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CASE REPORT – OPEN ACCESS

A Case of a Systemic-to-Pulmonary Artery Fistula and its Endovascular Treatment

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ABSTRACT

Background: Systemic-to-pulmonary artery fistula has been identified as a very rare condition of which the clinical course cannot be fully predicted. Thus, the precise indication for treatment remains to be unclear.

Case Report: A 42-year-old man, without any history of congenital heart disease or chest wall injury, presented with a 3-day history of vague chest pain. As the initial examinations were found unremarkable, the patient underwent coronary CT angiography revealing a fistula between the left internal mammary artery (LIMA) and the upper lobe pulmonary artery. After confirming the fistula via selective LIMA angiography followed by coronary angiography revealing normal coronaries without fistulous connection, the fistula was closed using cyanoacrylate glue injection. Although a nontarget pulmonary embolization was observed to have occurred to a small extent, the complaint of the patient entirely disappeared following the procedure.

Conclusion: Considering the potential complications, it seems more favorable to close a systemic-to-pulmonary artery fistula. For a safe and effective treatment, the flow dynamics should be considered when choosing the method of embolization.

Keywords: Arterio-arterial fistula, embolization, lung

INTRODUCTION

The lung is one of the two organs that receive blood via two distinct circulatory routes, i.e., systemic circulation and pulmonary arterial circulation. Apart from bronchial arteries, any systemic arterial supply to the lung is deemed unusual. On the other hand, in patients with congenital heart disease or pulmonary vascular abnormality, systemic-to-pulmonary artery (S-PA) connections may occur. Although more rare, such a connection may be detected even in individuals with normal bronchial and pulmonary vascular structures (1, 2). Here we report a case of S-PA fistula with speculative origin and further discuss its etiology and treatment strategy.

CASE REPORT

A 42-year-old man presented with a 3-day history of vague chest pain. The patient had been having an intermittent undiagnosed pain under his left rib cage since childhood; however, he had no history of congenital heart disease or chest wall injury. In addition, he experienced a lung infection almost 20 years ago and has undergone surgery for thyroid cancer. He had a 12.5 pack-year smoking history as well. Initial examination, chest X-ray, ECGs, cardiac enzymes, and other routine blood tests were found to be unremarkable. He continued with discomfort in the area; hence, a coronary CT angiography was performed on the following day to rule out coronary artery disease. There was no abnormality of heart and coronary arteries on CT; however, a tangle of vessels within the epicardial fat next to the heart apex was detected, incidentally. The tangle was found to be associated with the left internal mammary artery (LIMA) and the left upper lobe pulmonary artery (Figs. 1a, b). A thin-walled, 2 x 2.8 cm sized, air-filled cystic space next to the tangle was noticed in lung window settings, additionally. To evaluate the flow dynamics and also to decide the treatment strategy, selective LIMA angiography was performed via the left brachial arterial route using a 5 Fr catheter. The arteriograms revealed a mildly hypertrophied LIMA, with an enlarged and tortuous branch related to the tangled vessels, and a retrograde flow was seen in a pulmonary artery branch through a fistula. The venous drainage of the involved lung segment was by way of the left inferior pulmonary vein (Figs. 2a, b). Upon this finding, coronary angiography was performed a few days later, revealing normal coronaries without fistulous connection. Transthoracic echocardiography showed normal right heart chambers with indirect systolic right ventricle pressure of 25 mm/Hg. As the fistula could cause complications in the future, we preferred to close it. The method we opted to use was glue injection because of its simplicity, practicality, and cost-effectivity. Since there was an actually artery-to-artery fistula, it was considered that the pressure gradient would be relatively lower, allowing a safe injection.

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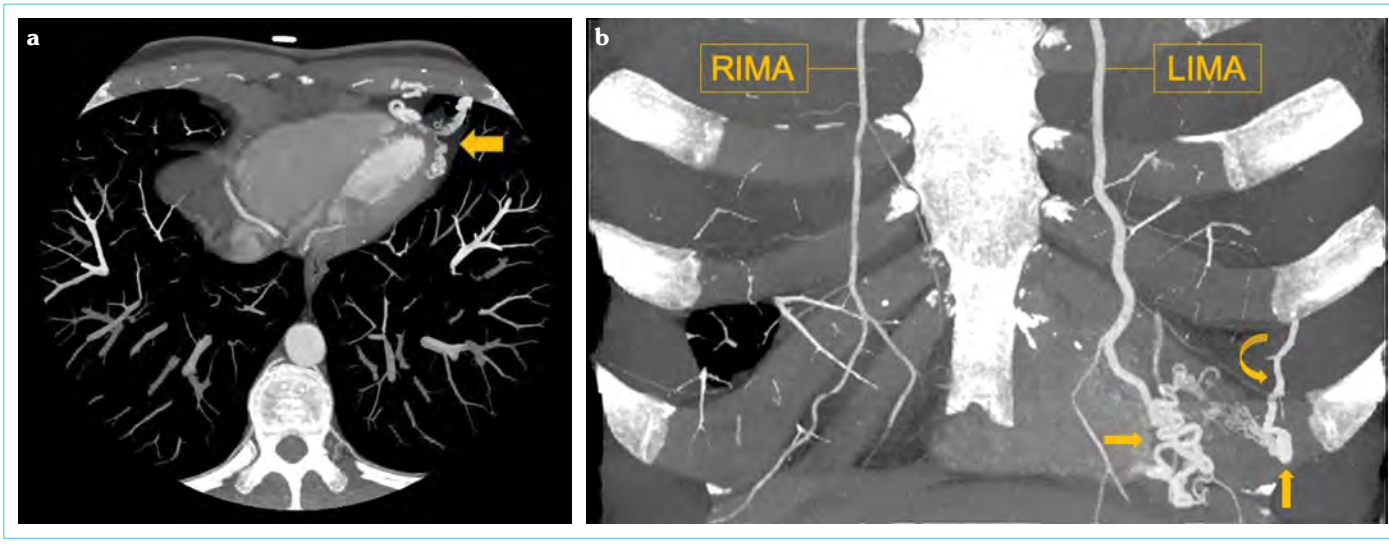

Figure 1. (a) Coronary CT angiography, axial minimum-intensity projection (MIP). A tangle of vessels within the epicardial fat next to the heart apex is seen (horizontal arrow). (b) Coronary CT angiography, coronal minimum-intensity projection (MIP). It can be seen that a tangle of vessels (horizontal arrow) and a segmentary branch of the left pulmonary artery (curved arrow) are connected via a fistula (vertical arrow). Note that the LIMA is hypertrophied. RIMA right internal mammary artery, LIMA left internal mammary artery

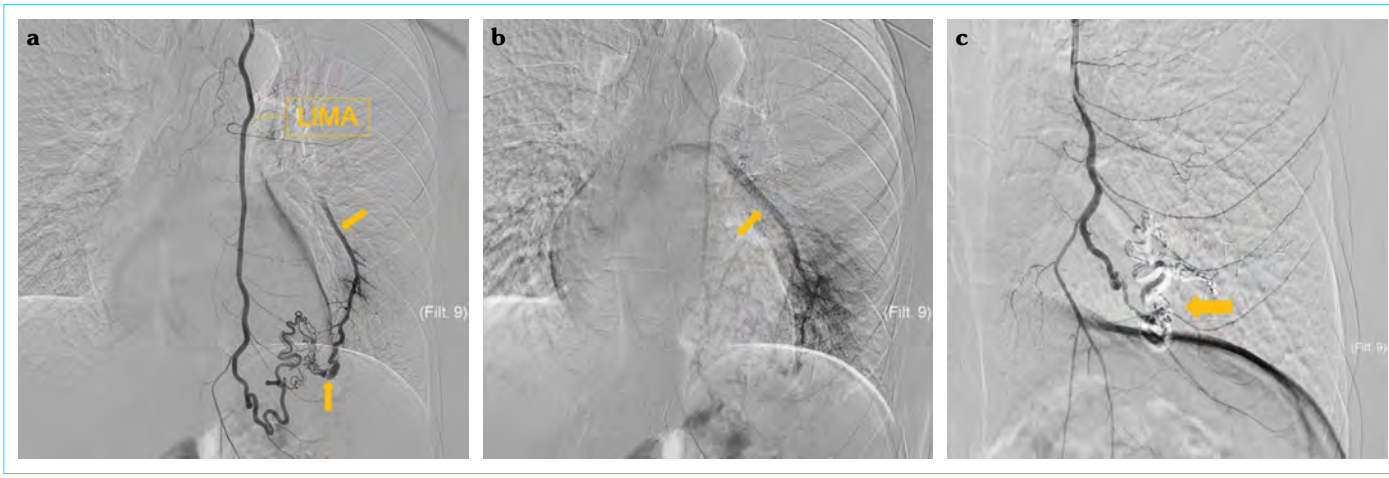

Figure 2. (a) Digital subtraction angiography. Selective angiogram well-depicts the fistulous connection (vertical arrow) between the LIMA and a segmentary branch of the left pulmonary artery (oblique arrow). LIMA left internal mammary artery. (b) Digital subtraction angiography. Note the pulmonary vein filling in the late phase, which is normal in caliber (oblique arrow). (c) Digital subtraction angiography. Final selective angiogram shows complete occlusion of the fistula (horizontal arrow)

At a different session, the LIMA was catheterized as previously, followed by superselective catheterization of the LIMA branch supplying the fistula using a 3 Fr coaxial catheter. After advancing the microcatheter as far as possible, a mixture of cyanoacrylate glue/iodized oil at a 1:8 ratio was injected. A small amount of the agent spread throughout the territory of the involved pulmonary artery at the beginning of the injection because of the lower concentration. After a short while to allow the agent to polymerize, the injection was continued until the tangled vessels were noted to embolize, ensuring the closure of the fistula. Although a nontarget pulmonary embolization occurred to a small extent, the whole procedure was completed using a total volume of 1.5 ml of the mixture thanks to gradual embolization. A control arteriogram demonstrated no further flow across the fistula (Fig. 2c). The patient was then admitted overnight for monitoring and discharged the next day. Seventeen days later, a chest CT scan was performed, because the patient complained of sudden onset chest pain. However, no acute abnormality was detected apart from the cast of embolic material filling the tangled vessels and dispersed linear opacities within the lingula, representing nontarget emboli (Fig. 3a, b). Since then (12-month follow-up at the time of writing), the patient has been doing well and has reported no complaints.

DISCUSSION

Since there is no finding suggesting a congenital anomaly such as pulmonary atresia with ventricular septal defect, scimitar syndrome, pulmonary arteriovenous malformation, and bronchopulmonary sequestration (1–5), there remains three possibilities that would correspond to the etiology of the shunt in this patient, and they are

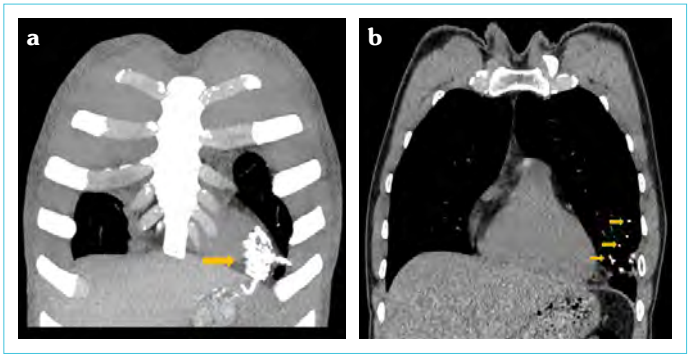

Figure 3. (a) Control chest CT, coronal minimal-intensity projection (MIP). The cast of embolic material filling the tangled vessels is seen (horizontal arrow). (b) Control chest CT, coronal reformation in mediastinal window. Note the dispersed linear opacities within the lingula, representing nontarget emboli (horizontal arrows)

as follows: [1] aberrant systemic arterial supply to the normal lung (ASANL), [2] pseudosequestration, and [3] fistulous connection.

Aberrant supply from the descending aorta in the left lower lobe with normal bronchial tree is one common finding of ASANL. With this abnormality, there may be a normal pulmonary supply to the involved parenchyma or may be absent (4). In contrast, in our case, the lingula was the involved segment, and the systemic supply was not from the descending aorta. Although both IMAs have been shown to be the source of systemic supply in several cases (4), neither the right nor the left IMA has not been the sole source. Hence, the patient cannot be considered to have ASANL.

It has been presumed that pleural adhesions subsequent to chronic inflammation may trigger the development of transpleural S-PA anastomoses by activating neovascularization from the systemic circulation (1). At the end of this process, chronically inflamed lung tissue turns into a mass of scar tissue, or the so-called pseudosequestration, that demonstrates peripheral contrast enhancement, with a tangle of anastomotic vessels supplied by a hypertrophied systemic artery, e.g., intercostal arteries, IMAs, inferior phrenic arteries (1, 4). In our case, the absence of such finding excluded the diagnosis of pseudosequestration. However, there was an emphysematous lesion suggestive of a previous inflammatory process. This process maybe the reason for fistula development or maybe a sequela of pre-existing fistula that facilitates infections owing to bacterial seeding (6, 7). It is hard to conclude which one is the causative of the other. In fact, congenital S-PA fistulas in adults, which often result from dilatation of normal precapillary or capillary anastomoses found in the normal lung due to genetic or external influences, and radiologically apparent fistulas without acquired etiology such as infection, trauma, surgery, and neoplasm are very rare (1, 2, 5–10). Nevertheless, the possibility of the fistula being congenital cannot be excluded because the majority of these abnormalities are congenital despite everything (5, 7).

Since the clinical course of S-PA fistulas cannot be fully predicted, the precise indication for treatment remains to be unknown. Considering the potential complications such as infection, hemorrhage, and pulmonary hypertension (2–10), it seems more favorable to close an S-PA fistula. With this thought, we decided to close the fistula by embolization. The calibration of the relevant pulmonary vein was normal, and the fistula was fed by numerous fine interconnecting channels, inferring a relatively low flow beyond the fistula. Hence, it was considered that glue injection alone would be sufficient for closure. Although a nontarget pulmonary embolization was noted to occur to a small extent, as expected, it did not affect the patient's overall condition. However, it would be better if we could have occluded the anastomotic part of the involved pulmonary artery using coils.

In conclusion, the clinical course of S-PA fistulas may be insidious and prolonged. Considering the potential complications, it seems more favorable to close such fistulas, although the precise indication for treatment remains controversial. For a safe and effective treatment, the flow dynamics should be considered when choosing the method of embolization.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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