A unique case of reversible right heart failure

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LEARNING OBJECTIVES

- Describe a case of hemodynamically significant anterior mediastinal mass
- Discuss the management options for malignant mediastinal rhabdomyosarcoma causing right heart failure

HISTORY OF PRESENT ILLNESS

A 38-year-old man with no significant past medical history presented with a one month history of worsening non-productive cough, orthopnea, and dyspnea with exertion (NYHA functional class III-IV). He also reported mid-upper back pain that radiated towards the retrosternal area.

PHYSICAL EXAMINATION AND INITIAL IMAGING

Vital Signs: HR 140, BP 120/70, RR 44, SpO2 90%
General: awake, alert, in respiratory distress
Neck: JVD present to the angle of the mandible
Cardiac: IV/VI murmur of pulmonic stenosis at the LUSB
Pulmonary: decreased breath sounds R mid-lower lung field
Extremities: 1+ pitting edema to mid-calf

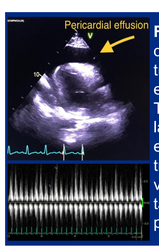

Figure 1. **Top**: 2-dimensional transthoracic echocardiogram (2-D TTE) demonstrating large pericardial effusion. **Bottom**: tricuspid inflow variation indicative of tamponade physiology.

HOSPITAL COURSE

Sonographic and clinical evidence of acute cardiac tamponade
- Emergent pericardio-centesis

Clinical deterioration with signs and symptoms or worsening right heart failure
- CTA with 17 cm anterior mediastinal mass **(Figure 2A)**
- TTE showed severe pulmonary stenosis and RVOT obstruction from anterior mediastinal mass **(Figure 2B-D)**.

Biopsy revealed rhabdomyosarcoma
- Not a surgical candidate due to risk for cardiovascular coll.
- Cycle 1 of AIM (adriamycin, ifosfamide, mesna started.

FINAL DIAGNOSIS

Prior to chemotherapy, transthoracic echocardiogram demonstrated severe right ventricular outflow tract obstruction (RVOT) with evidence of right heart failure, including hypokinesis of the right ventricular free wall, and global depression of the right ventricular systolic function due to extrinsic compression from the anterior mediastinal mass.

It also showed severe pulmonary arterial stenosis with an estimated peak systolic pulmonary artery pressure of 65-70 mmHg, suggestive of severe pulmonary stenosis.

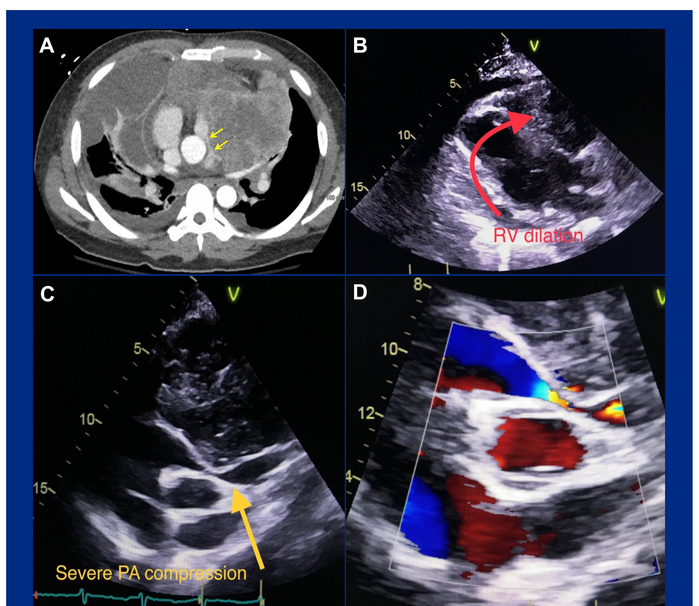

Figure 2. A. CT angiography demonstrated a large, heterogeneous anterior mediastinal mass compressing the main pulmonary artery (yellow arrow). **B.** Two-dimensional (2-D) transthoracic echocardiogram (TTE) showed dilatation of the right ventricle and plethoric IVC. **C**. Severe PA compression by mass. **D**. Doppler flow with elevated velocities at the area of stenosis.

HOSPITAL COURSE AND FOLLOW UP

Three days after chemotherapy, his respiratory failure and peripheral edema began to resolve and JVP began to normalize. The prior IV/VI murmur of pulmonary stenosis could no longer be auscultated. A repeat TTE was obtained nine days after chemotherapy which demonstrated an estimated peak systolic pulmonary artery pressure of 32-37 mmHg and normal right ventricular size and systolic function.

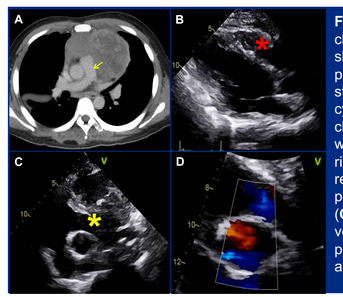

Figure 3. A. CT chest with contrast showed reversal of pulmonary artery stenosis after one cycle of AIM. Post-chemotherapy TTE without dilation of right ventricle (**B**), reversal of pulmonary stenosis (**C**), and normal velocity within pulmonary artery and RVOT (**D**).

DISCUSSION

Acquired pulmonary stenosis and RVOT obstruction is a rare complication of anterior mediastinal malignancy[1]. Surgical resection is the most important predictor of survival in mediastinal rhabdomyosarcoma[2]. However, in this case, surgery as well as radiotherapy was prohibitive due to high risk for cardiovascular collapse. In particular, adriamycin has been described to cause a high degree of intense cardiac dysfunction when used with radiotherapy[4].

Mediastinal rhabdomyosarcomas are most often resistant to chemotherapy[3]. 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.

CONCLUSIONS

Here we describe an unusual case of right heart failure, caused by extrinsic compression from a malignant mediastinal rhabdomyosarcoma, which responded dramatically with one cycle of chemotherapy and reversed sonographic and clinical signs and symptoms of right heart failure.

REFERENCES

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DISCLOSURES:

AUTHORS FOR THIS CASE PRESENTATION HAVE NO DISCLOSURES TO REPORT