

AMSER Case of the Month

August 2024

44-year old male with recurrent SNUC presenting with progressive dyspnea and hypoxemia.

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Patient Presentation

- **Brief HPI:** 44-year old male with history of right ethmoid polyp discovered in 2020, identified as sinonasal undifferentiated carcinoma (SNUC) s/p neoadjuvant chemotherapy, resection, and radiation at Huntsman Cancer Institute and Mayo-AZ, presenting in 2021 with **new onset diplopia/right upward gaze palsy with confirmed recurrence of SNUC.**
- Scheduled for palliative chemo 1 month later, but at visit is found to have progressive **severe dyspnea and hypoxemia** of several days.

Pertinent Labs

- **Labs on Initial Admission**

- **Significant for:**

- Thrombocytopenia (platelets- 83)
 - **Elevated D-dimer**
 - **Elevated Transaminases**
 - AST 199
 - ALT 162
 - Alkaline Phosphatase 232
 - Negative hepatitis panel
 - Negative autoimmune hepatitis panel, Wilson's

- **Labs on Subsequent Visit**

- **Significant for:**

- Negative COVID, Negative respiratory panel

What Imaging Should We Order?

ACR Appropriateness Criteria

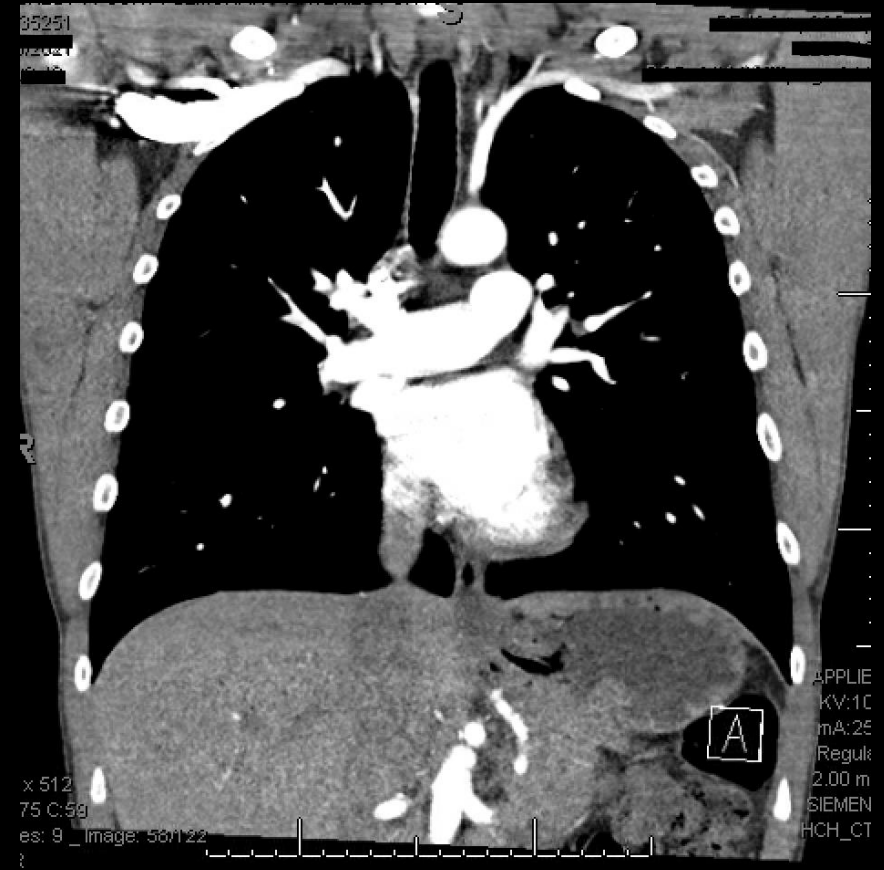
Variant 2:

Suspected pulmonary embolism. Low or intermediate pretest probability with a positive D-dimer. Initial imaging.

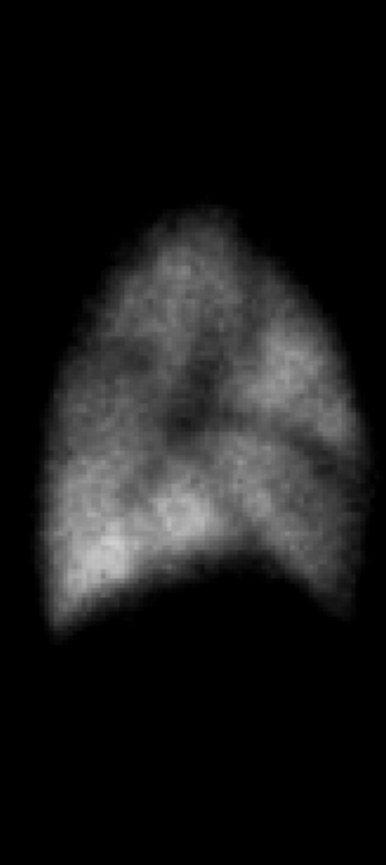
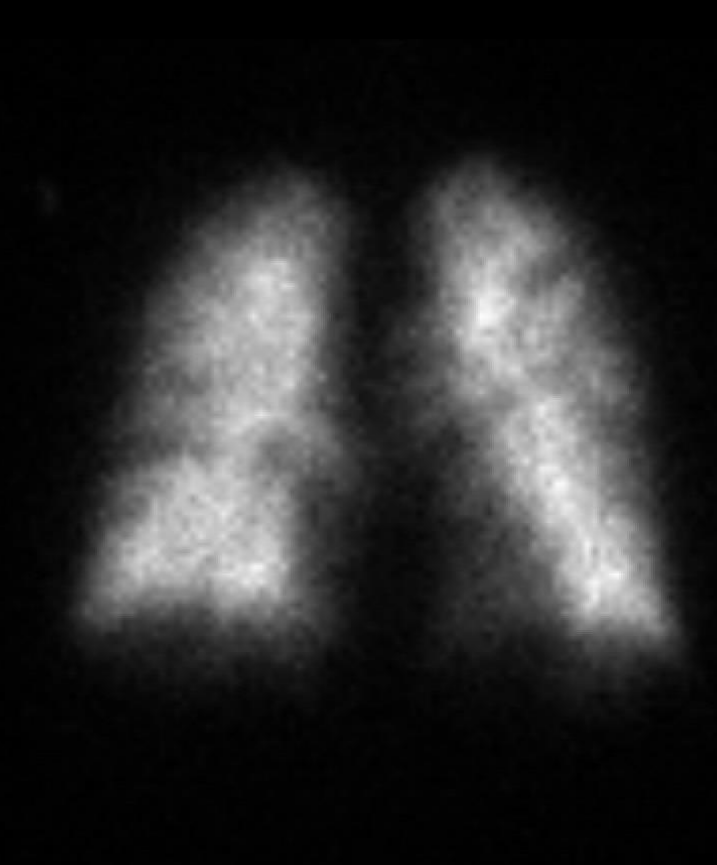
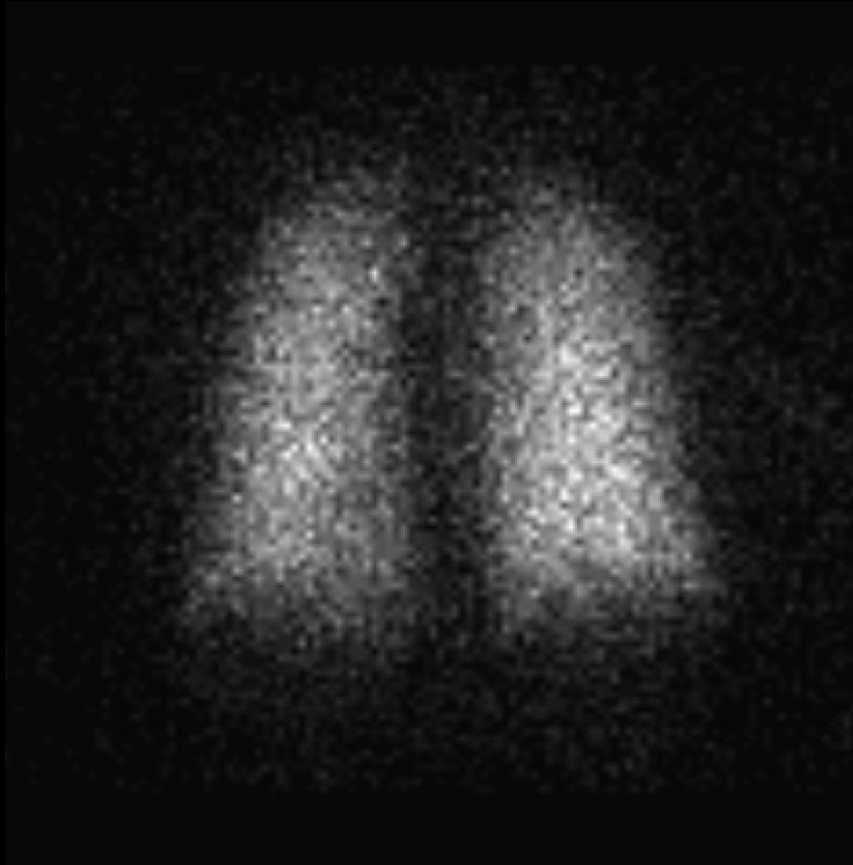
Procedure	Appropriateness Category	Relative Radiation Level
CTA pulmonary arteries with IV contrast	Usually Appropriate	☼☼☼
V/Q scan lung	Usually Appropriate	☼☼☼
MRA pulmonary arteries without and with IV contrast	May Be Appropriate	○
CTA triple rule out	May Be Appropriate (Disagreement)	☼☼☼
US duplex Doppler lower extremity	Usually Not Appropriate	○
US echocardiography transesophageal	Usually Not Appropriate	○
US echocardiography transthoracic resting	Usually Not Appropriate	○
Arteriography pulmonary with right heart catheterization	Usually Not Appropriate	☼☼☼☼
MRA pulmonary arteries without IV contrast	Usually Not Appropriate	○
CT chest with IV contrast	Usually Not Appropriate	☼☼☼
CT chest without and with IV contrast	Usually Not Appropriate	☼☼☼
CT chest without IV contrast	Usually Not Appropriate	☼☼☼
CTA chest with IV contrast with CTV lower extremities	Usually Not Appropriate	☼☼☼

We ordered both a CTPA and a V/Q lung scan

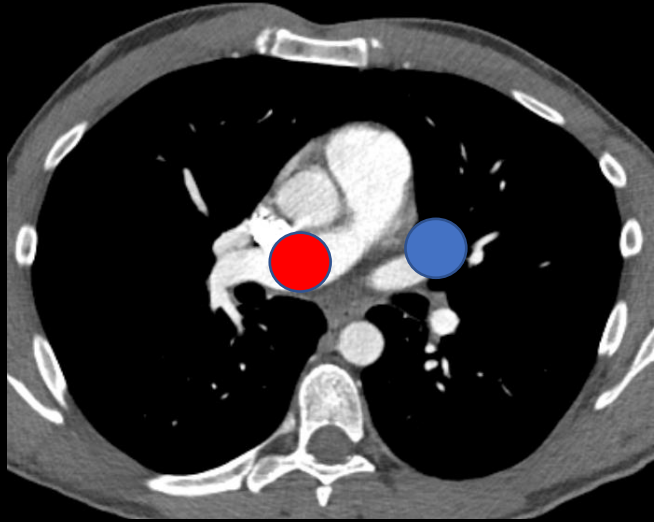
CTPA w/contrast for suspected PE (unlabeled)



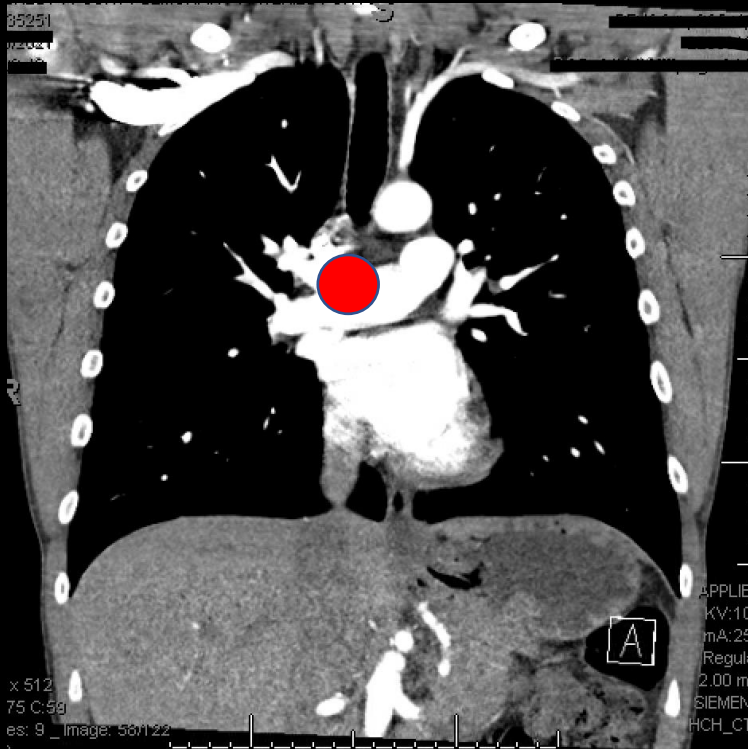
Radionucleotide Ventilation and Perfusion Scan Findings (unlabeled)



CTPA Axial View



CTPA Coronal View



CTPA w/contrast for suspected PE (labeled)

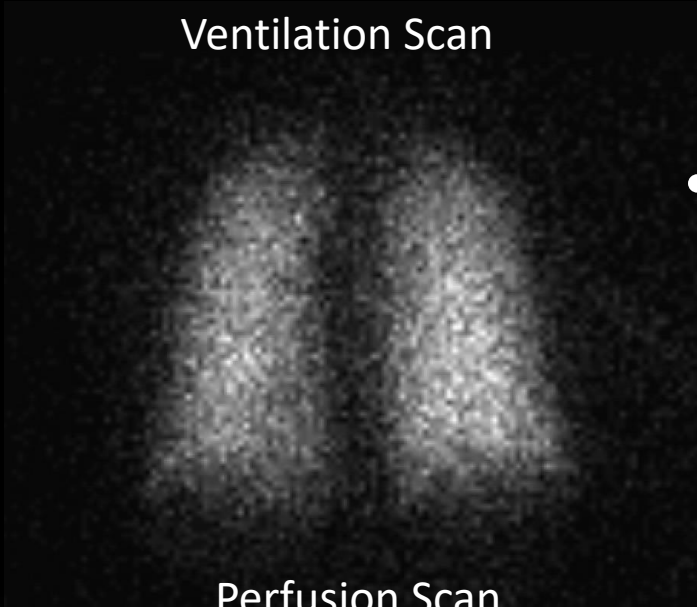
- On CTPA, the pulmonary arteries are patent, with **no pulmonary emboli identified** to the subsegmental pulmonary arteries bilaterally.
- No abnormal nodules are appreciated as well.

● Patent right pulmonary artery

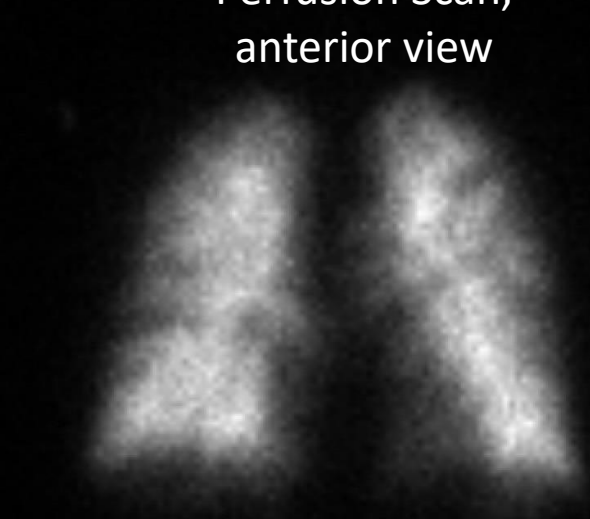
● Patent left pulmonary artery

Radionucleotide Ventilation and Perfusion Scan Findings (labeled)

Ventilation Scan

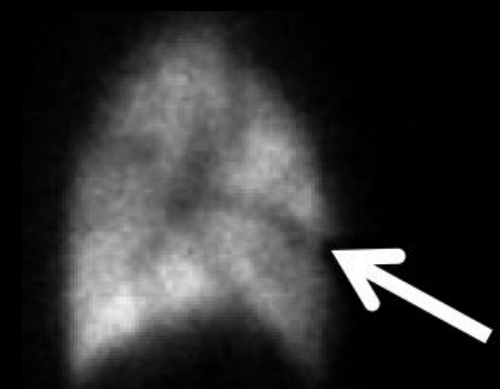


Perfusion Scan, anterior view



- **Ventilation Scan:** Diffuse, homogenous distribution of radionucleotides w/o evidence of ventilation defects.
- **Perfusion Scans:** Extensive pattern of **striated perfusion defects** extending diffusely and bilaterally from the periphery toward the hilum.

Perfusion Scan, lateral view



Final Diagnosis

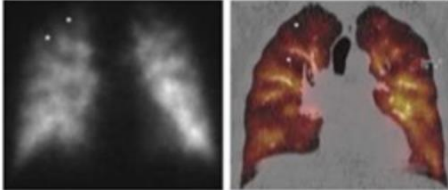
Tumor Thrombotic Microangiopathy from SNUC

“Segmental Contour Mapping”

- This **segmental pattern of perfusion restriction** in the lungs is known as ‘segmental contour mapping’ and is **pathognomonic** for tumor thrombotic microangiopathy (PTTP).
- These segmental contour patterns reflect diffusion restrictions due to microscopic tumor metastasis plugging up smaller arterioles.
- The consequence of which is **rapid multiorgan failure**. This diagnosis carries a **poor prognosis**.

SIGNS IN CARDIOPULMONARY IMAGING

The Segmental Contour Pattern
Laura B. Eisenmenger, MD and Howard Mann, MBBCh



Segmental Contour Pattern

Subsegmental-sized perfusion defects on lung perfusion scintigraphy representing the so-called segmental contour pattern, a consequence of tumor-associated occlusion of segmental pulmonary arteries and arterioles.

Differential Diagnosis

Pulmonary Tumor Embolism
Bland Pulmonary Embolism

FIGURE. (Tc-99m MAA perfusion scan, posterior-anterior projection), (Fused SPECT-CT, coronal reformation) demonstrate multifocal, subsegmental-sized perfusion defects (examples indicated by asterisks) that appear to “outline” bronchopulmonary segments.

Appearance: The segmental contour pattern is seen on lung perfusion scintigraphy. Bilateral, multifocal subsegmental-sized perfusion defects appear to outline the anatomic boundaries of bronchopulmonary segments.

Explanation: This scintigraphic pattern reflects a heterogenous distribution of increased vascular resistances when small segmental pulmonary arteries and arterioles are narrowed by intraluminal tumor and/or tumor-associated fibrocellular intimal proliferation.

Discussion: Dyspnea is a common symptom of patients with cancer. When sudden dyspnea occurs or worsens over a short period of time, usual diagnostic considerations include pulmonary embolism and diffuse lymphangitic spread of tumor. The latter is typically associated with interstitial lung abnormalities on chest radiography and computed tomography (CT), consequent on tumor obstruction of pulmonary lymphatics. When dyspnea is accompanied by findings of pulmonary arterial hypertension, particularly when this dominates the clinical presentation, the presence of diffuse occlusion of pulmonary microvessels should also be considered. The latter may reflect the occlusion of vessels by metastatic tumor or extensive fibrocellular intimal proliferation—the entity of tumor thrombotic microangiopathy.¹

Additional Finding – Multiorgan Failure

- In the setting of the patient's elevated transaminases and now this perfusion defect, an Abdominal MRI was performed, which showed a similar segmental pattern of restriction.



Axial T1 post-contrast with fat saturation MR of the liver shows **patchy enhancement in the periphery** of liver segments

Key Learning Points

- In the setting of a newly diagnosed malignancy and acute onset dyspnea and pulmonary hypertension **but no DVT/PE on imaging**, tumor thrombotic microangiopathy (TTM) should be on the differential.
- **Segmental contour mapping** is pathognomonic for tumor thrombotic microangiopathy and may be diagnosed with V/Q scan or dual-energy CT
- TTM carries a **very poor prognosis**, with many patients passing soon after diagnosis

References

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