The Boy's Not All Right

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Case

- Adolecent patient presenting with massive hemoptysis

Differential Diagnosis

- Rheumatologic disorders:
  - Granulomatosis and polyangiitis, anti-GBM disease, SLE, microscopic polyangiits, pulmonary capillaritis
- Ateriovenous malformation
- Oncologic / Hematologic
- Tuberculosis and infectious
- Inhalant abuse
- Idiopathic pulmonary hemosiderosis
- Trauma
- Foreign Body
- Bronchiectasis
- CPAM
- Pulmonary hypertension
- Pulmonary embolus

Systemic artery to pulmonary vein arteriovenous malfomations (SP-AVM)

A case-based review of the literature

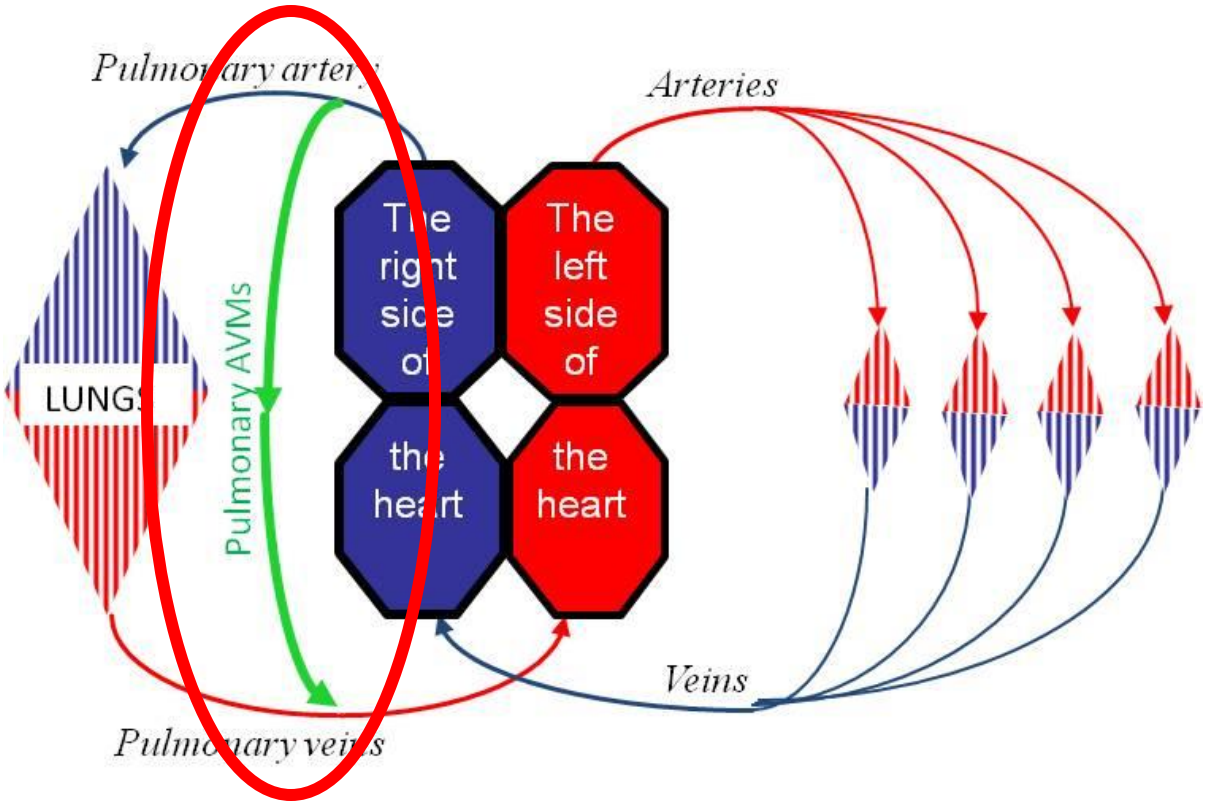
SP-AVM

- SP-AVMs extremely rare
- 3-4% of pulmonary AVMs
  - Dines[1] - 3/101
  - Bosher[2] - 12/350
- Systemic connections
  - Most frequently internal mammery arteries[3]
  - Anomalous aortic branches, bronchial, intercostal, lateral thoracic, esophageal

1) Dines DE, Seward JB, Bernatz PE. “Pulmonary arteriovenous fistulas”. Mayo Clin Proc. 1983 Mar;58(3):176-81
2) Bosher LH Jr, Blake DA, Byrd BR. “An analysis of the pathologic anatomy of pulmonary arteriovenous aneurysms with particular reference to the applicability of local excision”. Surgery. 1959 Jan;45(1):91-104
3) Pouwels HM, Janevski BK, Penn OC, Sie HT, ten Velde GP. “Systemic to pulmonary vascular malformation”. Eur Respir J. 1992 Nov;5(10):1288-91. Review

“Typical” Pulmonary AVM

- Pulmonary artery to pulomary vein
- Cyanotic



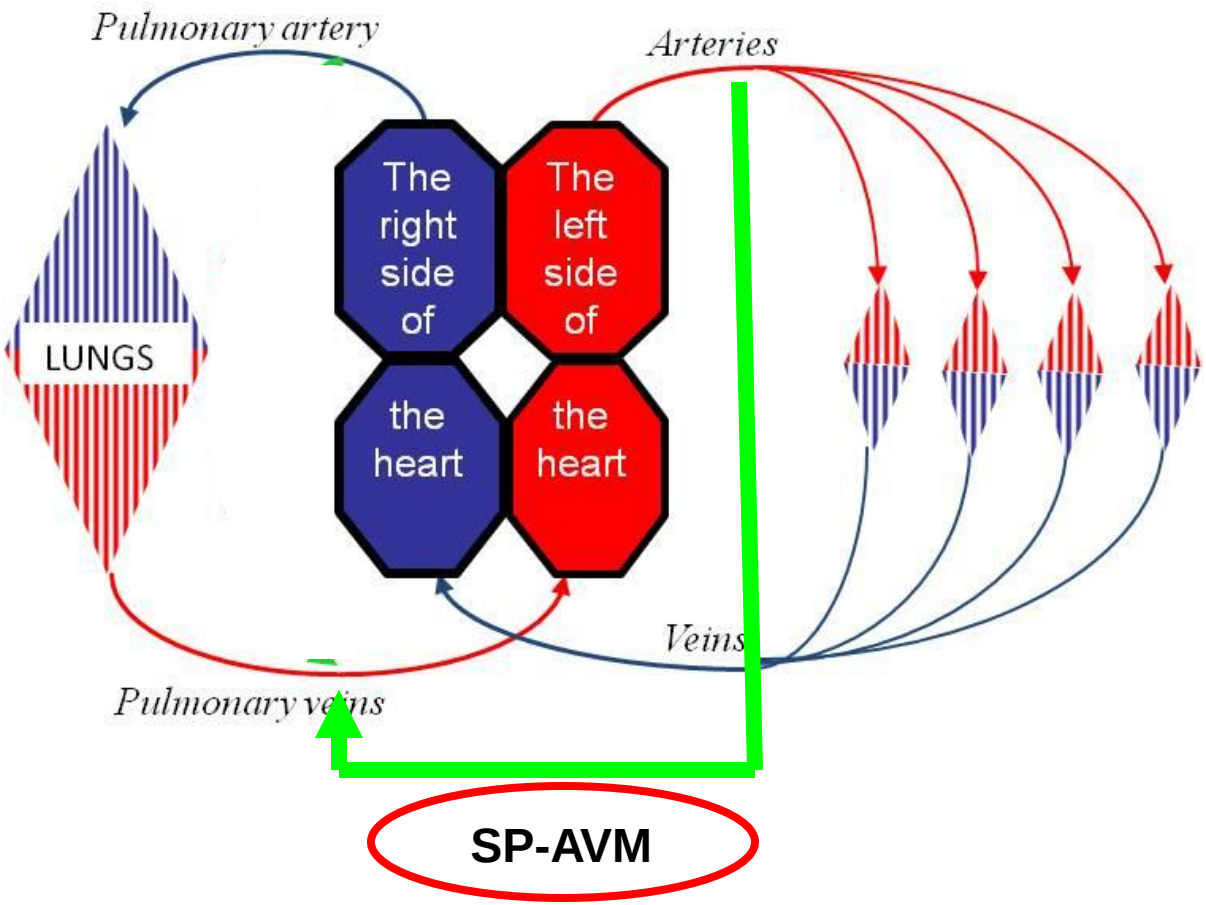
http://www1.imperial.ac.uk/resources/52F91CDC-2A31-43C7-B644-C79BF10E162D/pavms.jpg

Systemic to Pulmonary AVM

- Normal systemic artery to pulmonary vein

OR

- Aberrant aortic artery to pulmonary vein
- Acyanotic



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Difference Between Pulmonary AVM and SP-AVM

Pulmomary AVM	**Systemic to pulmonary AVM**
Female > Male	Male > Female
Related to HHT	No relationship
Right-to-left shunt	Left-to-left shunt
Single or multiple	Single
Local or diffuse	Local

3) Pouwels HM, Janevski BK, Penn OC, Sie HT, ten Velde GP. “Systemic to pulmonary vascular malformation”. Eur Respir J. 1992 Nov;5(10):1288-91. Review

SP-AVM - Presentations

- Not associated with hereditary hemorrhagic telangiectasia
  - No telangiectasia

- Does not present with classic triad of PAVM
  - Cyanosis
  - Clubbing
  - Desaturation

SP-AVM - Presentations

- Physical exam
  - Continuous systolic murmur
  - Bounding pulses
- Asymptomatic
  - Incidental finding
- Symptomatic
  - Heart failure
  - Hemoptysis

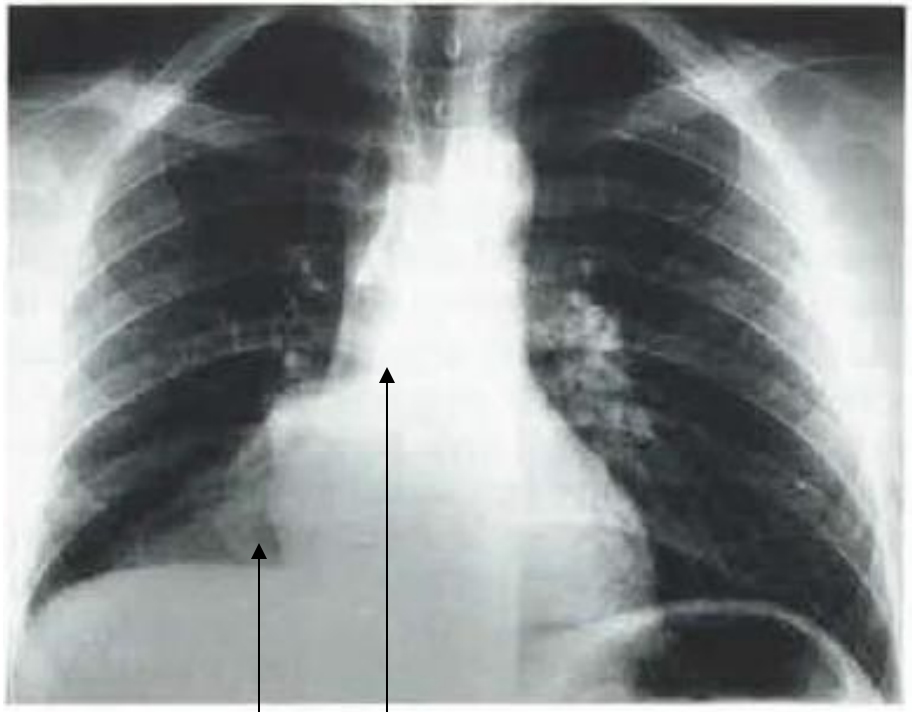
SP-AVM - Presentations

Chest radiographs

- Typically abnormal, often non-specific
- Single lesion
- May involve more than one lobe

Presentation – Hemoptysis

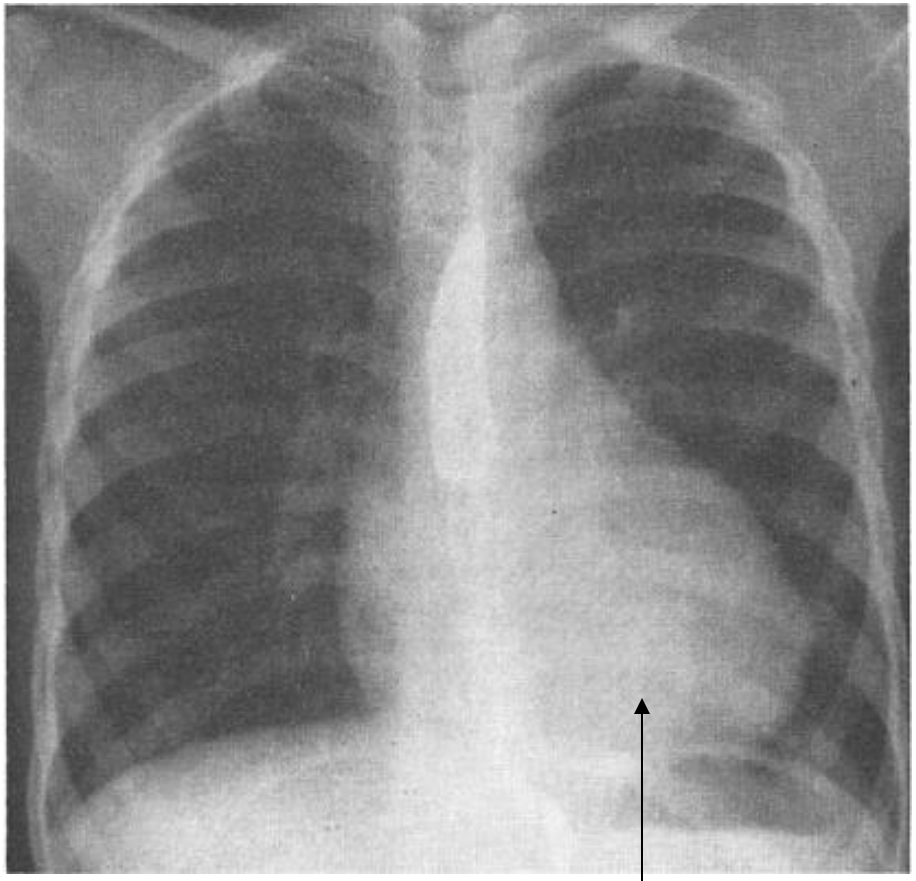
- 21 y.o. ♂
- Hemoptysis - 600cc upon awakening
- On exam:
  - Crackles to right base
  - No murmur
- Bronchoscopy – clots in RLL
- Diagnosis – Bronchial artery to pulmonary vein
- Treatment – bilobectomy



3) Pouwels HM, Janevski BK, Penn OC, Sie HT, ten Velde GP. “Systemic to pulmonary vascular malformation”. Eur Respir J. 1992 Nov;5(10):1288-91. Review

Presentation – Asymptomatic

- 6 y.o. ♀
- Continuous murmur, misdiagnosed as PDA
- Admitted for cardiac catheterization
- Hypertensive 130/60 with bounding pulses
- ECG: Left ventricular hypertrophy
- Diagnosis – SP-AVM, abnormal aortic branch
- Treatment – Left lower lobectomy, ligation of artery

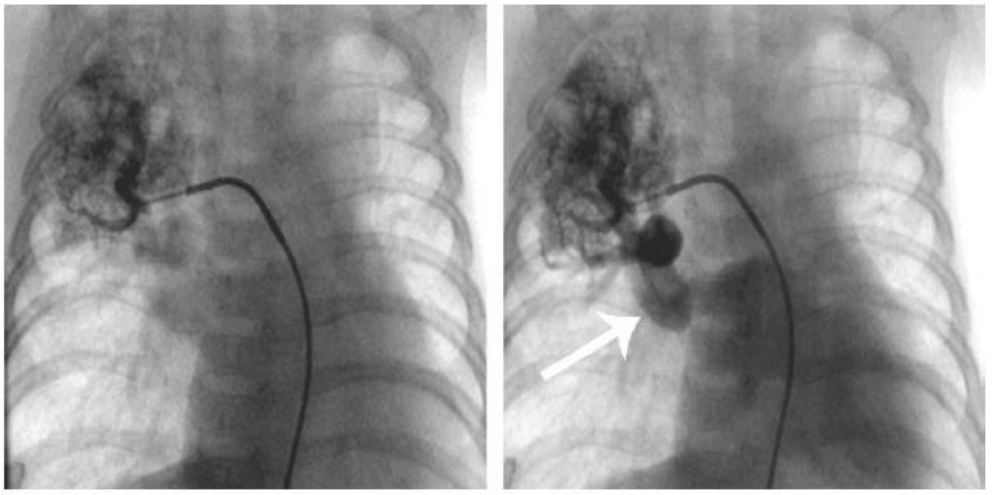


Increased density behind heart

5) Scott LP 3rd, Perry LW. “Systemic artery--pulmonary vein fistulas, congenital and acquired left to left shunt”. Am J Cardiol. 1969 Jun;23(6):872-6.

Presentation – Heart Failure

- Term neonate
- APGARS 5 at 1 min, 3 at 5 min
- Complicated neonatal course
  - Congestive heart failure, sepsis, mechanical ventilation, seizures, cerebral edema
- At 6 weeks
  - Congestive heart failure
  - Biphasic stridor
  - Tachypnea, Tachycardia
- Bronchoscopy – extramural vascular compression
- Diagnosis - 2 aberrant aortic vessel from descending aorta to pulmonary vein



a. **b.**

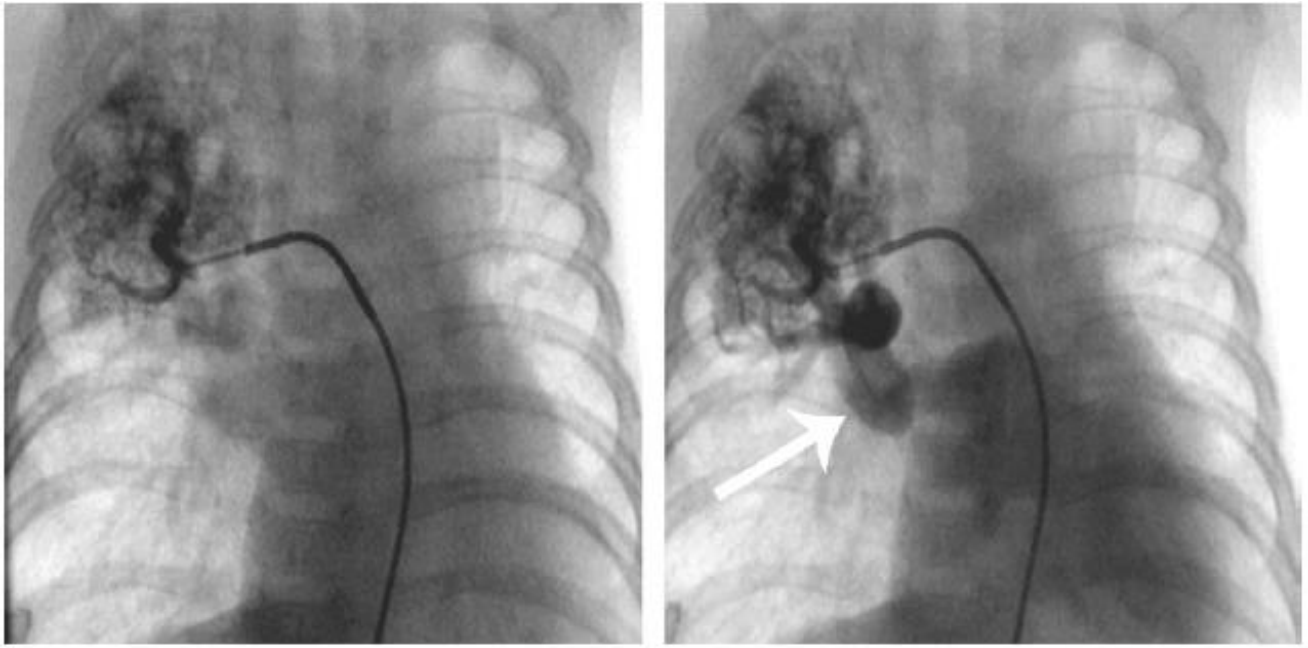
Figure 2. (**a**) Upper right lung lobe selective aberrant artery angiography. A "vascular sponge" in the upper right lung lobe is observed. (**b**) Enlarged upper right pulmonary vein is seen draining blood into the left atrium (*arrow*).

6) Kosutic J, Minic P, Sovtic A, Prijic S. "Upper lung lobe systemic artery-pulmonary vein fistula with signs and symptoms of congestive heart failure: successful treatment with coil embolization". J Vasc Interv Radiol. 2007 Feb;18(2):299-302.

SP-AVM - Diagnosis

Traditionally – selective angiography of systemic artery

- Contrast or bubble contrast
- Return to pulmonary vein, left atrium



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SP-AVM - Diagnosis

CT-angiography with 3D reconstruction

- Generally still confirmed with selective angiography

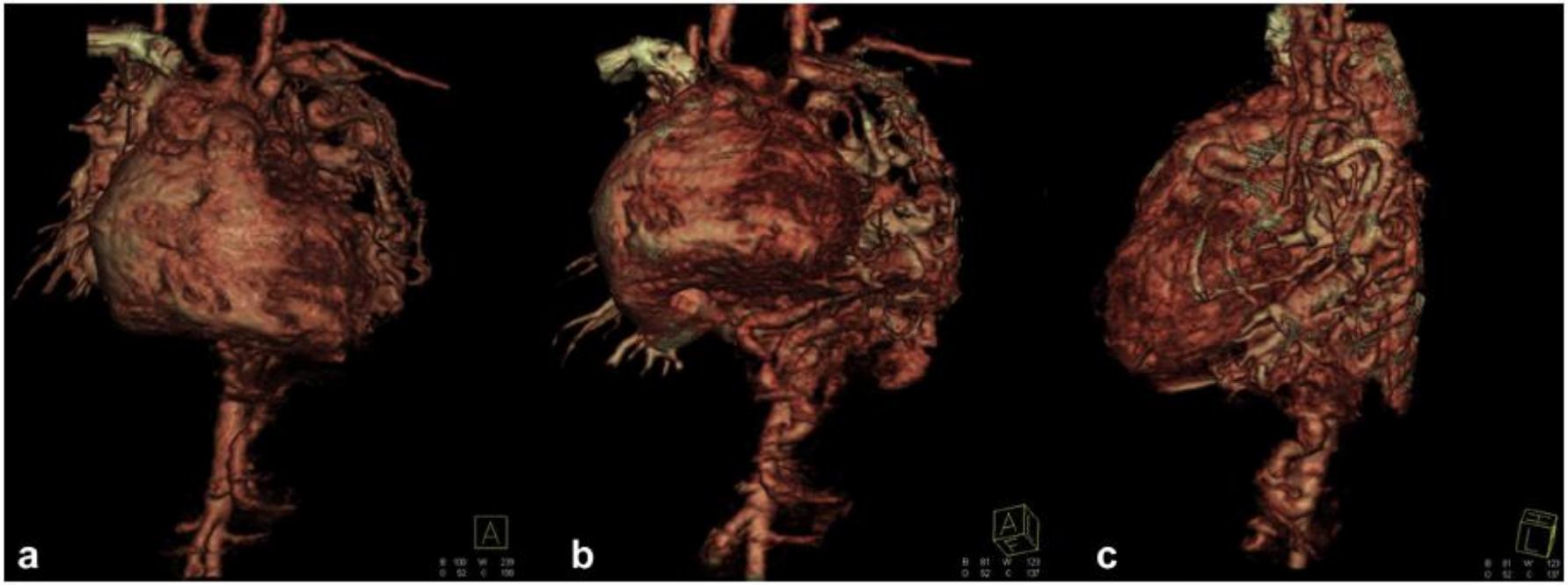


Fig. 2 – a–c: CT angiography with 3D volume rendering showing aberrant vessels originating from the left subclavian artery and abdominal aorta to the entire left lung which drained into the left atrium via normal pulmonary veins.

4) Jariwala P, Ramesh G, Sarat Chandra K. “Congenital anomalous/aberrant systemic artery to pulmonary venous fistula: closure with vascular plugs & coil embolization”. Indian Heart J. 2014 Jan-Feb;66(1):95-103. doi: 10.1016/j.ihj.2013.10.009. Review

SP-AVM Management

- Historically: Surgical
  - Surgical ligation
  - Lobectomy, only if lung damaged[7] or ligation not possible

- Interventional Radiology
  - Coil embolization
  - Vascular plug

Pernot C, Simon P, Hoeffel JC, Worms AM, Marcon F, Prevot J. “Systemic artery-pulmonary vein fistula without sequestration”. Pediatr Radiol. 1991;21(2):158-9.

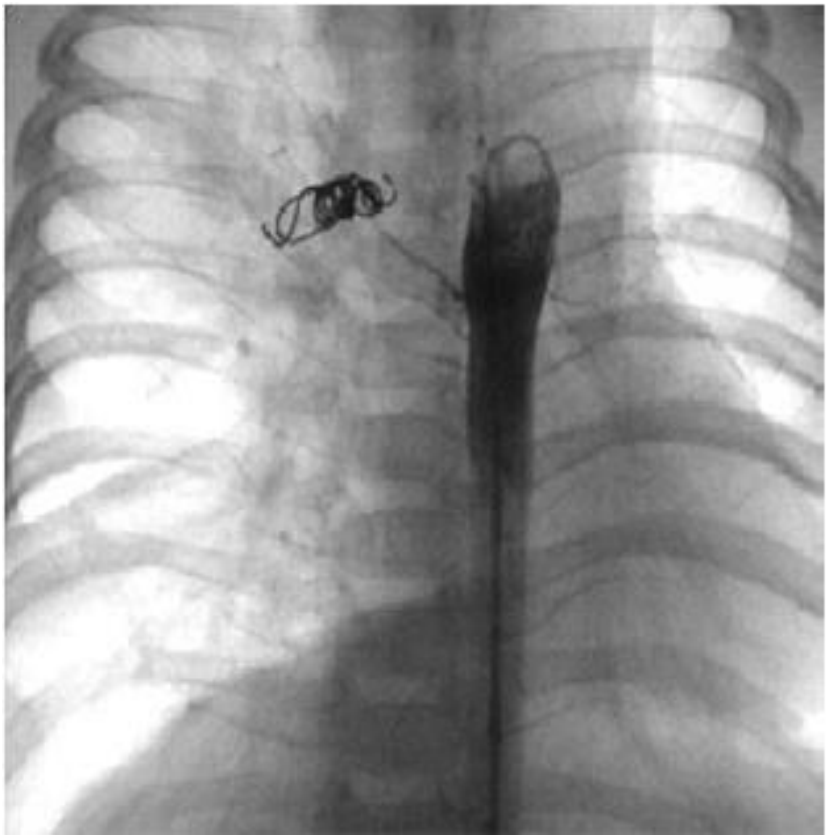
SP-AVM Management

A review of 21 cases of SP-AVM

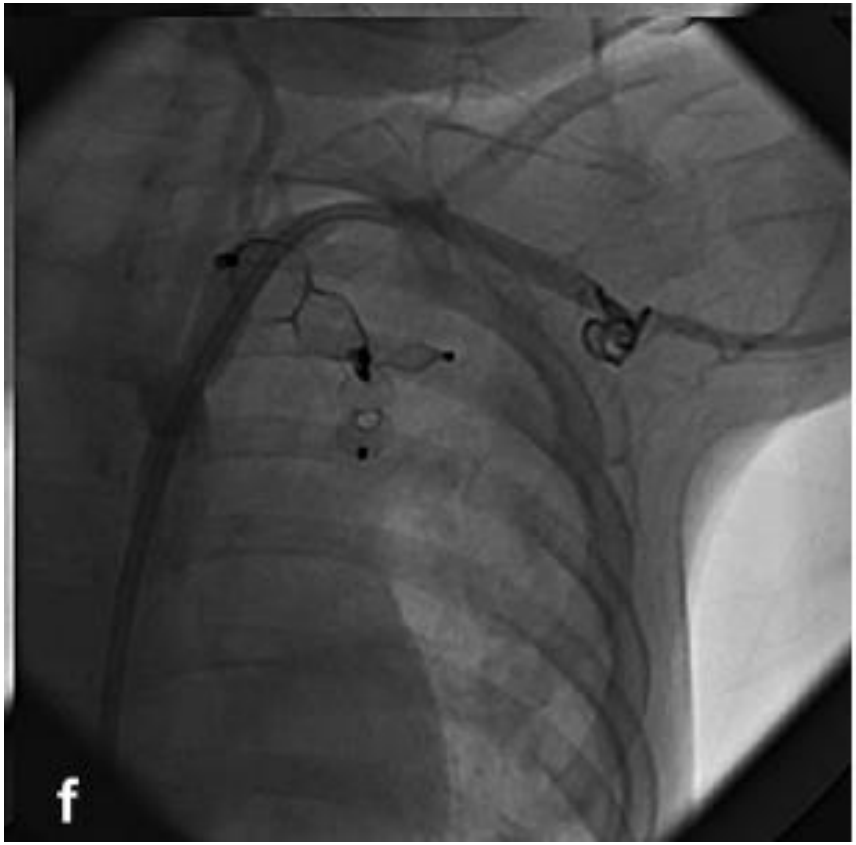
Management	Cases
Lobectomy	1
Surgical Ligation	11
Lobectomy and Surgical Ligation	3
Interventional	4
Refused Treatment	2

4) Jariwala P, Ramesh G, Sarat Chandra K. “Congenital anomalous/aberrant systemic artery to pulmonary venous fistula: closure with vascular plugs & coil embolization”. Indian Heart J. 2014 Jan-Feb;66(1):95-103. doi: 10.1016/j.ihj.2013.10.009. Review.

Interventional Management



Coil Embolization[6]



Vascular Plug & Coil[4]

4) Jariwala P, Ramesh G, Sarat Chandra K. “Congenital anomalous/aberrant systemic artery to pulmonary venous fistula: closure with vascular plugs & coil embolization”. Indian Heart J. 2014 Jan-Feb;66(1):95-103. doi: 10.1016/j.ihj.2013.10.009. Review.
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Risks of Embolization

- Treatment failure
  - Acute
  - Recurrence

- Migration of coil, material systemically → stroke
  - Higher risk than pulmonary AVMs

Summary - I

- Extremely rare, not associated with HHT
- Often asymptomatic, incidental finding
- May be hemodynamically significant SP-AVMs
- Not associated with cyanosis or increased pulmonary blood flow
- Theoretically increased risk of hemoptysis compared to other pulmonary AVMs

Summary - II

- Diagnosis: Selective angiography – passage of contrast to pulmonary vein and left atrium

- Management:
  - Arterial ligation
  - Lobectomy if lung damaged, ligation not possible
  - Interventional radiology used increasingly