Case Report

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대한영상의학회지

Endovascular Treatment of a Systemic-to-Pulmonary Artery Fistula: A Case Report

체-폐동맥루의 혈관 내 치료: 증례 보고

Eunbyul Lee, MD, Dong Jae Shim, MD*, Doyoung Kim, MD, Jung Whee Lee, MD

Department of Radiology, Incheon St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Incheon, Korea

Systemic-to-pulmonary artery fistulas are rare. This condition may be congenital, post-traumatic, or post-inflammatory and can cause infection, hemorrhage, or pulmonary hypertension. Here, we report a case of an intercostal-to-pulmonary artery fistula, incidentally detected during the evaluation of dyspnea in a 67-year-old female. Retrograde transcatheter coil embolization in a dilated draining pulmonary artery was initially attempted. However, another draining pulmonary artery developed after 5 months. The intercostal arteries or systemic feeders were successfully embolized through a transarterial access. At the 10-month follow-up, the abnormally dilated vessels had regressed, and dyspnea had improved. Sequential or simultaneous retro- and antegrade transcatheter embolization may successfully treat pleural arterio-arterial fistulas.

Index terms Vascular Fistula; Therapeutic Embolization; Arterio-Arterial Fistula

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***Corresponding author**
Dong Jae Shim, MD
Department of Radiology,
Incheon St. Mary's Hospital,
College of Medicine,
The Catholic University of Korea,
56 Dongsu-ro, Bupyung-gu,
Incheon 21431, Korea.

Tel 82-32-280-5236
Fax 82-32-280-5230
E-mail inharad@naver.com

ORCID iDs
Eunbyul Lee
https://orcid.org/0000-0002-9106-1647
Dong Jae Shim
https://orcid.org/0000-0001-9596-0765
Doyoung Kim
https://orcid.org/0000-0003-1937-0661
Jung Whee Lee
https://orcid.org/0000-0001-8949-5385

INTRODUCTION

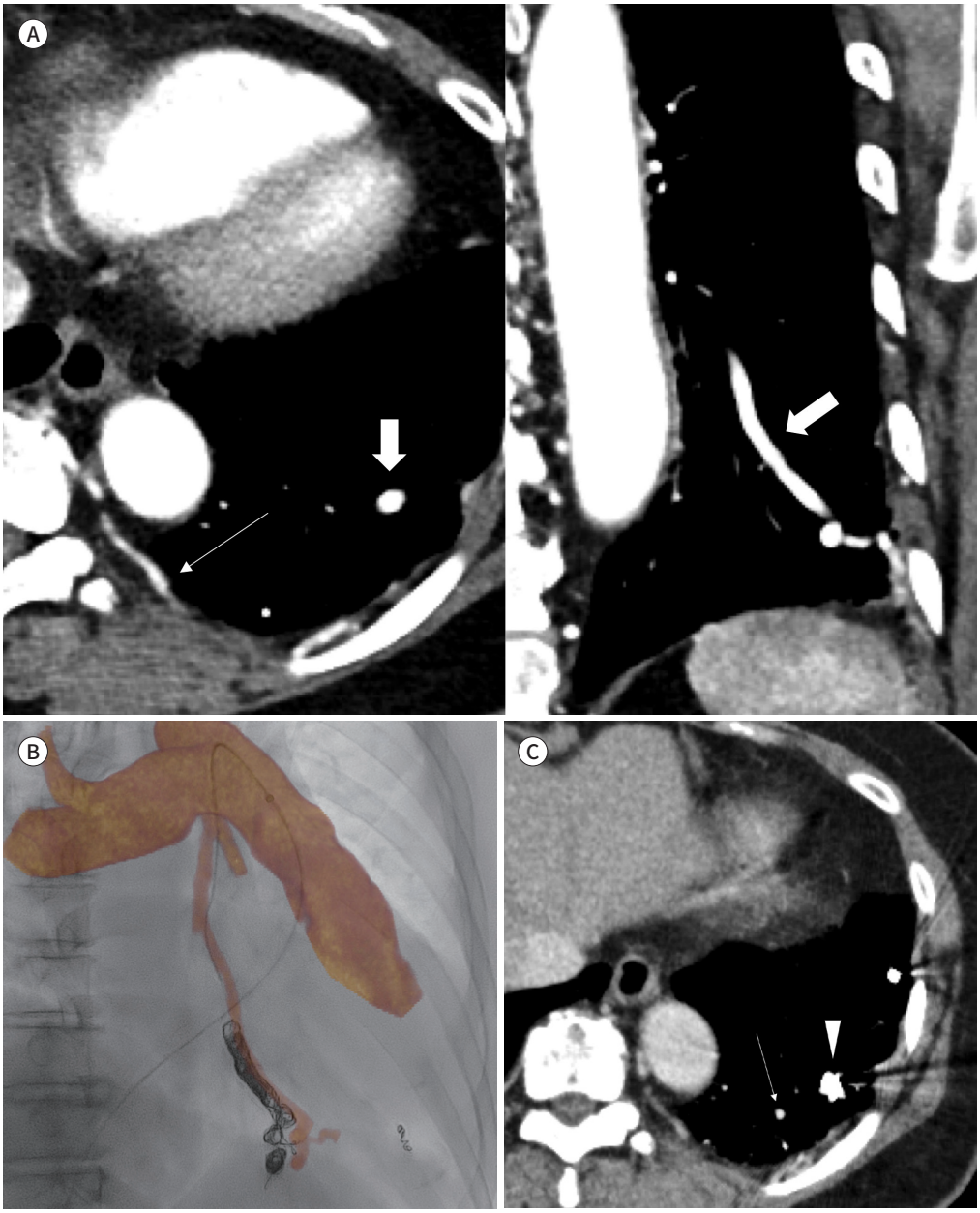
Systemic-to-pulmonary artery fistula (S-PAF) of unknown aetiology is rare. Symptoms may include haemoptysis, dyspnea, and heart failure (1). Although most of the patients are usually asymptomatic, S-PAF has risk of adverse hemodynamic effect, bacterial vegetation, and rupture (2). S-PAF differs from pulmonary arteriovenous malformation because pulmonary arteries serve as draining vessels, and there is neither a nidus nor a dilated pulmonary vein (3). S-PAFs are either congenital or of acquired aetiology (1). Congenital S-PAF is extremely rare. Most patients acquire S-PAF from trauma, neoplasms, or inflammatory lesions (4). Published reports show surgical or endovascular treatment options, with the latter favoured due to minimal invasiveness (5). Favourable endovascular treatments include systemic artery embolization, although the presence of multiple feeders and draining vessels may prove challenging (6). In addition, if all

feeding and draining vessels are not completely occluded, a fistula can easily recur (7). Herein, we present a case report of a patient with intercostal arteries to pulmonary artery fistula that was successfully treated by pulmonary artery occlusion and subsequent systemic artery embolization.

CASE REPORT

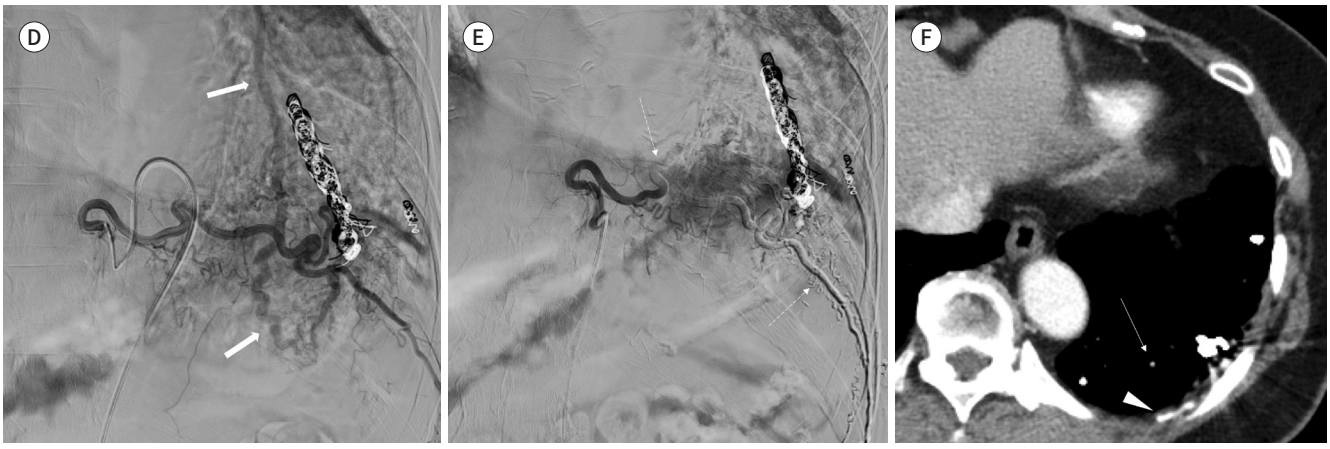
An exemption was granted from our Institutional Review Board. A 67-year-old female presented with mild dyspnea (Modified Medical Research Council Scale III), occurring for 3 months without history of trauma, cardiovascular disease, or lung disease (8). Examination showed no evidence of wheezing or cardiac murmurs, and oxygen saturation was 98%. The patient had a pulmonary function test, which showed no obstructive airway disease. A chest radiograph revealed a tortuous tubular opacity in the left-lower lung field, suggesting a dilated vascular shadow. A chest CT scan (Brilliance 64; Philips Medical Systems, Cleveland, OH, USA; acquisition parameters: tube voltage 120 kVp, effective tube current 200 mAs, collimation 0.625 mm, slice thickness 2 mm, pitch 0.763, field of view 288 mm) acquired two-day before procedure revealed an enlarged left-lower posterior basal segmental pulmonary artery and mildly dilated adjacent left-lower intercostal arteries (Fig. 1A). There was no evidence of previous inflammatory sequelae in the lung parenchyma, pleura, and chest wall. Neither a nidus nor a dilated pulmonary vein was found when tracing the dilated arteries, suggesting a case of S-PAF. However, the connection between the intercostal and pulmonary arteries were not evident on CT. The clinician considered either surgery or endovascular treatment, however the patient preferred less invasive treatment. We attempted to embolize the pulmonary artery at the first visit because the fistula had one draining pulmonary artery with multiple feeding systemic arteries suspected. In the case of an unsuccessful embolization, antegrade systemic arterial embolization would be attempted at a subsequent visit. Raw data of CT uploaded to angiography machine (AlluraClarity; Philips Healthcare, Best, the Netherlands) before procedure. After merging process of CT image with fluoroscopy by two-different angle, the real-time roadmap image guided the way to the lesion in accordance with dynamic change of fluoroscopy. Left-lower lobar pulmonary arteriography via right femoral vein access failed to demonstrate a dilated artery due to retrograde blood flow. Thus, the dilated draining pulmonary artery was catheterized with the guidance of a pre-acquired CT real-time three-dimensional roadmap (Vessel Navigator; Philips Healthcare). The pulmonary artery was then embolized with detachable micro-coils (Fig. 1B, one Interlock coil, 6 mm × 100 mm; Boston Scientific, Marlborough, MA, USA; five Concerto coils 7 mm × 300 mm × 2 and 8 mm × 300 mm × 3; Medtronic, Dublin, Ireland). A follow-up chest CT scan after 5 months revealed little change in the enlarged left-lower intercostal arteries and showed newly developed branches from the left-lower posterior basal pulmonary artery (Fig. 1C). At a subsequent visit, the left 10th intercostal arteriography showed retrograde filling into the pulmonary artery, which suggested recurrent S-PAF (Fig. 1D). The feeding artery was embolized with a mixture of N-butyl-2-cyanoacrylate (Histoacryl; B. Braun, Melsungen, Germany) and ethiodized oil (Lipiodol; Guerbet, Aulnay-sous-Bois, France) at a ratio of 1:2 (Fig. 1E). In addition, the 9th intercostal arteriography showed minor abnormal communication; thus,

Fig. 1. A 67-year-old female with mild dyspnea presented with systemic-to-pulmonary artery fistula.
A. Axial and coronal reconstructed CT images show a dilated left-lower posterior basal segmental pulmonary artery (solid arrows) and an adjacent left lower intercostal artery (arrow).
B. The dilated draining pulmonary artery was catheterized under the guidance of a real-time three-dimensional roadmap made of pre-acquired CT and then embolized with detachable micro-coils.
C. A CT image at the 5-month follow-up shows a newly enlarged left-lower posterior basal pulmonary artery (arrow) and coil-embolized pulmonary artery (arrowhead).



we performed embolization with polyvinyl alcohol (PVA, Contour 355–500 μm; Boston Scientific) particle. Arterio-arterial fistula was no longer present on the post-procedure arteriography. A follow-up chest CT scan after 10 months revealed a reduced size in the pulmonary artery and intercostal arteries and improved dyspnea in the patient (Modified Medical Research Council Scale II) (Fig. 1F).

Fig. 1. A 67-year-old female with mild dyspnea presented with systemic-to-pulmonary artery fistula.
D. The left 10th intercostal arteriography shows retrograde filling into the pulmonary artery (arrows), suggesting a systemic-to-pulmonary artery fistula.
E. Completion arteriography shows occlusion of the intercostal artery (arrows) and no remaining arterio-arterial fistulas.
F. A CT image at the 10-month follow-up shows reduced size of the pulmonary artery (arrow) and the glue-embolized left 10th intercostal artery (arrowhead).



DISCUSSION

S-PAFs at pleura are very rare in the absence of underlying disease (1). Most patients with S-PAFs fistulae are asymptomatic, however, there are several potential complications of S-PAFs such as bacterial seeding, enlargement with rupture, pulmonary artery hypertension and propensity for heart failure. So it is important to confirm the presence of these fistulae and treatment (2). Most reported cases were treated by surgery, and only a few received endovascular treatment (5). Diagnosis by CT is challenging because CT cannot show a hemodynamic state, such as retrograde flow in the pulmonary artery. Furthermore, communication between the systemic and pulmonary arteries was not clearly demonstrated in the patient presented here. Our patient was older than most reported in the literature, who ranged from 7–44 years old (4). The patient denied a history of risk factors; thus, the cause may be congenital S-PAF with symptoms at a late stage or acquired S-PAF from a recent unknown inflammatory disease. The natural course of S-PAF is unknown, and no established standard treatment exists. Although treatment is controversial, transcatheter embolization has shown success (5). Although retrograde access is technically challenging with a possibility of recurrence due to overflow to adjacent vessels, a stepwise retro- and antegrade approach was supported (9). Previous studies of endovascular treatment for peripheral arteriovenous malformations showed efficacy by embolization through transvenous retrograde access (6). Jackson et al. (7) suggest that transvenous embolization techniques may be helpful if multiple small feeding arteries supply the lesion or an extreme arterial tortuosity precludes successful catheterization. Similar to peripheral arteriovenous malformations, S-PAF with multi-feeders can be occluded by a retrograde approach. In addition, embolization of the draining artery can prevent inadvertent distal embolization. Detachable coils can be a safe embolic material for high flow vascular lesions. However, deployment of coil requires firm support of delivering catheter and microcatheter placement to the distal end of supplying artery. In this present-

ing case, stiff and tortuous course of the feeding arteries made it difficult to cannulate distally. We chose N-butyl-2-cyanoacrylate and PVA for the recurrent S-PAF, because the 10th intercostal artery had high flow and direct communication with draining pulmonary artery and 9th intercostal artery had relatively small diameter. Lois et al. (10) described that PVA is a good embolization agent to close smaller collateral vessels. In conclusion, although pleural S-PAFs without underlying disease are rare, close inspection of CT scans and systemic arteriography can aid in diagnosis. Sequential or simultaneous retro- and antegrade transcatheter embolization is a minimally invasive treatment option.

Author Contributions

Conceptualization, S.D.J.; data curation, L.E., K.D.; investigation, L.E., L.J.W.; project administration, S.D.J.; resources, L.J.W.; software, S.D.J.; supervision, S.D.J.; Validation, S.D.J.; visualization, L.E.; writing—original draft, L.E., K.D.; and writing—review & editing, S.D.J., L.J.W.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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체-폐동맥루의 혈관 내 치료: 증례 보고

이은별 · 심동재* · 김도영 · 이정휘

체-폐동맥루는 주로 외상, 감염, 종양 등에 의해 유발될 수 있으나 위험 인자가 없는 환자에서 생기는 체-폐동맥루는 매우 드물다. 이들은 감염이나 출혈 혹은 폐 고혈압 등을 유발할 수 있어 치료가 필요하다. 호흡곤란을 주소로 내원한 67세 여자 환자에서 늑간동맥-폐동맥루가 발견되었다. 먼저 배출혈관인 확장된 폐동맥으로 역행성으로 접근하여 코일 색전술로 치료하였으나 5개월 후 인접폐동맥의 확장이 관찰되어 공급동맥인 늑간동맥을 N-butyl cyanoacrylate를 이용하여 색전하였다. 10개월 추적검사에서 환자의 증상과 확장된 폐동맥은 호전되었다. 체-폐동맥루를 치료에서 순차적 혹은 동시에 역방향과 순방향 접근으로 혈관 내 치료하는 것이 효과적일 수 있겠다.

가톨릭대학교 의과대학 인천성모병원 영상의학과

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