Introduction

Determining the cause of pulmonary hypertension (PH) in an individual patient can be challenging as PH can be a result of multiple etiologies. We present a unique case of a patient with known hypertrophic cardiomyopathy (HCM) who was diagnosed with PH of unanticipated etiologies.

Case Presentation

HPI

• 70-year-old male with a history of non-obstructive HCM and remote pulmonary embolism (PE) presented to the hospital with worsening lower extremity swelling and dyspnea. Physical examination was significant for jugular-venous distention and 1+ pitting edema of the lower extremities.

Exam/Labs

• NT-proBNP 386 pg/mL
• high sensitivity troponin 396 ng/mL

Imaging

• ECG: sinus rhythm, 1st degree atrioventricular block, and bifascicular block
• High resolution chest CT showed dilation of main pulmonary artery, patchy groundglass infiltrates and some scarring in left lung. Due to history of prior PE, a ventilation perfusion (V/Q) scan was done which showed decreased perfusion in left lower lobe of the lung with homogenous ventilation bilaterally.
• A pulmonary angiogram was consistent with chronic thromboembolic pulmonary hypertension (CTEPH).

TTE

• Transthoracic echocardiogram revealed severe concentric left ventricular hypertrophy (LVH), LV end-diastolic diameter 3.6 cm, ejection fraction 65-70%, dilated right ventricle with severely reduced systolic function, severe tricuspid regurgitation and estimated peak systolic pressure 70 mmHg

RHC results as seen in Table 1
• Started on intravenous epoprostenol and diuretics

Further Testing

• ANA titers were mildly elevated with homogenous pattern (>1:160) but ANA screen with antibody returned negative.
• HIV antigen/antibody screen was negative.

Management

• Patient refused surgical or percutaneous treatment of CTEPH and wanted only medical therapy. He responded poorly to parenteral vasodilators with intolerable side effects. Riociguat was started but stopped due to hemoptysis. Subsequently he opted for comfort care and was enrolled in hospice.

Table 1: Right Heart Cath Measurements

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<table>
<thead>
<tr>
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<tbody>
<tr>
<td>Right atrium</td>
<td>17 mmHg</td>
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<tr>
<td>Right ventricle</td>
<td>90/16 mmHg</td>
</tr>
<tr>
<td>Pulmonary Artery</td>
<td>89/48/62 mmHg</td>
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<tr>
<td>Pressures</td>
<td></td>
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<tr>
<td>Pulmonary arterial wedge pressure (PAWP)</td>
<td>12 mmHg</td>
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<tr>
<td>Pulmonary Vascular Resistance (PVR)</td>
<td>17.5 wood units</td>
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<tr>
<td>Cardiac Index</td>
<td>1.3L/min/m²</td>
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Figure 1, 2, 3:

Transthoracic echocardiogram showing septal hypertrophy flattening in systole and diastole indicating pressure and volume overload in a parasternal short axis view. Apical four chamber view, LVH, septal hypertrophy and enlargement of right sided cardiac chambers.

Conclusion

This case of HCM with suspected predominant WHO group II PH is an example of how other less likely etiologies may be a cause of PH. Given positive ANA titers and HCM with severe LVH, WHO group I and II disease respectively could not be ruled out. Therefore, it is essential to do a thorough evaluation as it may alter management.