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Case Report

Systemic Artery-to-Pulmonary Artery Fistula Mimics Pulmonary Embolus

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ABSTRACT

Systemic artery-to-pulmonary artery fistula (SA-PAF) is a rare phenomenon that can resemble a filling defect on computed tomography angiography (CTA). SA-PAF can be due to congenital or acquired etiologies and can alter the hemodynamics of the pulmonary circulation, with the most serious reported complication being hemoptysis, requiring embolization. We describe a case of an unusual SA-PAF between the right inferior phrenic artery and the right lower lobe pulmonary artery that mimicked an unprovoked pulmonary embolus (PE) on standard CTA in a patient with cardiomyopathy. This SA-PAF was interpreted on CTA as PE due to the presence of a filling defect, revealing that not all filling defects are PE. SA-PAF should always be considered when the clinical context or the imaging findings are atypical, specifically with an isolated filling defect visualized in the inferior lower lobe pulmonary artery. The false-positive PE was the result of mixing of systemic non-opacified blood with opacified pulmonary arterial blood.

Keywords: Fistula, Pulmonary artery, Inferior phrenic artery, Pulmonary embolus, Arterial malformation

INTRODUCTION

Systemic artery-to-pulmonary artery fistula (SA-PAF) is a rare entity. Patients with this vascular anomaly are typically asymptomatic; however, serious complications can occur, such as hemoptysis, pulmonary hypertension, congestive heart failure, and chronic pulmonary infection from bacterial seeding.[1] The vast majority of reported cases involve the use of transcatheter embolization or surgery for life-threatening hemoptysis.[2-4] This vascular anomaly is typically diagnosed with conventional angiography; however, it can be identified with computed tomography angiography (CTA). SA-PAF can appear as an abnormal pulmonary artery filling defect on CTA, similar to an arteriovenous malformation or a pulmonary embolus (PE), resulting in a misdiagnosis.[1,4] CTA can be utilized to further investigate the cause of abnormal filling patterns.[1]

SA-PAF can be congenital or acquired. Acquired causes include chest trauma, thoracic surgery, chronic thromboembolic disease, malignancy, and infections.[5-8] Congenital etiologies are rarer in the general population in the absence of risk factors.[2] SA-PAF can alter the hemodynamics of pulmonary arterial circulation presenting with the aforementioned complications.

We present a case of SA-PAF between the right inferior phrenic artery and the right lower lobe (RLL) pulmonary artery, originally misdiagnosed as PE. The patient underwent conventional angiography due to the presence of hypertrophied arteries seen on CTA within the right lung base and upper abdomen.

CASE REPORT

A 59-year-old woman with a history of non-ischemic dilated cardiomyopathy was referred to our facility for further investigation of an unprovoked PE beginning 2 years prior. A small filling defect diagnosed as PE was reported within the RLL pulmonary artery on CTA imaging 2 years before presentation at our institution [Figure 1a]. Her only symptom at the time of diagnosis was dyspnea. Outside testing for thrombophilia was unrevealing. She was treated with anticoagulation for 6 months following the diagnosis of PE on CTA. She denied smoking, hemoptysis, headache, gastrointestinal bleeding, history of stroke or cancer, chest trauma, recent surgery, chronic lung infections, or long-distance travel. Vital signs, physical examination, and laboratory results were unremarkable. The patient presented at our institution 2 years following original diagnosis of PE. CTA at our facility was less convincing in demonstrating RLL pulmonary artery filling defect; however, hypertrophied arteries in the right lung base [Figure 1b] and upper abdomen [Figure 1c] were identified. The hypertrophied arterial structure in the right lung base was present on the CTA from 2 years prior. Our current CTA interpretation described no evidence for PE but suggested the possibility of a pulmonary arteriovenous malformation. In fact, careful retrospective evaluation of the CTA from our facility reveals the fact that hypertrophied arteries are present in both the thorax and the abdomen. This combination is highly suggestive of SA-PAF instead of a simple pulmonary arteriovenous malformation limited to the thorax. Regardless, conventional angiography is indicated and definitive for either diagnosis on CTA.

The patient underwent conventional angiography for further investigation. Abdominal aortogram demonstrated a hypertrophied artery originating from the right renal artery communicating with RLL pulmonary artery. The right renal angiography confirmed a hypertrophied right inferior phrenic artery [Figure 1d]. Selective arteriogram of the right inferior phrenic artery confirmed flow to the inferior RLL pulmonary artery [Figure 1e].

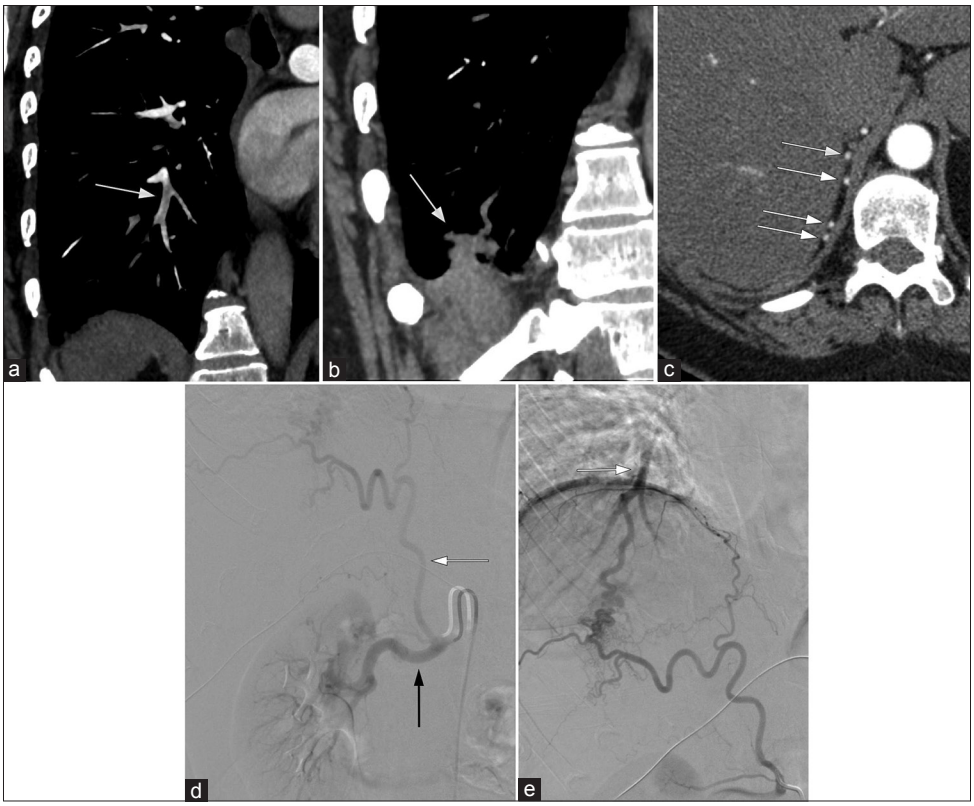

Figure 1: (a) A 59-year-old female with a history of non-ischemic dilated cardiomyopathy and symptoms of dyspnea diagnosed with pulmonary embolus on computed tomography angiography (CTA) 2 years prior. CTA coronal oblique reconstruction demonstrates an isolated linear defect in the right lower lobe pulmonary artery (white arrow). (b) Computed tomography (CT) angiogram demonstrates a hypertrophied arterial structure in the right lung base consistent with a vascular abnormality (white arrow). (c) CT angiogram demonstrates serpiginous hypertrophied arteries in the upper abdomen between the diaphragm and liver (white arrow). (d) AP digital subtraction right renal arteriogram demonstrates a hypertrophied right inferior phrenic artery (white arrow) originating from the right renal artery (black arrow). (e) Selective right inferior phrenic arteriogram demonstrates opacification of the inferior RLL pulmonary artery (white arrow).

Pulmonary angiography did not demonstrate a PE or pulmonary arteriovenous malformation; however, competitive inflow was seen within the inferior RLL pulmonary artery due to SA-PAF. The right pulmonary arterial pressures were mildly elevated. Given the absence of pulmonary arteriovenous malformation, PE, or significant pulmonary hypertension, a conservative approach was elected with the intent to have the patient return for embolization of SA-PAF if symptoms developed.

DISCUSSION

The most common cause of a filling defect within the pulmonary arteries is PE. In the setting of atypical presentations and imaging features, other etiologies should be considered before initiating anticoagulation. Respiratory motion and flow-related phenomenon can mimic pulmonary artery filling defects. Uncommon causes of a pulmonary artery filling defect include primary pulmonary artery sarcoma, tumor thrombus, and SA-PAF.[1] Atypical filling defects extending proximally within the pulmonary artery in the background of chronic inflammatory and fibrotic lung disease or hemorrhage and infarction should raise a concern for SA-PAF on CTA.[1] If SA-PAF is suspected or considered based on the clinical history or initial CT findings, delayed contrast-enhanced CT imaging can be helpful. If the pulmonary artery filling defect disappears with delayed imaging, it is indicative that a fistula is producing only an arterial phase filling defect. Delayed imaging demonstrates homogenous pulmonary artery enhancement as the systemic arterial component enhances with contrast overtime. This would never be the case with a true PE. Conventional angiography is also helpful and definitive for the diagnosis of SA-PAF; however, CTA is commonly used to diagnosis suspected PE and guide a treatment plan.[1]

Patients with SA-PAF have a wide range of clinical presentations including hemoptysis, pulmonary hypertension, congestive heart failure, and infection due to bacterial seeding. The majority of SA-PAFs are asymptomatic at the time of diagnosis, and no definitive treatment guidelines have been established. Some reports recommend prophylactic treatment due to risk of future complications, although the topic is debatable.[2] At present, the preferred treatment modality for symptomatic patients is transarterial embolization, particularly for the immediate control of hemoptysis.[2,3]

The etiology of SA-PAF is hypothesized to result when the pulmonary arterial circulation is altered through chronic inflammation, hypoxic vasoconstriction, or arterial thrombosis. Proliferation of adjacent bronchial and systemic arteries ensues.[7] The most common systemic artery to contribute to SA-PAF is a bronchial artery, with 90% of reported cases presenting with hemoptysis. Less than 10% of cases reported in literature involve the inferior phrenic artery.[3] The inferior phrenic artery should be suspected as an inflow vessel to SA-PAF when SA-PAF involves the lung base. In our case, the right inferior phrenic artery originated from the right renal artery [Figure 1d], which occurs in 10.5%–15.4% of reported cases.[9,10]

The etiology of SA-PAF in our case is indeterminate. The patient lacked typical risk factors for an acquired SA-PAF, such as chronic pulmonary infections, chronic lung disease, thoracic trauma or surgery, and malignancy. The only pertinent medical history was non-ischemic dilated cardiomyopathy. At the time of evaluation, the patient's echocardiogram revealed an ejection fraction of 53%, without signs of the right-sided heart failure or pulmonary hypertension. This SA-PAF was originally interpreted on CTA as PE [Figure 1a] due to the presence of an isolated arterial phase pulmonary artery filling defect. The false-positive PE was the result of mixing of systemic non-opacified blood with opacified pulmonary arterial blood from SA-PAF. This mimicked a filling defect within RLL pulmonary artery, and SA-PAF arteries in the peripheral RLL imitated an associated pulmonary infarct [Figure 1b]. Delayed contrast-enhanced imaging was not obtained before conventional angiography and this limited the diagnostic consideration of SA-PAF. In this case, the hypertrophied arteries in the thorax [Figure 1b] and abdomen [Figure 1c] were not described on CTA even though in retrospect they were present. In addition, subsequent CTA findings of a combination of hypertrophied arteries in both the thorax and abdomen should have led to a presumptive diagnosis of SA-PAF over consideration of a pulmonary AVM. Arteriography of the right inferior pulmonary artery and right pulmonary arteries confirmed SA-PAF [Figure 1e].

CONCLUSION

SA-PAF should be considered when the clinical context and the imaging findings are atypical for PE. Delayed contrast-enhanced CTA imaging, recognition of hypertrophied arteries in both the thorax and abdomen, and conventional angiography can be utilized to make the correct diagnosis of SA-PAF.

Declaration of patient consent

The authors certify that they have obtained patient consent. The patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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