







# Levoatriocardinal Vein Combined with Pulmonary Venous Varix Mimicking Arteriovenous Malformations: A Case Report

동정맥기형으로 오인되었던 폐정맥정맥류를 동반한  
Levoatriocardinal 정맥: 증례 보고

Joo Hee Jeun, MD<sup>1</sup> , Eun-Ju Kang, MD<sup>1\*</sup> ,  
Jeong-Hyun Jo, MD<sup>2</sup> , Ki-Nam Lee, MD<sup>1</sup> 

<sup>1</sup>Department of Radiology, Dong-A University Medical Center, Dong-A University College of Medicine, Busan, Korea

<sup>2</sup>Department of Radiology, Daedong Hospital, Busan, Korea

The levoatriocardinal vein is an uncommon pulmonary venous abnormality that connects the left atrium or pulmonary vein with the systemic vein. It is distinct from partial anomalous pulmonary venous return in that the former forms a connection with the left atrium through the normal pulmonary vein whereas the latter involves pulmonary venous drainage to the systemic vein. Herein, we describe a case of the levoatriocardinal vein initially misdiagnosed as a pulmonary arteriovenous malformation using chest radiography and chest CT. The levoatriocardinal vein combined with pulmonary venous varix was confirmed using pulmonary angiography. To the best of our knowledge, this unusual coexistence of the levoatriocardinal vein and pulmonary venous varix has not been reported in English literature.

**Index terms** Pulmonary Vein; Anomalous Pulmonary Venous Return; Varix

## INTRODUCTION

Levoatriocardinal vein refers to a connection between the left atrium (LA) or the pulmonary vein and the systemic vein (1-4), and it is a rare pulmonary anomaly. The vein is usually accompanied by obstructive left heart lesions such as mitral or aortic atresia or stenosis (1). Levoatriocardinal vein can be confused with partial anomalous pulmonary venous return (PAPVR), which occurs when anomalous veins drain into a systemic

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\*Corresponding author

Eun-Ju Kang, MD  
Department of Radiology,  
Dong-A University Medical Center,  
Dong-A University  
College of Medicine,  
26 Daesingongwon-ro, Seo-gu,  
Busan 49201, Korea.

Tel 82-51-240-5367

Fax 82-51-253-4931


E-mail medcarot@dau.ac.kr


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### ORCID iDs

Joo Hee Jeun 

[https://  
orcid.org/0000-0002-4012-8338](https://orcid.org/0000-0002-4012-8338)

Eun-Ju Kang   
[https://  
orcid.org/0000-0003-0937-3607](https://orcid.org/0000-0003-0937-3607)

Jeong-Hyun Jo   
[https://  
orcid.org/0000-0001-6964-5032](https://orcid.org/0000-0001-6964-5032)

Ki-Nam Lee   
[https://  
orcid.org/0000-0003-0848-3935](https://orcid.org/0000-0003-0848-3935)

vein. The difference between the two conditions lies in the connection with the LA, into which the normal pulmonary vein of the levoatriocardinal vein drains directly (1).

Herein we present a case of pulmonary venous varix (PVV) connected to the left innominate vein and also connected to the LA in the other direction, without a normal pulmonary vein. We describe this as a case of levoatriocardinal vein combined with PVV, even though there is no normal pulmonary vein between the systemic vein and LA. Chest radiography and CT are presented, as is angiography which depicted two sites of drainage into the left innominate vein and LA.

## CASE REPORT

A 77-year-old woman visited the hospital for evaluation of abnormal chest radiography results. She was a non-smoker and did not exhibit any respiratory symptoms. Her medical history included an aortic valve replacement performed 7 years prior due to bicuspid aortic valve and aortic stenosis, and a transient ischemia attack (TIA) that had occurred 6 years ago.

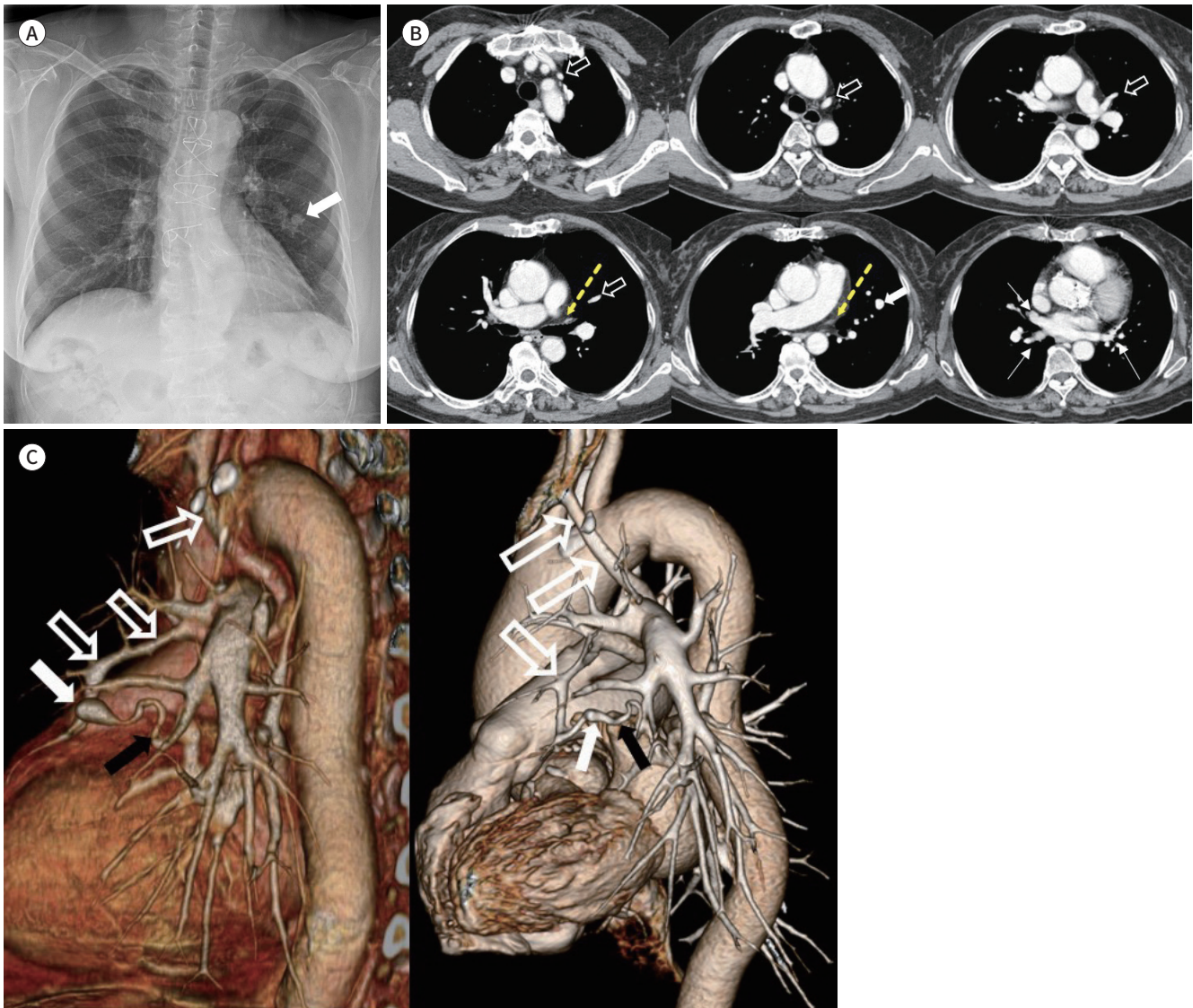
Chest radiography depicted a well-defined nodular opacity approximately 1 cm in size on the left middle lung field (Fig. 1A). Contrast-enhanced chest CT depicted a corresponding focally dilated vascular structure in the left lingular segment (Fig. 1B). The vascular structure exhibited drainage to the left innominate vein, and it was considered as PAPVR in the first place. The course from the dilated vasculature to the left innominate vein indicated that it entered the mediastinum at the left pulmonary artery and coursed upward, crossing the aortic arch inferiorly. Chest CT also revealed the absence of a normal left upper pulmonary venous structure, which is typically situated between the left side of the pulmonary trunk and the left main bronchus (Fig. 1B). Volume-rendered CT revealed that the dilated portion contained another connected vascular structure other than the vasculature drained to left innominate vein (left image in Fig. 1C). It was not clear, however, into which vessel this structure drained.

In a retrospective review of previous imaging, volume-rendered CT performed before the aortic valve replacement operation 7 years ago exhibited a smaller dilated vasculature (right image in Fig. 1C). It also depicted bidirectional drainage compatible with PAPVR and the other drainage flow was still obscured. Considering the incidence rate of pulmonary congenital anomaly, we interpreted the focally dilated vasculature as a connection between the pulmonary artery and vein, defined as a pulmonary arteriovenous malformation. In addition, we consulted it to an interventional radiologist for pulmonary angiography and embolization.

In the arterial phase, pulmonary angiography via femoral artery puncture depicted normal pulmonary arteries with no delineation of arteriovenous malformation (left image in Fig. 1D). In the venous phase, however, angiography revealed opacification of a round-shaped lesion corresponding with the dilated vessel visible on chest CT. And it also showed a solitary drainage to the LA through left lower pulmonary vein (middle image in Fig. 1D). Selective segmental pulmonary arteriography depicted two sites of drainage, into the left innominate vein and to the serpiginous vein, which was directed to the left lower pulmonary vein and subsequently to the LA (right image in Fig. 1D). Because this connection between the LA and the systemic vein is a defining characteristic of levoatriocardinal vein rather than PAPVR, the pa-

tient's condition could be diagnosed as levoatriocardinal vein combined with PVV. An additional skin puncture via the left basilic vein was conducted to approach the left innominate vein. Venography with the catheter tip near the left innominate vein did not indicate any flow toward the dilated portion, meaning that this portion was the source of the drainage flow. Approaching the focally dilated portion the flow receded, confirming bidirectional flow (left image in Fig. 1E). Embolization for the dilated vessel was recommended due to the pa-

**Fig. 1.** Levoatriocardinal vein combined with pulmonary venous varix in a 77-year-old woman, presenting with chest abnormalities on screening. **A.** Chest radiography depicts a well-defined, round increased opacity focus in the left middle lung field (arrow). **B.** Contrast-enhanced CT images depict focally dilated vasculature in the left lingular segment corresponding to the round increased opacity focus on chest radiography (white arrow), with a drainage course to the left innominate vein (blank arrows). There is no normal left upper pulmonary vein between the pulmonary trunk and the left main bronchus (yellow dotted arrows), and the normal right pulmonary veins and left lower pulmonary vein (white thin arrows) are evident. **C.** Three-dimensional volume-rendered CT image in the posterolateral view depicts a dilated sac (white arrow, left image) and smaller sac in the lateral view acquired 7 years before (white arrow, right image). Further, a drainage pathway into the left innominate vein (blank arrows) is depicted. Another tortuous draining vasculature is visible (black arrows), but its drainage course is unclear in both images.

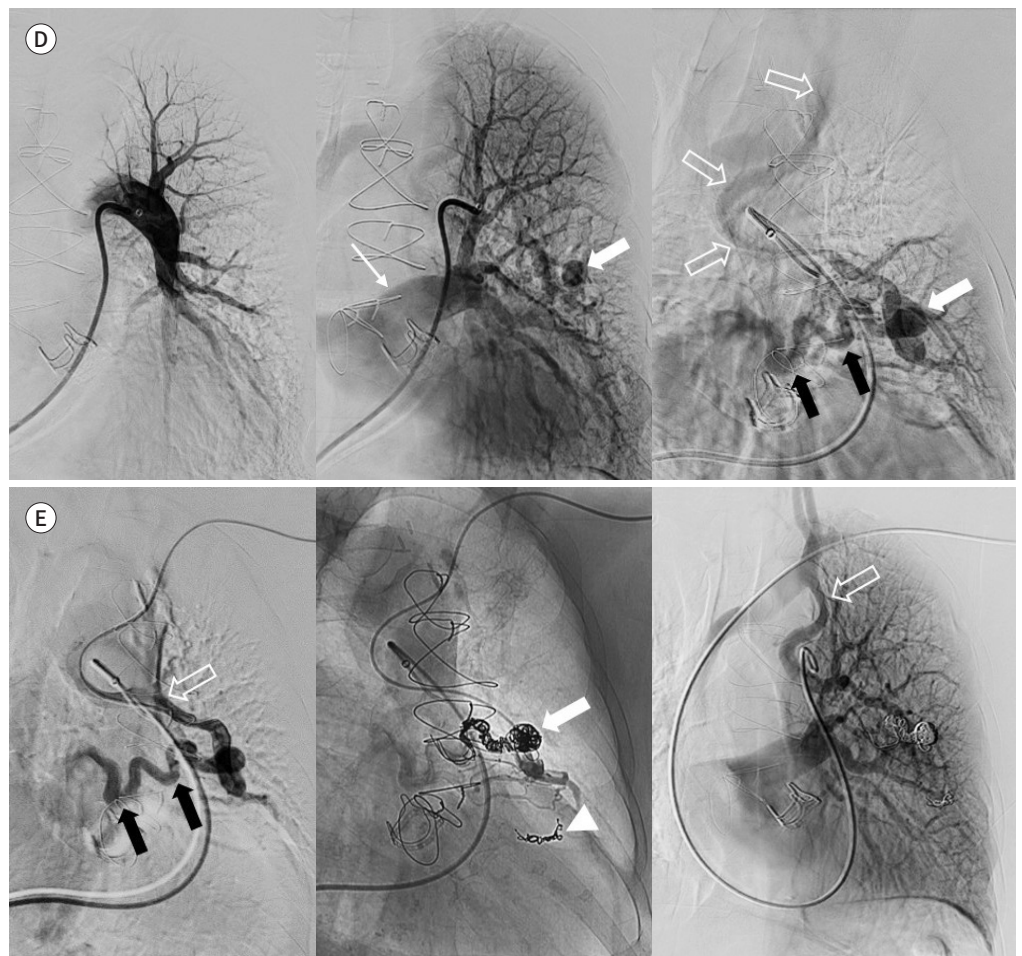




**Fig. 1.** Levoatriocardinal vein combined with pulmonary venous varix in a 77-year-old woman, presenting with chest abnormalities on screening.

**D.** Left pulmonary angiography via a femoral vein puncture in the arterial phase does not depict any evidence of vascular anomalies (left image). Delayed left pulmonary angiography (middle image) demonstrates the convergence of the left pulmonary veins into the left lower pulmonary vein (white thin arrow) but no evidence of the left upper pulmonary vein. Note a round abnormal vascular opacification focus (white arrows) in the left lingular segment. Selective angiography of the segmental pulmonary artery (right image) depicts two sites of drainage into the left innominate vein (black arrows), left lower pulmonary vein (blank arrows), and finally to the left atrium.

**E.** Venography via a left basilic vein puncture also depicts bidirectional flow (blank arrow and black arrows) of the venous varix (left image). Embolization was performed at the venous varix (white arrow) with microcoils (middle image), and another embolization with a microcoil was performed for an additional small connection (arrowhead). Left pulmonary angiography 3 days later via the left basilic vein puncture (right image) depicts systemic flow to the left innominate vein (blank arrow) and no flow to the left atrium, indicating adequate disconnection of flow.



patient's history of cerebral infarction, which meant that the structure had the potential to be the source of an embolism from systemic venous flow to the left heart, and also the size of the varix had increased substantially during the past 7 years. Coil embolization was performed to treat the PVV using microcoils (6 mm/14 cm  $\times$  1, 4 mm/14 cm  $\times$  3, Nester; Cook Medical, Bloomington, IN, USA and 8 mm/30 cm  $\times$  1, 6 mm/20 cm  $\times$  1, 5 mm/20 cm  $\times$  1, Concerto; Medtronic, Minneapolis, MN, USA). After embolization of the PVV a smaller con-

nection between the left innominate vein and the LA was observed, and an additional embolization was performed using a microcoil (4 mm/10 cm × 1, Tornado; Cook Medical) (middle image in Fig. 1E). Pulmonary arteriography 3 days after the procedure depicted venous flow of the left innominate vein without any flow to the LA, confirming embolization of the PVV (right image in Fig. 1E). Angiography was used to diagnose the patient with coexisting levoatriocardinal vein and PVV, because there was connection between the systemic vein and LA, leaving the absence of a normal pulmonary venous in between. The patient was treated with endovascular coil embolization. Follow-up chest radiography has indicated no further changes in embolization coils and no other notable symptoms for a period of 1 year following the procedure.

## DISCUSSION

Levoatriocardinal vein is defined as a connection between the LA and a systemic vein (left image in Fig. 1F). The developing lungs are drained to the cardinal vein for the first 2 months, and the capillary plexus near the LA subsequently becomes the pulmonary vein (1, 2). The pulmonary vein connects to the LA while obliterating the normal connection to the cardinal vein. If the primitive connection with the cardinal vein is a consistent alternative pathway in the case of obstructive left heart lesions, the condition becomes levoatriocardinal vein with the LA and systemic vein connection derived from the cardinal vein (1-3). The levoatriocardinal vein most commonly drains to the left innominate vein and courses dorsally to the left pulmonary artery and laterally to the aortic arch (1, 4).

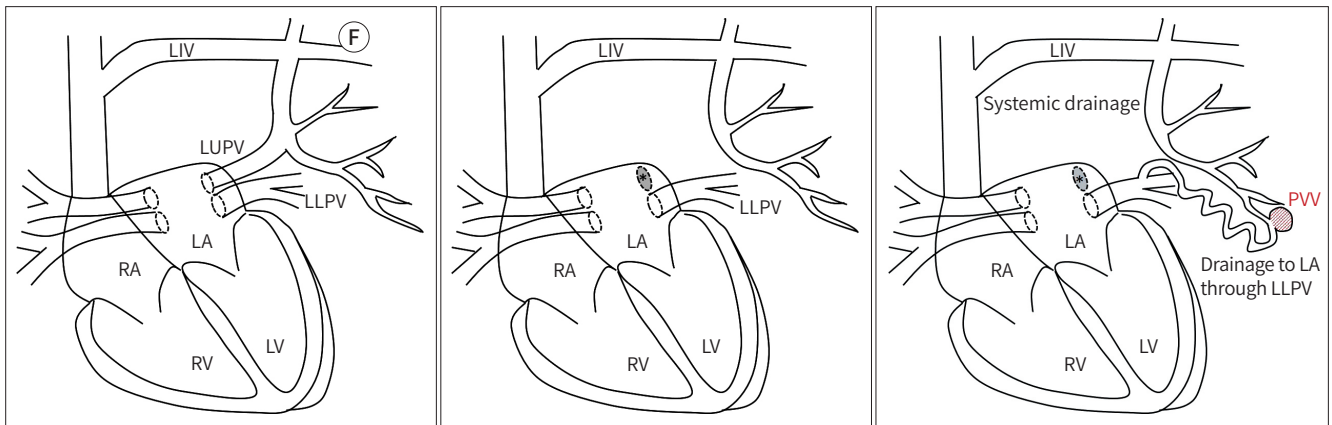
The other vascular anomaly that can be confused with levoatriocardinal vein is PAPVR. PAPVR exhibits anomalous pulmonary vein drainage to a systemic vein (middle image in Fig. 1F) and is only detected in 0.4–0.7% of autopsies (5). If the vein is located in the left lung the most common drainage site is the left innominate vein or the coronary sinus (2). The primary difference between levoatriocardinal vein and PAPVR is that the levoatriocardinal vein is connected with the normal pulmonary vein (1).

The current patient was initially diagnosed with PAPVR because there was clear drainage to the left innominate vein, whereas drainage to the LA was indeterminate in conventional chest CT. Subsequent angiography revealed another connection to the LA through serpiginous vasculature connected to the left lower pulmonary vein in the absence of a normal left upper pulmonary vein (right image in Fig. 1F). Because there was a connection to the LA through PVV, the term levoatriocardinal vein was more accurate to describe the condition. We ultimately diagnosed the condition as PVV of levoatriocardinal vein.

It is known that levoatriocardinal vein usually drains toward a cephalad course, and less commonly drains bidirectionally (1). In the present case the vein drained bidirectionally to both the LA and the innominate vein. Further, the focally dilated portion of the venous structure in the present case was between these two flows. That structure was also depicted as an increased oval-shaped opacity on chest radiography mimicking arteriovenous malformation, and it was determined to be venous dilatation via angiography. Focal dilatation of the pulmonary vein is a defining characteristic of PVV, and usually occurs near the entrance to the LA (3). To our knowledge there have been no reports of levoatriocardinal vein combined with

**Fig. 1.** A 77-year-old woman with abnormalities on screening chest radiography.

**F.** Schematic illustrations demonstrating pulmonary venous anomalies. In the levoatriocardinal vein, there is an abnormal connection between the LA or a pulmonary vein and the systemic vein (left image). It has a normal pulmonary vein connected to the LA. In partial anomalous pulmonary venous return, a pulmonary vein drains into the systemic vein without normal pulmonary venous connection with the LA (\*, middle image). This illustration shows drainage to the LIV. The right image shows a connection between the systemic vein and the LA of the present case. Unlike the typical levoatriocardinal vein, it is connected to the LA not through a normal pulmonary vein but through abnormal tortuous drainage vasculature directed to the LLPV. Further, it is also combined with PVV. Note the absence of the normal LUPV (\*, right image). LA = left atrium, LIV = left innominate vein, LLPV = left lower pulmonary vein, LUPV = left upper pulmonary vein, LV = left ventricle, PVV = pulmonary venous varix, RA = right atrium, RV = right ventricle



venous varix formation.

PVV is a rare condition involving anomalous pulmonary vessels, and there are two types, primary and secondary (6, 7). The secondary type is more common, and cardiovascular conditions such as mitral valve disease or another chronic heart disease are common causes (8). The underlying chronic bicuspid aortic valve and aortic stenosis in the current case may have caused the varix formation. The characteristic features of the present case such as the bidirectional flow and the unusual drainage passage to the LA, and consequent limitation of drainage flow may have been another reason for the formation of PVV and tortuous vasculature. Varix formation is generally benign but in rare cases there are complications such as rupture and systemic emboli due to thrombosis within the varix (5). In addition, a levoatriocardinal vein in the absence of hypoplastic left heart typically does not require management (1). In the current case however the growing entity and the patient's previous history of TIA suggested a high risk of complications, so treatment with embolization was performed.

In summary, we have described a rare case of levoatriocardinal vein combined with PVV. The rare vasculature course involved was identified via angiography. A vascular anomaly was considered given the observation of a nodular opacity on a chest X-ray. Regular follow-ups to assess changes in shape or size should be conducted in the treatment of patients with the above-described condition and history.

#### Author Contributions

Conceptualization, all authors; investigation, all authors; supervision, K.E.; visualization, J.J.H., K.E.; and writing—original draft, J.J.H.

### Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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## 동정맥기형으로 오인되었던 폐정맥정맥류를 동반한 Levoatriocardinal 정맥: 증례 보고

전주희<sup>1</sup> · 강은주<sup>1\*</sup> · 조정현<sup>2</sup> · 이기남<sup>1</sup>

Levoatriocardinal 정맥은 폐정맥 기형의 드문 형태로, 좌심방 혹은 폐정맥과 체정맥을 연결해 주는 구조이다. 이는 폐정맥이 체정맥으로 연결되는 기형을 뜻하는 부분 폐정맥 환류 이상과 구분되는데, Levoatriocardinal 정맥은 정상 폐정맥을 통한 좌심방과의 연결성이 있다는 차이점이 있다. 저자들은 폐정맥정맥류가 흉부 엑스선 및 전산화단층촬영에서 동정맥기형으로 오인되었던 증례를 보고하고자 하며, 이는 혈관조영술을 통해 폐정맥정맥류와 동반된 Levoatriocardinal 정맥으로 진단된 경우이다. 이러한 폐정맥정맥류를 동반한 Levoatriocardinal 정맥을 보이는 증례는 영문 문헌상 보고된 바 없기에 이를 보고하고자 한다.

<sup>1</sup>동아대학교 의과대학 동아대학교의료원 영상의학과,

<sup>2</sup>대동병원 영상의학과