

Pulmonary Hypertension

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Medicine

Disclosures

- None



Case Presentation



40 year old female with history of neuroendocrine tumor (s/p surgical resection) and bilateral pulmonary emboli (1 year prior) presents with dyspnea on exertion.



Workup:

Echo: LVEF 50-55%, RVSP 20-25mmHg, normal diastology
ESE: 7.5 METS (limited exercise capacity), no inducible ischemia

Cardiopulmonary stress: 6.7 METS, Max HR 98% predicted,
Peak VO₂ 23.3 (118% pred), RER 1.08, VE-VCO₂ 37 (n),
Breathing reserve 24.4% (abnormal)

PFTs: FEV₁/FVC 80.43 %, FEV₁ 89% pred, DLCO 73% pred

V/Q Scan: Large segmental perfusion defects in the apical right upper lobe and anterior basal segment of right lower lobe—chronic PE

Lab work negative for HIV, rheumatologic disease, drug screen



Case presentation

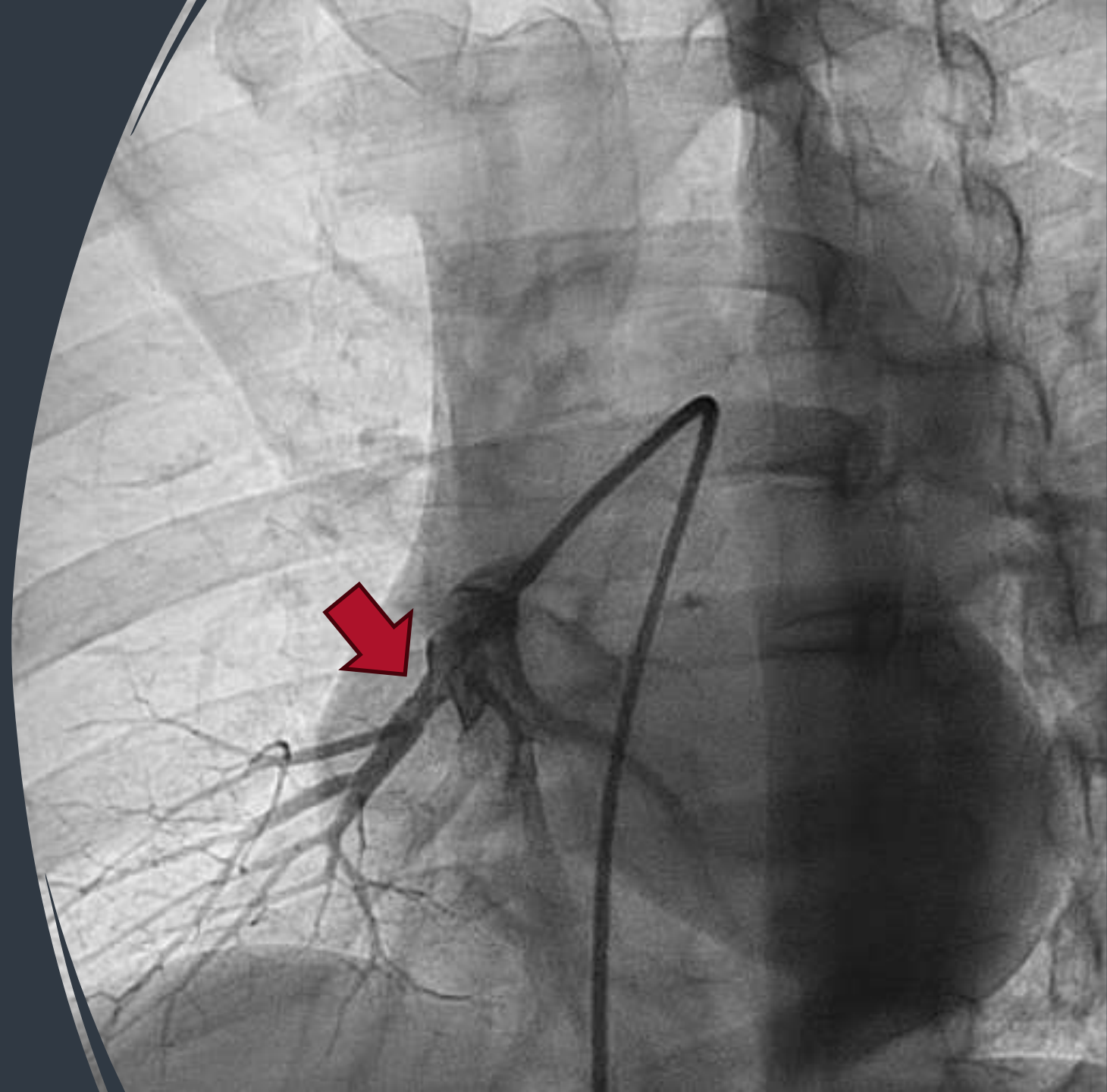
- Right heart catheterization

- RA: 18 RV: 47/7 PA: 41/16, mean 28 PCWP:19
- CO: 4.12 CI: 2.57 PVR: 3.5woods

- 6 minute walk test: 1519 ft

- Pulmonary angiogram:

- Main PA: Dilated
- RUL: A3 stenosis with distal tapering
- RML: Patent
- RLL: A8 distal webs, A9 proximal stenosis with ring lesion



Treatment and outcome

Riociguat started

Balloon angioplasty

- Balloon angioplasty to right A8 segment and subsegmental branches of A8
- Balloon angioplasty to right A9 segment and subsegmental branches of A9, A10 subsegments and subsegmental branches
 - Pre-angioplasty PA pressure: 25mmHg
 - Post-angioplasty PA pressure: 16mmHg

3-month follow up RHC

- RA: 10 RV: 34/4 PA: 31/13, mean 20
PCWP: 13
- CO: 5.1 CI:2.27 PVR: 1.3 woods

6 Minute walk: 1650 feet

Symptoms of dyspnea resolved



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