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CLINICAL QUIZ

Pulmonary hypertension in an adolescent with end-stage-renal disease—a diagnostic challenge: Questions

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Introduction

The prevalence of pulmonary hypertension (PH) in patients with end-stage-renal disease (ESRD) is higher than in controls matched for age, but the pathophysiology remains poorly understood. Here, we report the case of a 16-year-old boy who underwent two renal transplantations and who was referred to our institution to explore PH fortuitously diagnosed on echocardiography.

Case report

Our patient was diagnosed at 13 months old for congenital nephrotic syndrome with diffuse mesangial sclerosis on the kidney biopsy secondary to *PLCE1* mutation. He quickly progressed to ESRD and underwent bilateral nephrectomy because of severe hypertension. Following an initial period of hemodialysis on a jugular catheter, he underwent a first renal transplantation in 2001. Four years later, he had acute renal failure, nephrotic syndrome, and clinical and laboratory features of systemic thrombotic microangiopathy. He progressed rapidly to ESRD and initiated dialysis. Multiple hemodialysis catheter changes severely impaired jugular and sub clavicular venous accesses precluding arteriovenous (A-V) fistula creation on the upper limbs. Thus, a femoral arteriovenous (A-V) fistula was created in 2005. A second renal transplantation was performed in 2007. Fifteen months later, he had acute antibody-mediated rejection with deterioration of graft function.

During follow-up, he did not have systemic hypertension. An echocardiography performed in 2008 only showed a dilated left ventricle (LV) and normal systolic pulmonary pressure. In August 2012, at 16 years of age, he was admitted to the intensive care unit for rapidly progressing dyspnea. B-type natriuretic peptide was increased at 1462 ng/mL. Chest X-ray and a chest CT scan showed pulmonary edema. The echocardiography showed a dilated LV (LVEDD 56 mm, *z* score + 2.76) with normal systolic function and an ejection fraction of 65%. The maximum tricuspid regurgitation velocity showed an elevated systolic pulmonary pressure of 60 mmHg and signs of increased right atrial pressure above 10 mmHg. He improved with diuretics.

The patient's nephrologist asked the cardiologist to perform heart catheterization. Right heart catheterization confirmed PH with a mean pulmonary artery pressure of 50 mmHg and a pulmonary wedge pressure of 21 mmHg, while systemic pressure was 149/80 (mean 108) mmHg. Oxygen saturation in the inferior vena cava was 96% and cardiac index was at 10 L/min/m^2. Pulmonary vascular resistance (PVRi) was 2.8 WoodU*m^2. Acute vasoreactivity testing was performed by oxygen and NO inhalation and showed a drop in PVRi to 1.6 WoodU/m^2.

1) What is the cause of the pulmonary hypertension in this patient?
2) How should the pulmonary hypertension in this patient be treated?

The answers to these questions can be found at https://doi.org/10.1007/s00467-018-3939-x.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.