Systemic Artery to Pulmonary Artery Shunts: A rare mimicker of Pulmonary AVMs

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PURPOSE:

To describe the presentation, imaging appearance and management of systemic artery-to-pulmonary artery shunts.

METHODS:

An imaging database search for systemic artery to pulmonary artery shunts , between January 2007 and February 2022, was performed. This was defined as CT and angiographic evidence of systemic artery to pulmonary artery communications resulting in retrograde flow in discrete pulmonary arteries. Pulmonary AVMs with post-embolization systemic artery collaterals, chronic lung disease patients with only micro-fistulous shunting, and CTEPH patients with systemic collateral flow were excluded. A descriptive analysis was then performed.

BACKGROUND:

Systemic arterial shunting in the setting of chronic lung disease and chronic thromboembolic pulmonary hypertension is well established and understood to be caused by dilation of collateral arteries, primarily of bronchial artery origin. However, unlike these bronchial artery-to-pulmonary artery shunts, the transpleural-supplying vessels in transpleural systemic artery to pulmonary artery shunts is hypothesized to represent angiogenesis, rather than dilation of collateral vessels.[2] Transpleural systemic artery to pulmonary artery shunts may be congenital but more often occur in patients with history of thoracic surgery or trauma and in most cases without a history of chronic inflammatory lung disease[3, 2]. Perhaps most significantly, transpleural systemic artery to pulmonary artery shunts may superficially mimic the appearance of classic PAVMs[4]. However, unlike classic PAVMs, indications for treatment of transpleural systemic artery to pulmonary artery shunts is unknown and there is no association with HHT. It is therefore important to understand the imaging characteristics of these lesions to avoid mismanagement.

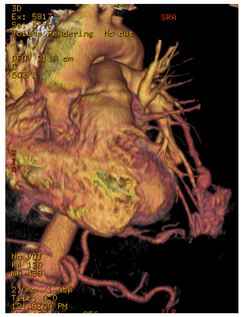
IMAGING CHARACTERISTICS

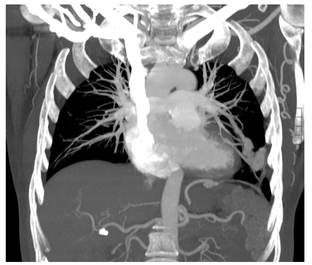
Retrospectively, all cases for which CT was obtained (7/8) could be diagnosed with the following features:

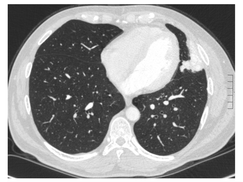
I - Tubular structure communicating with the pulmonary arteries; II – Hypertrophy of systemic arteries in the chest wall and diaphragm, greater than expected with chronic lung disease. III – Absence of associated chronic lung parenchyma disease (i.e., ground glass, fibrosis). Figures 1-3 demonstrate CT findings in a typical transpleural Systemic Artery to Pulmonary Artery Shunt.

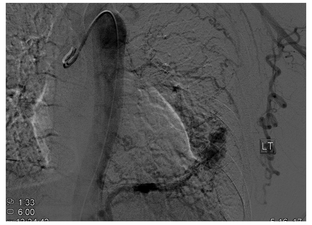
Angiographically, all cases (8/8) demonstrated:

I - hypertrophy of systemic arteries; II – high-flow shunting from the systemic arteries into pulmonary arteries; III – A more “nidus” like appearance than the less discrete blushing seen with chronic inflammatory lung disease. Figures 4 and 5 demonstrate angiographic findings in a typical Transpleural Systemic Artery to Pulmonary Artery Shunt.









PATIENT CHARACTERISTICS

Patient Number	Sex	Age	Etiology	Clinical presentation	Co-morbidities	History of HHT or other AVMs	Family History of AVM	Malformation Type	Outflow	Feeding vessels
1	F	57	Trauma	Incidental	Mitral valve prolapse	None	HTN, Asthma	Complex, Nidus	Pulmonary artery subsegmental and pulmonary venous	Internal thoracic, bronchial
2	M	54	Congenital	Incidental	Nasopharyngeal carcinoma	None	Nasopharynx carcinoma	Complex, Nidus	Left lingular/Left lobar artery	Inferior phrenic, internal thoracic/intercostal and long thoracic
3	M	70	Congenital	Incidental	COPD, SVT, Raynaud's syndrome	None	HTN, COPD, Raynaud	Complex, Nidus	Pulmonary artery segmental	Descending thoracic aorta and inferior phrenic
4	M	10	Iatrogenic	Incidental	Tricuspid atresia, Protein losing enteropathy	Protein losing enteropathy	Congenital heart disease	Complex, Nidus	Left lobar arteries	Posterior intercostal, thoracoacromial, internal thoracic, costocervical, and inferior phrenic
5	F	44	Iatrogenic	Incidental	Pulmonary cystic-bullous disease	None	Pulmonary cystic-bullous disease	Complex, Nidus	Lingular subsegmental artery	Long thoracic, intercostal branches
6	M	53	Iatrogenic	Incidental	Lymphoma, Paroxysmal Afib	None	Esophageal rupture 2/2 Lymphoma, Afib	Complex, Nidus	Pulmonary artery, lingular branch	Inferior phrenic, internal thoracic
7	F	26	Iatrogenic	Incidental	Biliary dyskinesia, Pulmonary atresia	None	Biliary dyskinesia, Congenital heart disease	Complex, Nidus	Pulmonary artery, left apical segmental, & segmental venous	Inferior phrenic, bronchial, splenic
8	M	73	Iatrogenic	Incidental	Bicuspid aortic valve, complete heart block	None	None	Complex, Nidus	Subsegmental pulmonary A, RUL	Inferior phrenic, bronchial

DESCRIPTION OF IMAGES:

***Top Left**: 3D Rendering of a T-SAPAS, demonstrating a nidus-like malformation feeding into a branch of the left pulmonary artery. A hypertrophied phrenic artery is also seen, which was subsequently shown to be feeding into the shunt.*

***Top Right**: Coronal MIP of the chest demonstrates tubular soft tissue density in the LLL, hypertrophy of the long thoracic, and hypertrophy of the inferior phrenic artery which is seen crossing the diaphragm.*

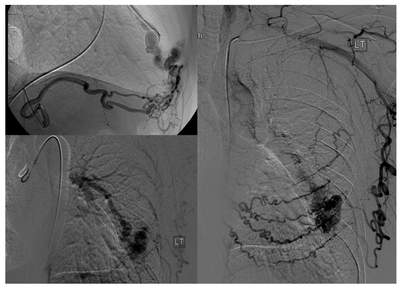
***Bottom Left**: Tubular soft tissue density seen in the LLL. On lung window the surrounding lung parenchyma demonstrates no evidence of inflammatory lung disease.*

***Bottom Right**: Aortogram showing hypertrophy of the left long thoracic, intercostal arteries and inferior phrenic artery, which feed into a “nidus-like” pulmonary artery shunt.*

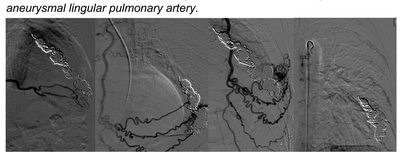
RESULTS

A total of eight patients with systemic artery-to-pulmonary artery malformations were identified. Etiology was presumed secondary to thoracic surgery in five cases (62.5%), trauma in one case (12.5%), and congenital in two (25.0%). Seven patients (87.5%) were referred for incidentally discovered lesions on CT, five diagnosed as pulmonary AVMs (62.5%). CTs demonstrated hypertrophied peripheral vessels with only pulmonary arterial communication. In all cases, CTs underestimated the complexity of the lesion and the number of vessels involved.

Angiography revealed all eight cases (100%) to involve several (in some cases numerous) dysplastic systemic arteries discretely filling multiple (in some cases numerous) pulmonary arteries. Pulmonary arterial outflow ranged from subsegmental to lobar. Systemic arterial inflow was wide ranging. The average mean arterial pressure was 16.1 ± 4.2 mm Hg. Two patients (25%) had pulmonary hypertension (greater than 20 mm Hg). Coil embolization was performed in seven cases. In four cases (57.1%) embolization of the outflow pulmonary artery was performed. There were no major adverse events. All patients remained asymptomatic at follow-up, and there were no episodes of pulmonary hemorrhage noted.



Patient number 2, presenting with a large complex transpleural systemic artery to pulmonary artery shunt. Aortogram (bottom left), superselection of the left inferior phrenic artery (top left), and selection of the left proximal subclavian artery (right) demonstrate dominant supply from the left inferior phrenic, internal mammary and long thoracic arteries, shunting into an aneurysmal lingular pulmonary artery.



Patient number 2, now status post coil embolization of the aneurysmal lingular pulmonary artery. Selection of the inferior phrenic, internal mammary, and long thoracic arteries (left to right) demonstrate no contrast extending beyond the coiled lingular pulmonary artery. Aortogram on delayed imaging (far right) demonstrates no retrograde flow into the pulmonary arterial system.

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DISCUSSION OF MANAGEMENT

The need for treatment in transpleural systemic artery to pulmonary artery shunts remains unknown. Given the theoretical risk of pulmonary hypertension and bleeding, all eight patients were embolized, primarily with coil embolization by targeting the outflow pulmonary artery (in four cases) and inflow systemic arteries (in three cases). Figures 6a and 6b demonstrate coiling of the pulmonary artery. Pulmonary pressures were checked, with two patients (25%) having pulmonary artery pressures between 20-25 mm Hg, meeting criteria for pulmonary hypertension according to the newest criteria[6]. However, in at least one of these cases this may have been confounded by COPD. Additionally, patients may be at risk for segmental pulmonary hypertension due to the different pulmonary inflow sources. Although, segmental pressures were not checked, this could potentially be studied in the future. While it is unclear what the management of transpleural systemic artery to pulmonary artery shunts should be at this time, it is clear that their natural history, management, and clinical implications differ from Pulmonary AVMs. As such it is important to be able to identify these lesions on CT and angiography. Whereas Pulmonary AVM patients should be referred into the HHT pathway, transpleural systemic artery to pulmonary artery shunts do not require genetic testing and may be able to be observed without intervention. Increasing awareness of transpleural systemic artery to pulmonary artery shunts may lead to more cases being identified and a greater understanding of the ideal management.