Pulmonary Hypertension in an ESRD patient : More than meets the eye

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Introduction

Pulmonary hypertension is a heterogeneous group of disorders characterized by a mean pulmonary artery pressure exceeding 20 mmHg.

We describe an interesting case of a patient with an arterio-venous fistula having worsening hypoxia that was initially attributed to progression of her COPD and heart failure. However, after a meticulous work up, we found an interplay of complex factors driving her hypoxia as described below.

Case Presentation

- 72 year old female with history of hypertension (managed with carvedilol, amlodipine, hydralazine and isosorbide mononitrate), COPD recently initiated on 2 L of oxygen for dyspnea and hypoxia, prior ESRD status post renal transplant in 2008 (her AV fistula was in her left arm and was unligated) presented to the ED with an episode of acute dyspnea. Physical examination and imaging were indicative of volume overload. She was admitted and diuresed aggressively. Despite achieving euvolemia, she only had a marginal improvement in dyspnea with minimal improvement in her hypoxia.
- 2D Transthoracic echocardiogram revealed preserved LV and RV Function with some diastolic septal flattening and grade II diastolic dysfunction;
- PFTs showed normal lung volumes with an FEV1/FVC ratio of 90%, DLCO of 24% and her HRCT showed mild centrilobular emphysema. At this point, it was clear that her shortness of breath and hypoxia was out of proportion to her COPD.
- A Right heart catheterization without fistula occlusion demonstrated the following:

RA: 1 mmHg,

PA: 59/15 with a mean of 34 mmHg,

PCWP: 10 mmHg,

CO/CI: 8.43/5.65,

TPG: 24 mmHg, PVR: 4.3 Woods Units,

DPG: 9 mmHg and

The Shunt run was negative.

- The patient had a AV fistula Doppler ultrasound that showed flows **in excess of 2.8 liters per min**

Imaging

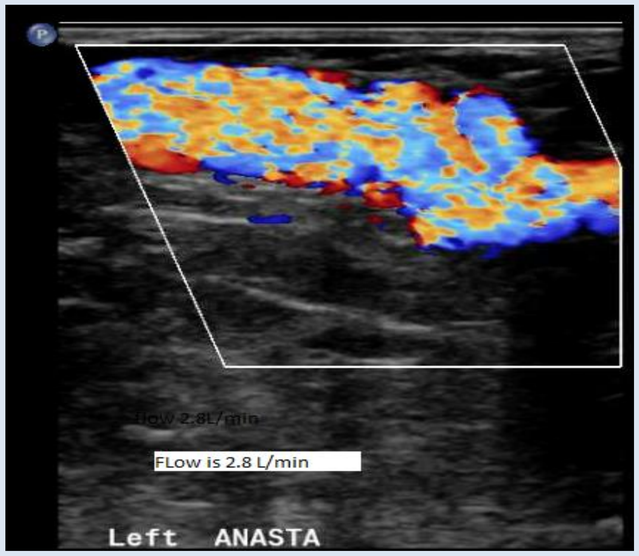

Image 1: high flow through the AV fistula

Management

We hypothesized that her underlying COPD in conjunction with the high output state from her hyperdynamic AV fistula resulted in pulmonary vascular remodeling and consequent pulmonary HTN (WHO 2 and 3).

The combination of hydralazine and isosorbide mononitrate (both of which are weak pulmonary arterial vasodilators) in conjunction with the high output state created by the AV fistula resulted in increased blood flow through areas of diseased lung parenchyma worsening her V/Q mismatch accounting for the progressive hypoxia.

The patient was evaluated by the vascular surgery team and had the AV fistula ligated. Closure of the fistula would eliminate the hyperdynamic circulation and hence reduce the pulmonary blood flow.

She was seen in the office after the procedure and was gradually weaned off the hydralazine and isosorbide mononitrate to a different anti-hypertensive regimen . She had regular 6 min walk tests for home oxygen determination in the office. Within a couple of weeks, the patient's symptoms improved and was successfully weaned off supplemental oxygen.

Mechanisms of pulmonary hypertension in ESRD patients

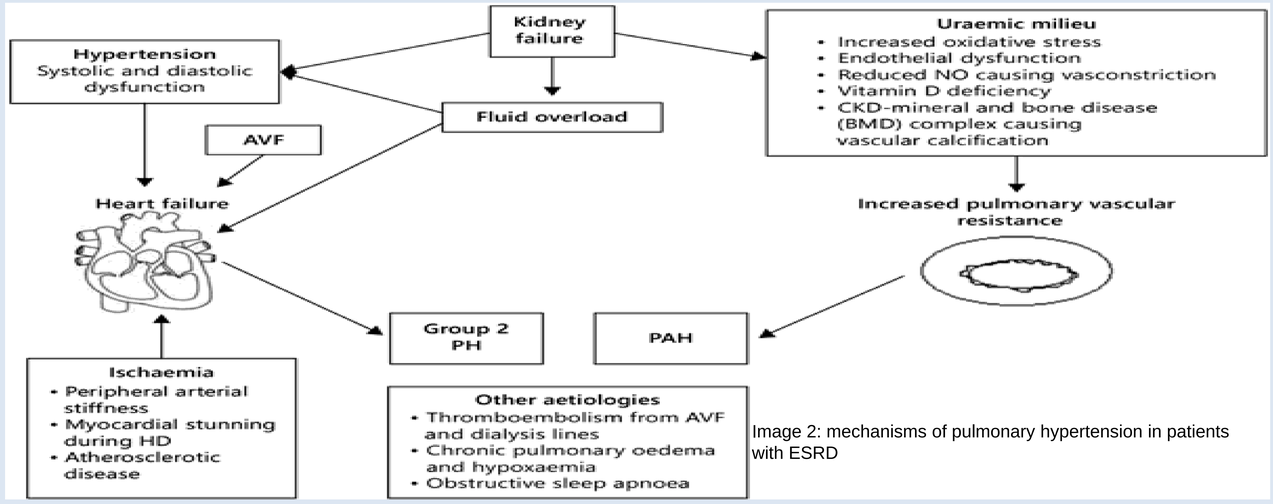

Image 2: mechanisms of pulmonary hypertension in patients with ESRD

Kawar B, Ellam T, Jackson C, Kiely DG. Pulmonary hypertension in renal disease: epidemiology, potential mechanisms and implications. Am J Nephrol. 2013;37(3):281-90. doi: 10.1159/000348804. Epub 2013 Mar 16. PMID: 23548763.

Discussion

Pulmonary hypertension in ESRD patients with fistulae has been described in several case series. [1,2] They are typically more likely to occur in patients with proximal fistulae with flows exceeding 2.2 l/min, which was the case for our patient. [3]

Her hypoxia was initially attributed to progression of her COPD and HFpEF; while they contributed to the acute presentation , the etiology of her hypoxia was more complex than expected.

The factors contributing to the development of pulmonary hypertension in ESRD patients are numerous as illustrated in image 2. [3]

The first step in management of our patient consisted in closing her fistula. Interestingly, in the paper of Forfia and al. they mentioned that occlusion of the AV fistula in the cath lab reduced the cardiac output by about 1 L/min and may actually under estimate the effects of a hyperdynamic circulation on the pulmonary vasculature.[1] Moreover, they found that definitive closure of the fistula resulted in a net decrease in cardiac out put of about 4L/min.[1] Closing the fistula of our patient restored normal pulmonary blood flow which facilitated the next step.

The patient was gradually weaned off the pulmonary vasodilators which in theory stopped the shunting and restored adequate VQ ratio and obviated the need for supplemental oxygen.

Conclusion

This case demonstrates the multiple pathways for the development of pulmonary hypertension in patients with ESRD. Most of the times, ESRD patients have numerous common diseases that may cause them to become hypoxic. However, having tunneled vision may delay accurate diagnosis and management of hypoxia & pulmonary hypertension in this patient population.

References

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2) Yigla M, Nakhoul F, Sabag A, Tov N, Gorevich B, Abassi Z, Reisner SA. Pulmonary hypertension in patients with end-stage renal disease. Chest. 2003 May;123(5):1577-82. doi: 10.1378/chest.123.5.1577. PMID: 12740276.

3) Kawar B, Ellam T, Jackson C, Kiely DG. Pulmonary hypertension in renal disease: epidemiology, potential mechanisms and implications. Am J Nephrol. 2013;37(3):281-90. doi: 10.1159/000348804. Epub 2013 Mar 16. PMID: 23548763.

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