided lung space

is the subject of the current article, generally appear to be unilateral [5]. Our case showed left-sided lung hypoplasia.

Although the certain cause is not entirely understood, deficiency of some elements, such as thyroid transcription factor-1, hepatocyte nuclear factors, epidermal growth factor, and its mitogen-activated protein kinase receptor, are presumed hypotheses. Besides that small fetal thoracic volume, prolonged oligohydramnios, early rupture of membranes, congenital heart diseases, and trisomies 13, 18, and 21 may play role in the development of this entity [6, 7].

Considering the literature, a small number of left lung hypoplasia cases have been reported, so far. Here, we present a left lung hypoplasia patient who developed cardiac arrhythmia. It is the first case in the literature that presented to the cardiology clinic with an occasional palpitation rather than pulmonary insufficiency symptoms and that diagnosed incidentally by a cardiologist.

Usually, pulmonary hypoplasia cases presented with recurrent chest infections or symptoms of cardiopulmonary insufficiency [4]. In our case, dissimilarly, there was no respiratory complaint. ECG showed two and 24-hour ambulatory rhythm Holter showed a significant number of ventricular extra beats. In this case, it is a question point whether the ventricular extra beats were a result of lung hypoplasia or completely coincidental. Even though it is hard to link arrhythmia to lung malformation, we presume it was due to elevated pulmonary artery systolic pressure and right ventricular enlargement which was a result of left lung hypoplasia. After all, it should be kept in mind that such patients could have a cardiological complaint and disorder. These patients should be followed up by a cardiologist besides a pulmonologist to prevent future possible cardiological complications.

To associate arrhythmia with lung malformations further studies and similar case examples are needed in this field.

Limitation

Due to the transposition of the heart, it was hard to reveal appropriate echocardiographical views, especially the apical four-chamber view.

Conclusion

Lung hypoplasia cases could manifest with a cardiological sign. Respiratory and circulatory systems sustain a closely correlated function, and if the function of one is impaired, the other will inescapably be affected. Therefore, such patients should be followed-up by a cardiologist beside a pulmonologist.

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Contributions

Elevation of the patient in the cardiology clinic, MK; preparation of the manuscript, TO.

Conflict of interest

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Streszczenie

Hipoplazja płuca jest wadą wrodzoną. Układy oddechowy i krwionośny pełnią ściśle skorelowane funkcje; jeżeli funkcjonowanie jednego z tych układów jest zaburzone, to ma to wpływ na drugi układ. W pracy przedstawiono przypadek jednostronnej hipoplazji płuca, która przejawia się zaburzeniami kardiologicznymi (arytmia komorowa), a nie objawami związanymi z układem oddechowym. Zalecamy, aby tacy pacjenci byli monitorowani zarówno przez kardiologa, jak i pulmonologa.

Słowa kluczowe: hipoplazja płuca, niemiarywość, echokardiografia, wrodzona wada płuc

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