

Coil embolization of a fistula from the right inferior phrenic artery to the right pulmonary artery with involvement of further arteries: A rare case report

Juergens LJ, Thalhammer A, Gruber-Rouh T, Koch V, Vogl TJ, Martin SS

Goethe University Frankfurt, University Hospital, Department of Diagnostic and Interventional Radiology, Germany

*Correspondence: Lisa Joy Juergens, Department of Diagnostic and Interventional Radiology, University Hospital Frankfurt, Theodor-Stern-Kai 7, 60596 Frankfurt, Germany, Tel: +49 151 58814870, Fax: +49 69 6301-7258,

✉ lisajoy@juergensfamily.de

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ABSTRACT

A 51-year-old female patient was presenting dyspnea for more than a year with no previous lung infections or surgery. Initially, a diagnostic computed tomography was made, showing a rare arterio-arterial malformation between the right inferior phrenic and right pulmonary artery leading into a vascular bundle in the middle lung lobe. Due to the patients' dyspnea and massive extent of malformation, the indication for transcatheter arterial embolization was made. The first transcatheter arterial embolization procedure involved the inferior phrenic and a selective branch of the internal thoracic artery. Interventional angiography as well as computed tomography revealed further extend of the malformation showing a connection of right lateral thoracic, hepatic, and inferior epigastric artery to the fistula. After one month, a second transcatheter arterial embolization of these arteries as well as a second approach of the proximal internal thoracic artery was performed. In the follow-up the patient described a substantial improvement of her dyspnea and showed no signs of infections.

A phrenic artery to pulmonary artery fistula is an extremely rare case occurring congenital or acquired. Patients may be asymptomatic or present, among others, dyspnea, hemoptysis, pulmonary infections and congestive heart failure. Symptomatic patients require treatment using transcatheter arterial embolization or surgical resection. The patient had dyspnea and a substantial extent of malformation with possibly complicated clinical course. The recommended less invasive treatment using transcatheter arterial embolization was successfully performed. In conclusion, our patient represented a rare congenital case of systemic and pulmonary artery communication, which we were able to treat sufficiently with coil embolization.

CASE REPORT

CASE REPORT

A 51-year-old woman was presented to our hospital with dyspnea during minimal exercise for more than a year. The patients' medical history shows a COVID pneumonia in May 2022, and no signs of surgery, trauma or smoking. A computed tomography (CT) revealed an arterio-pulmonary arterial malformation of the right pulmonary artery and inferior phrenic artery with vascular bundle in the middle lung lobe. Additionally, imaging showed an enlargement of the right inferior phrenic artery as well as right internal thoracic artery (Figures 1A,1B). Due to the patients' exertional dyspnea, the massive extent of the fistula and possible complications in the clinical course, the

indication of a transcatheter arterial embolization (TAE) was made. The TAE procedure was executed with a transfemoral approach under local anesthetic. First, the right inferior phrenic artery located above the coeliac trunk was approached. After the application of the contrast media, it showed the substantial extent of the shunt between inferior phrenic artery and the right pulmonary artery. Further, its vascular bundle located in the middle lobe of the right lung was revealed. A selective approach into the enlarged inferior phrenic artery with a microcatheter was performed. Initially, several coils (Tornado 5/2 mm, Cook Medical, Bloomington USA) were applied into the center of the vascular bundle and, subsequently, into the main branch of the inferior phrenic artery using two removable coils (Concerto 6mm x 20cm and 7mm x 30cm, Medtronic, Irvine USA).

Thereafter, the right internal thoracic artery was approached using a vertebralis catheter. The same vascular bundle showed enhancement over the right internal thoracic artery. Therefore, another embolization using coils (Tornado 5/2 mm, Cook Medical) and two removable coils (Concerto 5mm x 15cm, Medtronic) were applied in the same manner (Figure 2A). In our final control, all coil embolized artery were properly sealed. The final application of contrast media via catheter showed a filling of the right inferior epigastric artery with a slight supply into the embolized vascular bundle. A follow-up and a reevaluation via CT took place a week after the TAE. It showed an additional connection of the right lateral thoracic, hepatic artery and inferior epigastric artery and further extent from the internal thoracic artery to the arterio-pulmonary arterial shunt described before. In this first reevaluation, the patient described an improvement of her dyspnea and well-being. After one month a second TAE procedure was conducted, using the same technic as managed before. During this procedure, initially, the right hepatic artery was approached, and embolized with coils (Tornado 3/2 mm, Cook Medical). Subsequently, a selective approach of the right inferior epigastric artery using a vertebralis catheter and a microcatheter showed a connection to the vascular bundle (Figure 2B). Likewise, a coil embolization was performed (Tornado 3/2mm, Cook Medical). Afterwards, the right subclavian artery was approached with an embolization of two branches of the lateral thoracic artery and a new embolization of the proximal internal thoracic artery, using multiple coils (Tornado 3/2 mm and 5/2 mm, Cook Medical). The final application of contrast media showed all arteries leading to the vascular bundle were sealed properly. In total 74 coils were used during the two transcatheter arterial embolization procedures. In the next follow-up, our patient showed a further improvement of her exertional dyspnea with no signs of infection.

DISCUSSION

Etiology and demographics

The inferior phrenic artery to pulmonary artery fistula is a highly unusual case of trans pleural connection [1]. This rare arterio-arterial malformation can occur congenital, however is most frequently acquired due to some pulmonary diseases, including pulmonary tuberculosis, bronchiectasis or lung carcinoma, but can also be caused due to surgery or trauma [1,2]. There appears to be no gender-predilection and the symptoms tend to present during middle age [3].

Clinical and imaging findings

Usually, this sort of fistula is asymptomatic and is primarily recognized incidentally during radiological imaging such as CT [4]. However, patients may present dyspnea, major hemoptysis, recurrent pulmonary infections, pulmonary hypertension and congestive heart failure [1,3].

Treatment and prognosis

Minor malformations as well as asymptomatic patients do not require further treatment and are mainly observed

thoroughly. Symptomatic patients and those with a substantial extent of malformation require treatment using transcatheter arterial embolization (TAE) or surgical resection [2].

To our best knowledge, a single digit number of cases of this left- to right shunt have been reported [3]. The connection of systemic and pulmonary artery was of substantial extent, presenting a symptomatic patient with dyspnea, which led to our recommendation for a coil embolization. This decision was also driven due to the possibly complicated clinical course with increased right ventricular afterload and right heart insufficiency. The recommended approach to treat this sort of fistula is a TAE, a surgical resection, or a combination of both [1]. We determined the TAE to be less invasive and more beneficial for our patient, with the possibility of further alternative approaches after this procedure. In conclusion, our patient represented a rare congenital case of systemic and pulmonary artery communication, which we were able to treat sufficiently with coil embolization.

Differential diagnosis

The patients' symptom of chronic exertional dyspnea allows for a range of diagnoses. Our patient has a history of COVID pneumonia but no signs of pulmonary or cardiac diseases. Clinically there were no further significant findings. The radiological imaging shows the inferior phrenic to pulmonary artery fistula very prominently. Based thereon, we determined the patients' symptoms most likely originated from this fistula. Radiological imaging also allows to differentiate between arterio-arterial fistula and arterio-venous fistula. Arterio-arterial fistula are an unusual occurring mimicker of the arteriovenous fistula [5]. A list of differentials and their imaging findings are elaborated in the differentials table.

TEACHING POINT

Patients with arterio-arterial malformation will benefit and present significant improvement of symptoms and well-being, using the minimal invasive procedure of transcatheter arterial embolization, showing it's essential role in treatment of these diseases.

AUTHORS 'CONTRIBUTIONS

- LJJ has written the case report and assisted in the two procedures.
- AT evaluated the images, performed the two procedures.
- SSM evaluated the images, performed the two procedures, and controlled all written content.
- TGR has evaluated the images and advised in the procedures.
- VK has evaluated the images.
- TJV has evaluated all images and advised to the procedure.

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DISCLOSURES

- No financial funding
- The authors declare that they have no competing interests.

CONSENT

- Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

HUMAN AND ANIMAL RIGHTS

- All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

QUESTIONS

Question 1: How often does the inferior phrenic artery to pulmonary artery fistula occur? Choose the correct answer:

- a) often
- b) rarely (applies)
- c) not at all
- d) depends on the gender
- e) no correct answer

Explanation Question 1: The correct answer is b). The inferior phrenic artery to pulmonary artery occurs rarely. To our best knowledge so far only a single digit of numbers has occurred.

[The inferior phrenic artery to pulmonary artery fistula is a highly unusual case of trans pleural connection.]

Question 2: What may cause an arterio-arterial fistula? Choose the correct answer:

- a) pulmonary tuberculosis, bronchiectasis, or lung carcinoma
- b) surgery or trauma
- c) pulmonary diseases
- d) a + b + c is correct (applies)
- e) no answer is correct

Explanation Question 2: The correct answer is d). Pulmonary diseases, surgery and trauma may cause an arterio-arterial fistula.

[This rare arterio-arterial malformation can occur congenital, however is most frequently acquired due to some pulmonary diseases, including pulmonary tuberculosis, bronchiectasis or lung carcinoma, but can also be caused due to surgery or trauma].

Question 3: What treatment is required in arterio-arterial malformation with substantial extent? Choose the correct answer:

- a) no therapy is required
- b) surgery
- c) transcatheter arterial embolization

d) b + c is correct (applies)

e) no answer is correct

Explanation Question 3: The correct answer is d). Substantial extent and patients with symptoms may require transcatheter arterial embolization or surgery.

[Symptomatic patients and those with a substantial extent of malformation require treatment using transcatheter arterial embolization (TAE) or surgical resection.]

Question 4: A patient with no symptoms and a minor extent of malformation, wants to know how to treat the malformation? Choose the correct answer:

- a) we will mainly observe the patient thoroughly (applies)
- b) we give the patient medications
- c) we recommend a surgery
- d) we recommend staying in the hospital due to its rapid progression
- e) no answer is correct

Explanation Question 4: The correct answer is a). Patients with a minor extent of malformation or even asymptomatic patients may be observed thoroughly, not requiring direct treatment.

[Minor malformations as well as asymptomatic patients do not require further treatment and are mainly observed thoroughly.]

Question 5: A patient with massive hemoptysis presents a massive extent of arterio-arterial malformation in the computer tomography imaging. What does apply to this disease? Choose the correct answer:

- a) it occurs rarely
- b) other symptoms may include dyspnea
- c) transcatheter arterial embolization is a less invasive method to treat this patient
- d) b+ c is correct
- e) a + b + c is correct (applies)

Explanation Question 5: The correct answer is e). The inferior phrenic artery to pulmonary artery fistula occurs rarely, it has a variety of symptoms such as dyspnea and can be treated with the transcatheter arterial embolization.

[The inferior phrenic artery to pulmonary artery fistula is a highly unusual case of trans pleural connection.]

[However, patients may present dyspnea, major hemoptysis, recurrent pulmonary infections, pulmonary hypertension and congestive heart failure.]

[The recommended approach to treat this sort of fistula is a TAE, a surgical resection, or a combination of both].

REFERENCES

- [1] Wynne DM, Jefferson XD. Jefferson Coil Embolization of an inferior Phrenic Artery-to-Pulmonary fistula. *J Vasc Interv Radiol.* 2022; 33(6): 736-738. PMID: 35314369.

[2] Zhang W, Jin X, Wang B, Wang J. Phrenic artery with fistula formation to pulmonary artery and vein: a rare case report. *Asian J Surg.* 2022; 45(12): 2991-2992. PMID:35803892.

[3] Kawakado K, Yabuki T, Nishi T, et al. Successful transcatheter arterial embolization of asymptomatic aneurysm associated with left inferior phrenic artery-to-pulmonary artery fistula: a case report. *Respir Med Case Rep.* 2021; 33: 101444. PMID:34401284.

[4] Souza AS, Soares Souza LV, Marchiori E. Asymptomatic arterio-arterial fistula between pulmonary and phrenic arteries. *Eur J Cardiothorac Surg.* 2019; 56(4): 816. PMID: 31220235.

[5] Fish A, Chan SM, Pollak J, Schlachter T. Transpleural Systemic Artery-to-Pulmonary Artery Shunts: A Potential Mimicker of Pulmonary Arteriovenous Malformations. *J Vasc Interv Radiol.* 2023; 34(2): 300-303. PMID: 36283590.

FIGURES

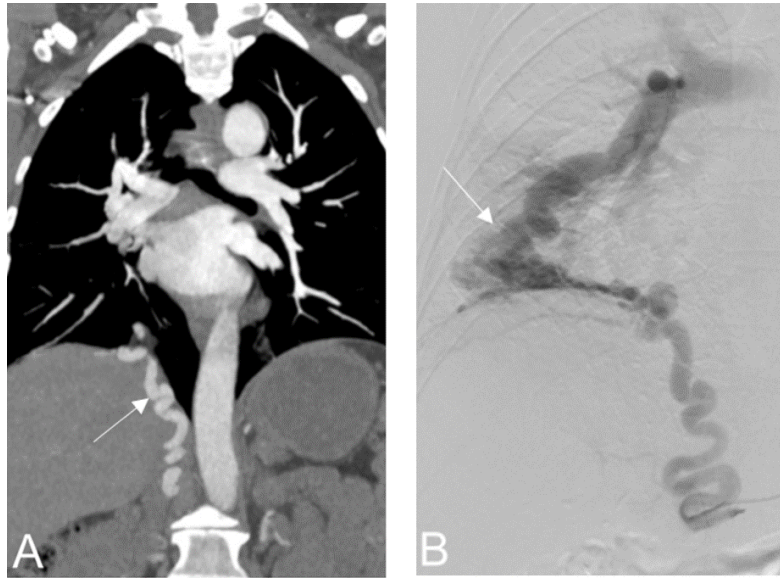


Figure 1: 52 -year-old female with a fistula from the right inferior phrenic artery to the right pulmonary artery

Findings and Technique:

(A) Contrast enhanced coronal computed tomography showing the enlargement of the right inferior phrenic artery (white arrow).

(B) Subsequent angiography revealed a malformation between the enlarged right inferior phrenic artery to the right pulmonary artery, forming a left-to right shunt (white arrow).

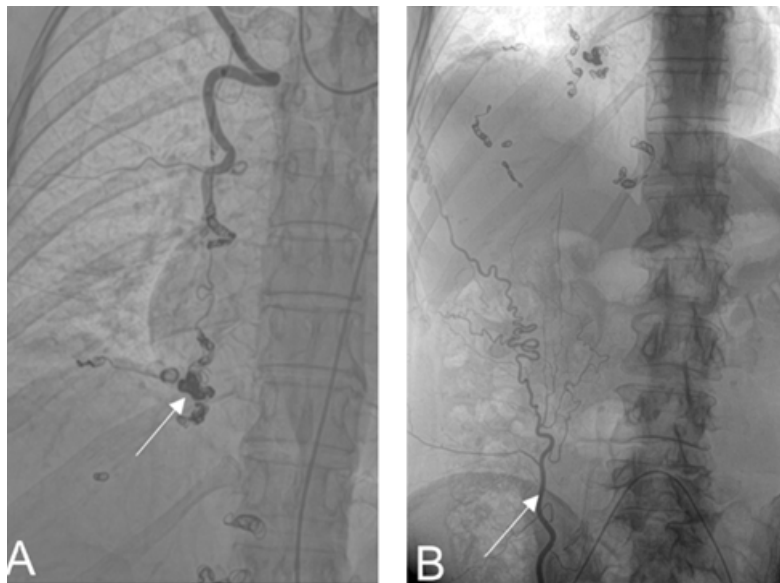


Figure 2: 52 -year-old female with a fistula from the right inferior phrenic artery to the right pulmonary artery

Findings and Technique:

(A) Patients' angiographic images with transcatheter to the right lower chest illustrating an enlargement of the right thoracic artery connecting with the malformation of phrenic artery to pulmonary fistula after coil embolization, showing a complete occlusion of the fistula (white arrow).

(B) Patients' angiographic images illustrating an enlargement of the inferior epigastric artery (white arrow) connecting with the malformation of phrenic artery to pulmonary fistula after coil embolization.

Summary table	Inferior phrenic artery to the pulmonary artery fistula
Etiology	Congenital or acquired
Incidence	single digit number of cases known to our best knowledge
Gender ratio	No gender predilection
Age predilection	Middle age
Risk factor	The fistula can be acquired due to some pulmonary diseases, including pulmonary tuberculosis, bronchiectasis, or lung carcinoma, but can also be caused due to surgery or trauma
Treatment	Minor malformations/asymptomatic patients <input type="checkbox"/> no further treatment required, mainly observation. Substantial extent malformations/ symptomatic patients <input type="checkbox"/> require treatment using transcatheter arterial embolization or surgical resection.
Prognosis	The transcatheter arterial embolization is less invasive and showed to improve the patients' symptoms with no side effects and infections in the follow ups up to date. Without treatment symptoms and the malformation may worsen
Findings on imaging	CT: <ul style="list-style-type: none"> the enlargement of the right inferior phrenic artery Angiography: <ul style="list-style-type: none"> malformation between the enlarged right inferior phrenic artery to the right pulmonary artery, forming a left-to-right shunt, enlargement of the right thoracic artery connecting with the malformation of phrenic artery to pulmonary fistula after coil embolization Coil embolization of the fistula enlargement of the inferior epigastric artery connecting with the malformation of phrenic artery to pulmonary fistula after coil embolization.

Table 1: Summary table of inferior phrenic artery to the pulmonary artery fistula

	X-Ray findings	CT findings	Angiography findings	MRI findings
Arterio-arterial Fistula	Dilated pulmonary vessels may appear as non-specific soft tissue white- grey mass lesion	Enlargement of vessels communicating with the pulmonary arteries. Hypertrophy of systemic arteries in the chest wall and diaphragm, greater than expected with chronic lung disease.	Enlargement of vessels communicating with the pulmonary arteries, good assessment of extend of the malformation, treatment	MR angiography considered MR technique of choice for imaging vascular structures in the thorax. Enlargement of vessels communicating with the pulmonary arteries.
Arterio- venous Fistula	Dilated pulmonary vessels may appear as non-specific soft tissue white mass with relatively unusual orientation compared to other vessels, nodular shadow	Tubular structures communicating with the pulmonary veins, hypertrophy of systemic arteries in the chest wall and diaphragm, greater than expected with chronic lung disease. Absence of associated chronic lung parenchyma disease	Angiography confirms diagnosis, showing extend of the complex vascular structures	MR angiography considered MR technique of choice for imaging vascular structures in the thorax. Tubular structures communicating with the pulmonary veins.
Lung Cancer	May appear as mass lesions in lungs, or pulmonary nodule	Lung mass/ focal lesion with possible blurred borders, no calcifications, spicule, enlarged lymph nodes, pleural effusion, atelectasis and consolidation.		Lung mass/ focal lesion enlarged lymph nodes, pleural effusion, atelectasis and consolidation.
Pulmonary vein varix	Nonspecific, may present as lung mass, round to oval homogenous shadow	CT angiography showing the arterial and venous vessels, hyperdense mass with continues vessels, pulmonary consolidation, pleural effusion	Allows for accurate delineation if arterial and venous anatomy, important to make definite diagnosis	MR angiography showing the arterial and venous vessels
Pulmonary tuberculosis	Patchy consolidations, caverns, pulmonary nodules	Parenchymal consolidation in any lung lobe, caverns, pulmonary nodules, predominance in lower and middle lobe, lymphadenopathy, pleural effusion	Uncommonly used in pulmonary tuberculosis diagnosis, pulmonary angiography rather used in therapy with coil embolization due to symptoms such as hemoptysis	Parenchymal consolidation in any lung lobe, caverns, pulmonary nodules, predominance in lower and middle lobe, lymphadenopathy, pleural effusion

Table 2: Differential table for arterio-arterial fistula

KEYWORDS

Arterio-arterial fistula; Transcatheter arterial embolization; arterio-arterial malformation; Interventional radiology; inferior phrenic artery to pulmonary artery fistula

ABBREVIATIONS

CT = Computed Tomography; TAE = Transcatheter Arterial Embolization

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