

Case Objectives

Case: A patient who presented chronic thromboembolic pulmonary hypertension with a large coronary fistula

1. To demonstrate possibilities of collaterals and fistulas in CTEPH patients.
2. To highlight the importance of LHC in addition to RHC for clinical imaging and diagnosis.

Background

- Chronic Thromboembolic Pulmonary Hypertension (CTEPH) is classified as WHO group 4 pulmonary hypertension.
- Major cause of PH and RV dysfunction → right heart failure → increased morbidity and mortality
- Coronary fistulas/collaterals are rare occurrences with CTEPH
- The fistulas shunt blood from coronaries into pulmonary circulation potentially compromising cardiac muscle blood flow → ischemia
- Multidisciplinary approach is necessary to care for the complex nature of these patients: from radiology and CT surgery to hematology and anesthesia to the nursing staff

Case Description

40 y/o female patient presents with worsening dyspnea and respiratory failure.

Required 4L O₂ per minute.

Prior diagnosis of RSV and Pneumonia. PMH revealed recurrent Pulmonary Embolisms after giving birth.

Patient was on indefinite Xarelto for anticoagulation

Differentials

- Chronic Thromboembolic Pulmonary Hypertension
- Pulmonary Embolism

Diagnostic Tests

- CTA Chest revealed occluded right pulmonary artery
- V/Q scan demonstrating complete absence of perfusion to right lung despite complete ventilation
- Cardiac Catheterization (Left & Right) confirmed pulmonary hypertension. No significant CAD present
 - Large Coronary Fistula originating from the Left Circumflex to the Right Pulmonary Artery

Patient Management & Intervention

- Patient elected to undergo surgical intervention for treatment → pulmonary thromboendarterectomy
 - Right Main pulmonary artery was divided and extended to hilum
 - Large AVM did not allow further dissection
 - Collateral vessels from AVM were divided
 - Mass was identified in the mRAP down to A1-A3 branches, right middle lobe branch, superior segment, basilar segment and all anteromedial and posterolateral branch
 - Specimen removed and otherwise uneventful surgical course
 - Extubated next morning

Images

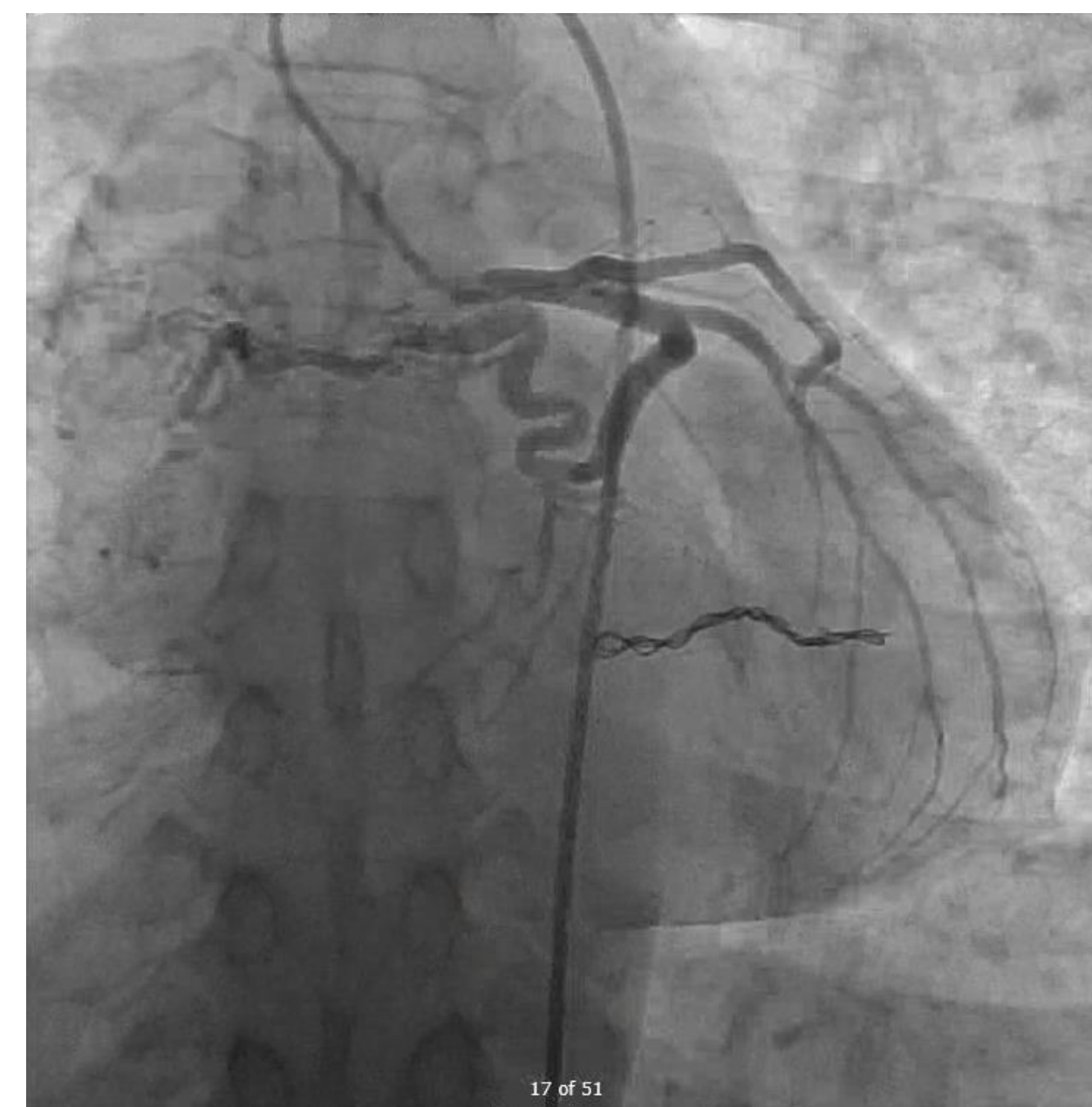


Figure 1: Coronary fistula take off connecting to right main pulmonary artery

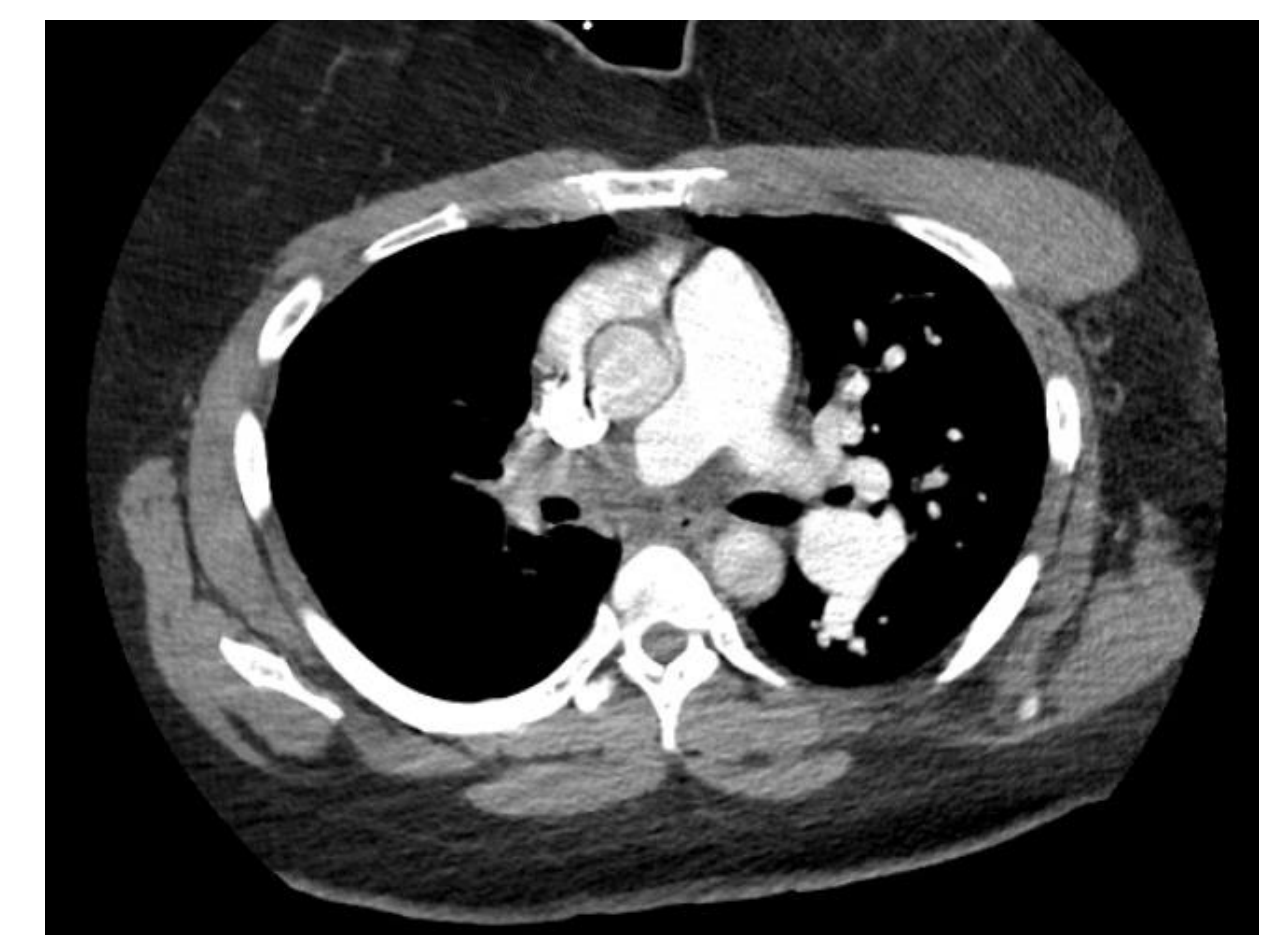


Figure 2: CT scan showing complete obstruction in the right pulmonary artery going to the right lung.

DLCO		Pred.	Best	% Pred.
DLco	mL/min/mmHg	24.33	16.84	69%
VA	L	4.97	3.93	79%
DLco/VA	mL/min/mmHg/L	4.94	4.29	87%
BHT	sec	---	12.69	---
VI	L	---	2.21	---
DL QA		---	D	---

Figure 3: DLCO measurements demonstrating minimal gas exchange in right lung pre operatively.

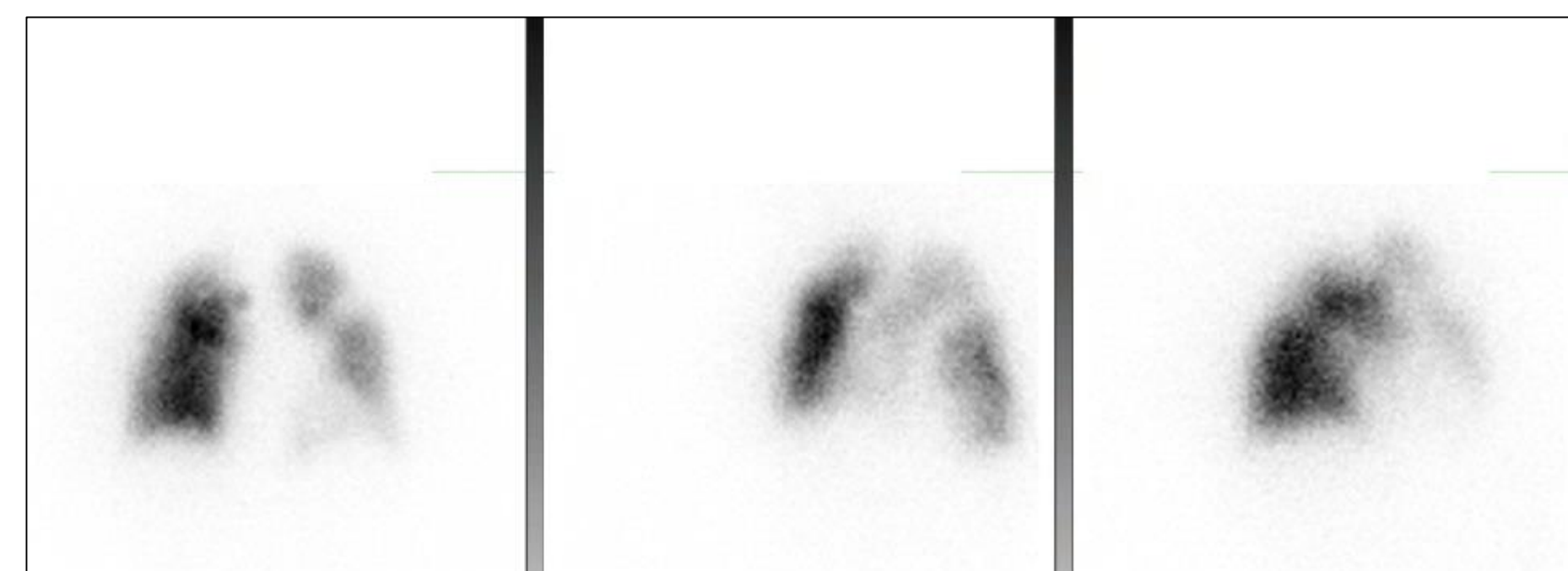


Figure 4: VQ scan post op showing increased perfusion to right lung post pulmonary thromboemblectomy and endarterectomy.

Discussion

- Increased prevalence of coronary to pulmonary fistulas in CTEPH patients → suggest potential benefit of attempts to maintain adequate blood supply in distal areas to the pulmonary arterial embolism (1)
- Formation of fistula could be through extension of SA nodal artery towards PA (2)
- 35% of SA nodal arteries stem from LCx and 65% from RCA → our patient was LCx (2)
- Patients with less distal CTEPH may experience a greater pressure difference between the systemic circulation and the pulmonary circulation distal to the occlusion, which can stimulate the development of either bronchopulmonary or coronary-pulmonary collaterals (3)

Conclusions

- Completing all diagnostic tests, including cardiac MRI and LHC will ensure proper diagnosis and a more comprehensive and complete patient assessment when planning for surgical pulmonary thromboemblectomy and endarterectomy
- Further studies are warranted to comprehend the correlation between coronary pulmonary fistulas and CTEPH better.

Contact

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References

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