PVOD Mimic in the Setting of Malignancy, A Case Report

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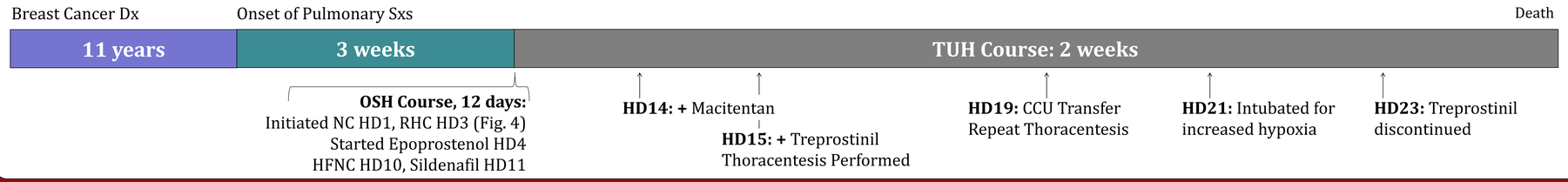
Introduction

Pulmonary hypertension (PH) related to cancer is a phenomenon that is not well understood. Definitive diagnosis with biopsy is often not feasible in these patients due to elevated operative risk in the setting of severe cardiopulmonary compromise, and thus the diagnosis is largely a clinical one. Prompt identification of the correct disease phenotype is necessary because initiation of PH-specific targeted therapy can worsen a patient's clinical status. We present a patient who developed severe PH in the setting of malignancy, and subsequently worsened in a manner that mimicked Pulmonary Veno-Occlusive Disease (PVOD).

Case

A 74-year-old female presented with progressive dyspnea on exertion and hypoxia. Medical history included breast cancer and paroxysmal AF. The breast cancer was diagnosed over 20 years prior and successfully treated with radiation. On admission, she had an irregularly irregular rhythm and was breathing comfortably with oxygen supplementation via nasal cannula (NC). Dopplers and ventilation/perfusion (V/Q) scan were negative for deep vein thrombosis (DVT) and pulmonary embolism (PE), respectively. CTA showed an enlarged pulmonary artery (PA) and mosaic pattern of densities of new pulmonary nodules, concerning for metastatic breast cancer (Fig. 2). Echocardiogram revealed a dilated RV and right heart catheterization was consistent severe PH (Fig. 1, Fig. 4). The patient underwent bilateral thoracentesis for pleural effusions. Cytology was negative, however there was still a high clinical suspicion for recurrence of malignancy. She was initiated on medical therapy in a step-wise fashion with Sildenafil, Macitentan, and Treprostinil which paradoxically led to declining respiratory status. The patient ultimately required ventilator support and maximal oxygenation for hypoxic respiratory failure, presumed to be secondary to cancer-related PVOD-like syndrome given her clinical deterioration with pulmonary vasodilator therapy.



Discussion

PH related to cancer has been described by a number of case reports, and at least seven different pathophysiologic mechanisms have been proposed. This discordance with regard to the underlying disease process highlights that it is not well-understood. This case offers an example of cancer-associated pulmonary disease resulting in pulmonary hypertension, which behaved similar to PVOD, with clinical worsening after starting pulmonary vasodilators. Despite the ultimate complexity of the case, this patient initially presented with dyspnea on exertion, which re-demonstrates the challenging nature of diagnosis with vague symptomatology.

There are no treatment guidelines for cancer-associated PVOD. Given the poor outcome typically seen in these cases, definitive treatment with bilateral lung transplantation is often not a feasible option, and therapy mainly relies on treatment of the underlying malignancy and supplemental oxygen for hypoxia. Unfortunately, some studies have linked chemotherapy treatment to malignancy-associated PVOD which makes therapy even more of a challenge.

This case also highlights the progression from initial onset of dyspnea to death in 5 weeks. A similarly rapid timeline is demonstrated in the literature, further emphasizing the importance of early suspicion for cancer-associated PVOD.

Clinical Data

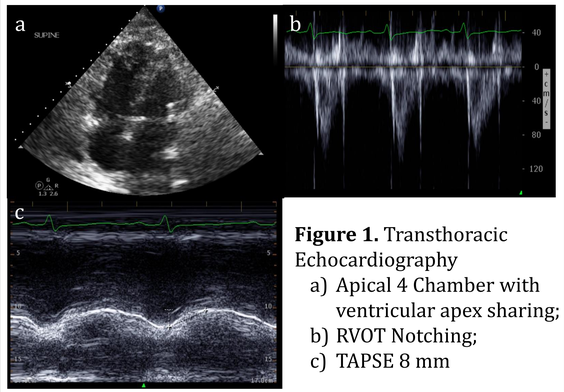


Figure 1. Transthoracic Echocardiography
a) Apical 4 Chamber with ventricular apex sharing;
b) RVOT Notching;
c) TAPSE 8 mm

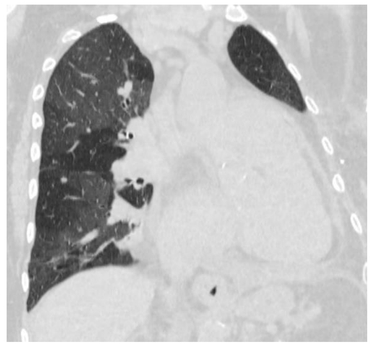


Figure 2. CT chest coronal

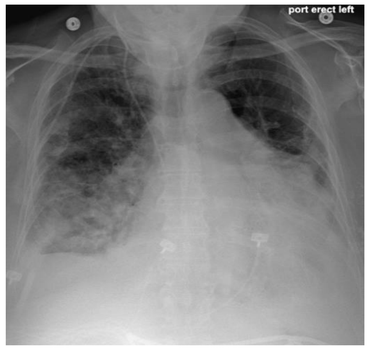


Figure 3. Chest x-ray

Hemodynamic Parameter	Values
Right Atrial Pressure (mmHg)	12
Systolic/Diastolic (Mean) Right Ventricular Pressure (mmHg)	107/10 (16)
Systolic/Diastolic (Mean) Pulmonary Artery Pressure (mmHg)	110/34 (64)
Pulmonary Capillary Wedge Pressure (mmHg)	15
Cardiac Output (L/min)	3.3
Cardiac Index (L/min/m^2)	1.65
Systemic Vascular Resistance (dynes-sec/cm^{-5})	1384
Pulmonary Vascular Resistance (woods units)	11

Figure 4. Right Heart Catheterization

Key Points

- Cancer-associated PVOD can present with vague symptomatology and rapidly progress to death.
- It is mainly a clinical diagnosis, so suspicion for this disease process is essential to detecting it.
- Avoiding pulmonary vasodilators in these patients is important as this has been shown to worsen their respiratory status.
- More research is needed to pinpoint the mechanism behind this disease process.

References

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