Acquired transpleural systemic artery-to-pulmonary artery communication mimicking a pulmonary arteriovenous malformation and causing a false-positive diagnosis of a pulmonary embolus

Dear sir,

The diagnosis of pulmonary arteriovenous malformations (PAVMs) on CT is usually straightforward, but there are several 'mimics' that may cause confusion, the imaging features of which have been described previously [1]. A rare vascular mimic is an acquired transpleural systemic artery-to-pulmonary artery communication as described in this report.

Case history:

Institutional review board approval was not required for publication of this case report.

A 43-year-old man was referred for management of a lingular PAVM incidentally diagnosed on two CT pulmonary arteriograms (CTPA) performed to investigate episodes of left-sided pleuritic chest pain. Both of these examinations, which were performed a year apart, demonstrated a single linear filling defect extending from the left main pulmonary artery into an inferior lingular pulmonary arterial branch (**fig.1**); this was interpreted as a pulmonary embolus (PE). The second PE-positive study was performed whilst the patient was on anticoagulants and for this reason, together with a diagnosis of Protein C deficiency and a heterozygous prothrombin gene mutation, the patient was commenced on long-term anticoagulation.

There was no family or clinical history to suggest underlying HHT, and physical examination was unremarkable including normal average arterial oxygen saturations of 96%. CT imaging was reviewed and the imaging features of the vascular abnormality appeared to be consistent with a PAVM as there was a slightly dilated and tortuous pulmonary artery and an adjacent prominent pulmonary vein (**fig. 2**). The linear filling defect interpreted as thrombus involved the proximal portion of this abnormal

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pulmonary arterial branch. There were no other pulmonary parenchymal abnormalities. The patient was referred for pulmonary angiography with a view to embolization.

Angiography demonstrated flash filling of only the proximal portion of the prominent lingular pulmonary artery seen on CT without opacification of its distal portion (**fig. 3 and online supplement**) consistent with reversed flow due to the presence of a transpleural systemic arterial supply. This abnormal lingular artery was selectively catheterized distally: angiography confirmed reversed flow within this vessel (**fig. 4**), and demonstrated no direct communication with a pulmonary vein.

The first of the referral hospital CT studies showed that the transpleural systemic arterial supply to the lingular pulmonary artery was provided by hypertrophied branches of the left inferior phrenic artery (**fig. 5**) and the linear filling defect seen in the pulmonary artery was recognized as being an artefact due to a jet of unenhanced systemic blood rather than a PE. On subsequent questioning, the patient admitted to having suffered left-sided chest trauma as a teenager for which he required insertion of a chest drain. The final diagnosis was of an acquired systemic artery-to-pulmonary artery communication.

Discussion:

The importance of non-bronchial systemic arteries and their contribution to life-threatening hemoptysis in chronic lung disease is well-recognized. It is less well-known that focal transpleural collaterals may also develop in response to surgical or accidental trauma, or at sites of localized pleural scarring caused by adjacent pulmonary inflammatory disease. Although uncommon, they are being increasingly incidentally recognized on CT and are an important mimic of PAVMs.

Systemic bronchial artery-to-pulmonary artery anastomoses are present in normal lung and these will increase in size in response to a number of stimuli including chronic inflammatory lung disease and pulmonary emboli. In severe, longstanding inflammation as occurs with chronic TB and/or bronchiectasis, these will result in reversal of pulmonary arterial flow in the diseased portion of lung. Such reversal of flow is best appreciated on selective bronchial arteriography [2] but may also be recognized on CTPA. This has been described as mimicking pulmonary embolic disease in two case

reports; one of a patient with chronic cavitary tuberculosis complicated by an aspergilloma and the other with bronchiectasis secondary to childhood pertussis [3, 4] . To our knowledge, this has not been previously reported in a focal lesion as described here.

Non-bronchial systemic arteries, including anterior and posterior intercostal arteries, inferior phrenic arteries and thoracic branches of subclavian and axillary arteries, may also provide a collateral supply to normal peripheral pulmonary arteries, but such anastomoses will only develop through an area of diseased pleura.

To date, there is no consensus regarding the management of focal transpleural systemic artery-to-pulmonary artery communications that have developed in response to localized pleural inflammation or trauma. Although reversal of pulmonary arterial flow may be dramatic, hemoptysis is rare. This is unlike similar degrees of systemic artery-to-pulmonary artery shunting seen in areas of chronically diseased lung, which are more commonly complicated by hemorrhage. It is the authors' opinion, therefore, that these focal transpleural shunts do not require treatment in the absence of bleeding complications.

The case presented here is of a transpleural systemic-to-pulmonary artery shunt mimicking a PAVM and resulting in the false positive diagnosis of a pulmonary embolus due to the reversal of blood flow within a pulmonary artery branch.

References:

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2. Hutchin P, Terzi RG, Peters RM. Bronchial-pulmonary artery reverse flow. Angiographic demonstration in bronchiectasis. Ann Thorac Surg. 1967;4(5):391-8.
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Declaration:

No conflict of interest.

The patient has given his written informed consent to the publication of this case report.

Figure Legend:

Figure 1 Coronal (a) and axial (b) images from a CTPA examination demonstrating an apparent low-density filling defect extending from the inferior lingular pulmonary arterial branch into the left main pulmonary artery mimicking a PE.

Figure 2 a. Axial image from a CTPA examination demonstrating a small tortuous inferior lingular pulmonary arterial branch. b. Reconstructed maximum intensity projection (MiP) sagittal image from the CTPA examination demonstrating a dilated, tortuous inferior lingular pulmonary artery branch with an apparent draining pulmonary vein.

Figure 3 Single image from left pulmonary arteriography demonstrating flash filling of the proximal part of an inferior lingular pulmonary arterial branch (arrow).

Figure 4 Selective pulmonary arteriography of the hypertrophic inferior lingular pulmonary artery branch which demonstrated reversed flow but no arteriovenous communication.

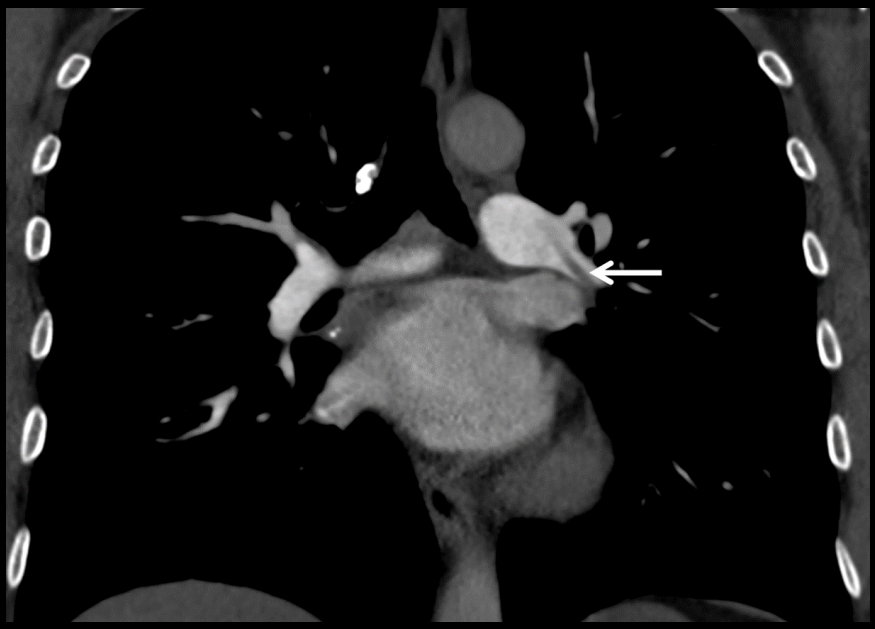
Figure 5 Axial MiP image from the original CTPA examination demonstrating an hypertrophied left inferior phrenic artery (white arrow) which provided the major systemic arterial supply to the dilated inferior lingular pulmonary artery (white arrowhead).

Online Supplement:

Video of a left pulmonary arteriogram performed in a right anterior oblique projection demonstrates flash filling of the proximal portion of an inferior lingular pulmonary arterial branch on a single image (compare with figure 3), which then clears on the subsequent images due to the presence of a transpleural systemic arterial supply to this vessel.

Figure 1

a



b

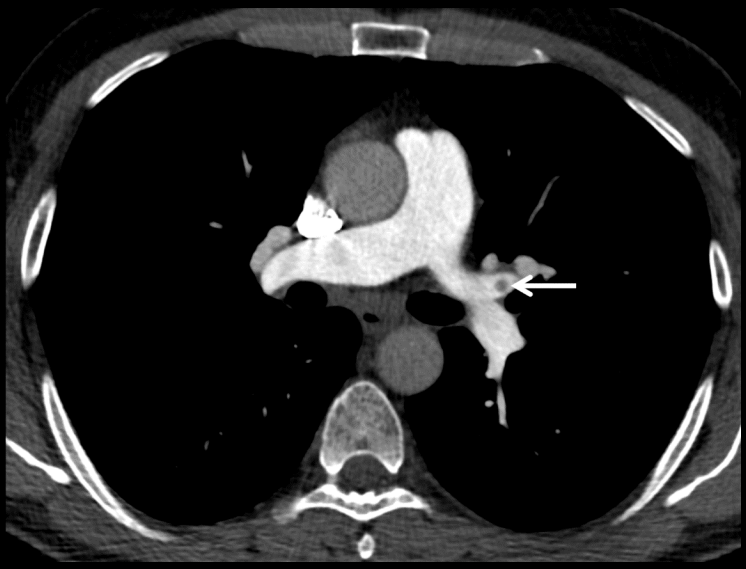


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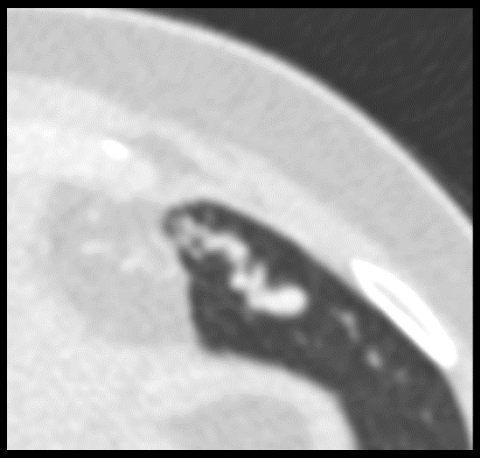
Figure 2

a



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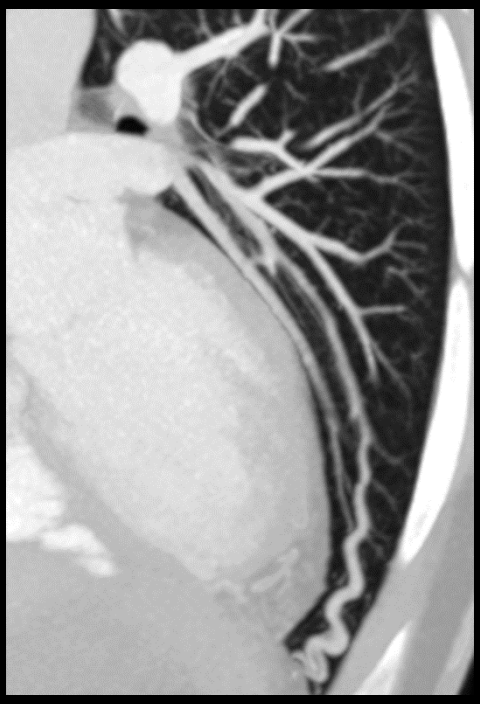

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Figure 3

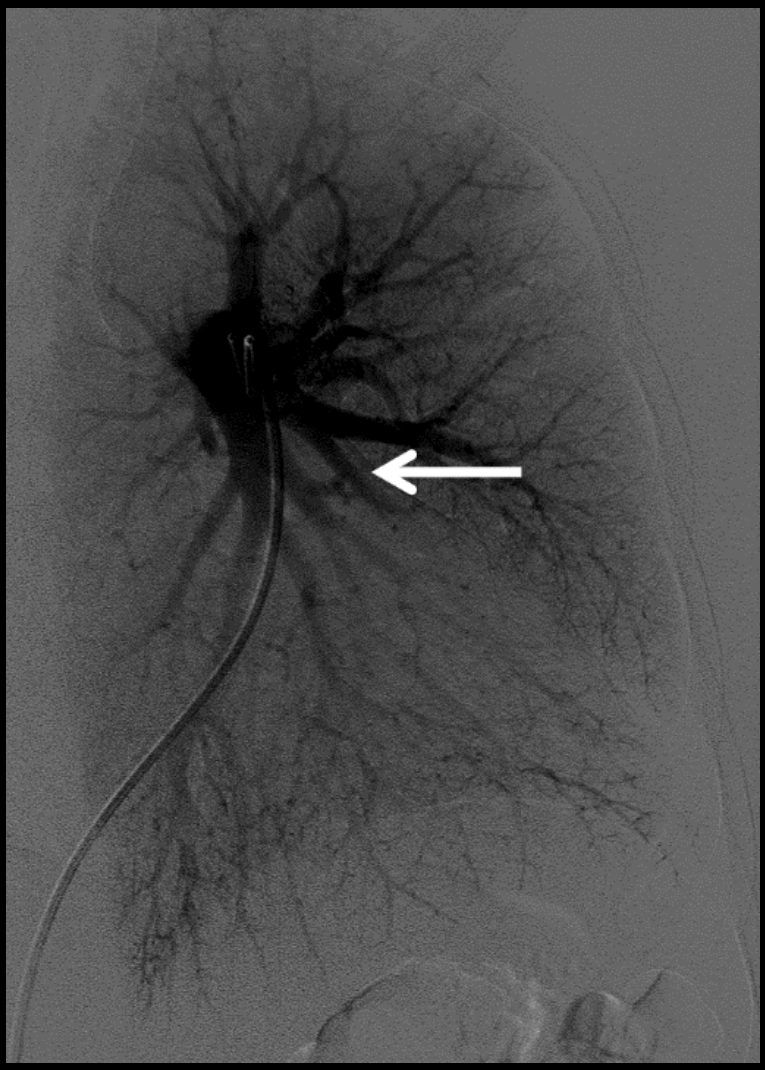

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Figure 4

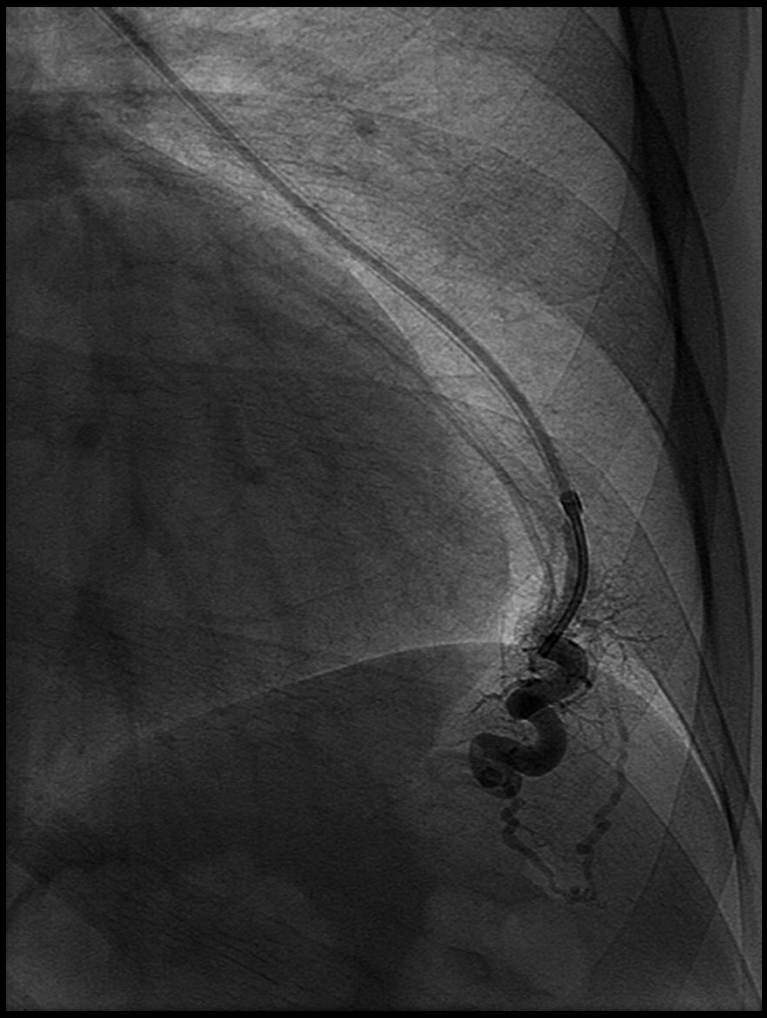

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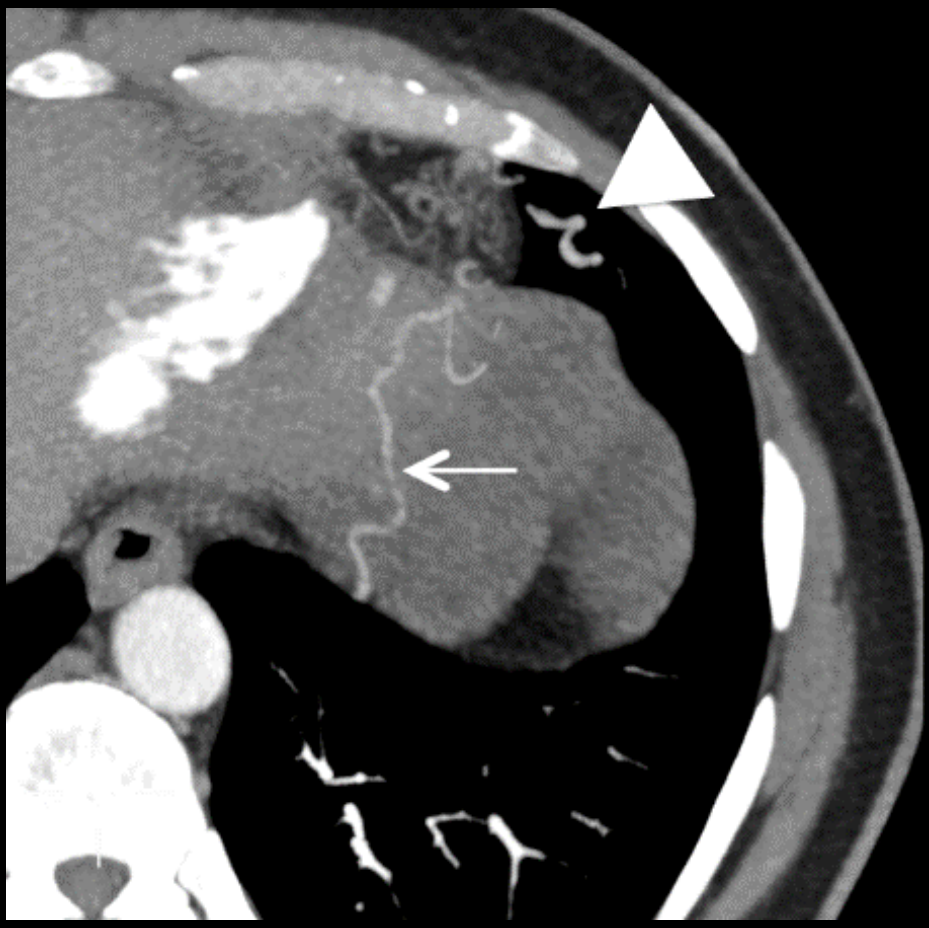
Figure 5



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