

Control Number: 32

Abstract Category: Clinical Case Challenge in Cardio-Oncology

Title: A unique case of reversible right heart failure

ABSTRACT BODY

Background and Purpose

Acquired pulmonary stenosis and right ventricular outflow tract obstruction due to extrinsic compression is infrequently described in the literature. Here we report a case of reversible severe pulmonary stenosis and right ventricular outflow tract (RVOT) obstruction from a large mediastinal rhabdomyosarcoma.

Case Description and Outcomes

A 38-year-old man with no significant past medical history presented with a one-month history of cough, orthopnea, and dyspnea with exertion. Physical exam demonstrated tachycardia, respiratory failure requiring high flow oxygen, elevated jugular venous pressure (JVP) and peripheral dependent edema. CT angiography demonstrated a 17-centimeter anterior mediastinal mass causing significant stenosis of the main pulmonary artery. A transthoracic echocardiogram (TTE) was obtained which showed dilatation of the right ventricle, hypokinesis of the right ventricular free wall, and global depression of the right ventricular systolic function due to extrinsic compression by the anterior mediastinal mass. Estimated peak systolic pulmonary artery pressure (PAP) was 65-70 mmHg. Biopsy of the mass revealed rhabdomyosarcoma, and he was treated with one cycle of adriamycin and ifosfamide. Three days after chemotherapy, respiratory failure and peripheral edema began to improve and JVP began to normalize. A repeat TTE was obtained which demonstrated an estimated peak systolic PAP of 32-37 mmHg and normal right ventricular size and systolic function.

Discussion

Acquired pulmonary stenosis and RVOT obstruction is a rare complication of anterior mediastinal malignancy. Surgical resection is the most important predictor of survival in mediastinal rhabdomyosarcoma. However, in this case, surgery as well as radiotherapy was prohibitive due to high risk for cardiovascular collapse. Additionally, mediastinal rhabdomyosarcomas are most often resistant to chemotherapy. Nevertheless, in this case, the tumor was indeed chemosensitive, and the resultant cytoreduction after a single cycle of chemotherapy led to a dramatic reversal of severe pulmonary stenosis and RVOT obstruction.

References

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3. Vyas, V., et al. "Primary mediastinal pleomorphic rhabdomyosarcoma: a case report." *Med principles and practice* 17.2 (2008): 154-156.

Image 1

