A Case of Chronic Thromboembolic Pulmonary Hypertension in a Heart Transplant Patient, Successfully Treated with Pulmonary Endarterectomy

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INTRODUCTION

We present a case of symptomatic chronic thromboembolic pulmonary hypertension (CTEPH) in a patient 15 years post heart transplantation, who underwent pulmonary endarterectomy, with significant improvement in symptoms and pulmonary hypertension (PH) post procedure.

CASE HISTORY AND BACKGROUND

- 71-year-old male presenting with dyspnea.
- Past medical history includes;
  - Heart transplantation 15 years prior for idiopathic dilated cardiomyopathy
  - Pulmonary embolism (PE) after traumatic pulmonary contusion 10 years prior
  - Mild renal impairment
  - Type 2 diabetes mellitus
  - Hypertension
  - Obstructive sleep apnea
- Regular medications include; warfarin, cyclosporin, everolimus, mycophenolate mofetil and prednisolone.

INVESTIGATIONS

- Transthoracic echocardiogram demonstrated elevated pulmonary pressures.
- Right heart catheterization confirmed the diagnosis of PH;
  - Pulmonary artery systolic pressure (PASP) of 66 mmHg
  - Pulmonary artery diastolic pressure (PADP) of 28 mmHg
  - Mean pulmonary artery pressure (mPAP) of 45 mmHg
  - Mean right atrial pressure (RAP) of 9 mmHg
  - Mean pulmonary capillary wedge pressure of 8 mmHg
  - Pulmonary vascular resistance (PVR) of 562 dynes/sec/cm^5
  - Ventilation/perfusion scan and CT pulmonary angiogram demonstrated bilateral chronic PE.
  - Formal pulmonary angiography confirmed bilateral chronic PE.
- 6-minute walk test was 317 meters.

INTERVENTION

- Riociguat was commenced and was warfarin continued.
- Pulmonary endarterectomy performed after discussion at the multidisciplinary meeting.

OUTCOMES

- Dyspnea improved, with patient returning to normal activities.
- Right heart catheterization 11 months post-procedure demonstrated improvement;
  - SPAP of 47 mmHg
  - DPAP of 23 mmHg
  - mPAP of 33 mmHg
  - RAP of 10 mmHg
  - PVR of 240 dynes/sec/cm^5
- 6-minute walk test improved to 397 meters.
- Riociguat was continued due to demonstration of residual PH.

CONCLUSIONS

This demonstrates the first described case of CTEPH in a patient post-heart transplantation. As an eminently treatable cause of PH, the importance of a high index of suspicion for CTEPH is clear.