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

동정맥기형으로 오인되었던 폐정맥정맥류를 동반한 Levoatriocardinal 정맥: 증례 보고

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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; Pulmonary Vein

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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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-
A patient'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.
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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 (blank arrows), left lower pulmonary vein (black 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.

The 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 × 1, 4 mm/14 cm × 3, Nester; Cook Medical, Bloomington, IN, USA and 8 mm/30 cm × 1, 6 mm/20 cm × 1, 5 mm/20 cm × 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 embo-
lization 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 levoat-
riocardinal 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 follow-
ing 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 levoatriocardi-
nal 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 pri-
mary 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 serpigi-
nous 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 struc-
ture 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 pulmo-
nary 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
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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.

REFERENCES
1. Agarwal PP, Mahani MG, Lu JC, Dorfman AL. Levoatriocardinal vein and mimics: spectrum of imaging findings. AJR Am J Roentgenol 2015;205:162-171
2. Dillman JR, Yarram SG, Hernandez RJ. Imaging of pulmonary venous developmental anomalies. AJR Am J Roentgenol 2009;192:1272-1285
3. Porres DV, Morenza OP, Pallisa E, Roque A, Andreu J, Martínez M. Learning from the pulmonary veins. Radiographics 2013;33:999-1022
4. Hyun D, Chae EJ, Seo JB, Kang JW, Do KH, Lee CW, et al. Partial anomalous pulmonary venous return via a levoatriocardinal vein in association with rheumatic mitral stenosis: MR demonstration and successful surgical repair. J Korean Soc Radiol 2010;63:339-343
5. Maillard JO, Cottin V, Etienne-Mastroianni B, Frolet JM, Revel D, Cordier JF. Pulmonary varix mimicking pulmonary arteriovenous malformation in a patient with Turner syndrome. Respiration 2007;74:110-113
6. Hochhegger B, Zanetti G, Marchiori E. Pulmonary venous varix presenting as a pulmonary nodule. Ann Thorac Surg 2017;103:e459
7. Hanson JM, Wood AM, Seymour R, Petheram IS. Anomalous unilateral single pulmonary vein: two cases mimicking arteriovenous malformations and a review of the literature. Australas Radiol 2005;49:246-251
8. Kumazoe H, Komori M, Ochiai R, Egashira R, Nakazono T, Kudo S. Pulmonary varix mimicking arteriovenous malformation. Clin Imaging 2008;32:61-64