

Minimally invasive mitral valve replacement for severe primary mitral regurgitation in a patient with lung neuroendocrine tumor

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LEARNING OBJECTIVES

- Present a unique case of left-sided valvular disease in a patient with lung neuroendocrine tumor.
- Discuss the potential etiology and pathogenesis of mitral regurgitation in a patient with carcinoid syndrome.
- Encourage cardiologists to consider minimally invasive intervention in patients with underlying malignancy, if appropriate.

HISTORY OF PRESENT ILLNESS

A 66-year-old woman presented with 6 months of persistent cough. Cross-sectional imaging demonstrated stage IIIa (T3N2M0) well-differentiated, low-grade neuroendocrine tumor (NET) of the left upper lobe of the lung, measuring 5.3x5.0cm, with encasement of the proximal thoracic descending aorta, pulmonary trunk, and left pulmonary artery.

LABORATORY DATA

- Chromogranin A 235 ng/mL (<96 ng/mL)

HOSPITAL COURSE

TTE demonstrated new moderate mitral regurgitation

TEE demonstrated severe mitral regurgitation (EROA 0.48cm², RVol > 60mL)

Patient was too high risk for surgical resection NET due to valvular heart disease

Decision was made for patient to undergo a minimally invasive mitral valve replacement

FINAL DIAGNOSIS

Severe primary mitral regurgitation due to carcinoid syndrome

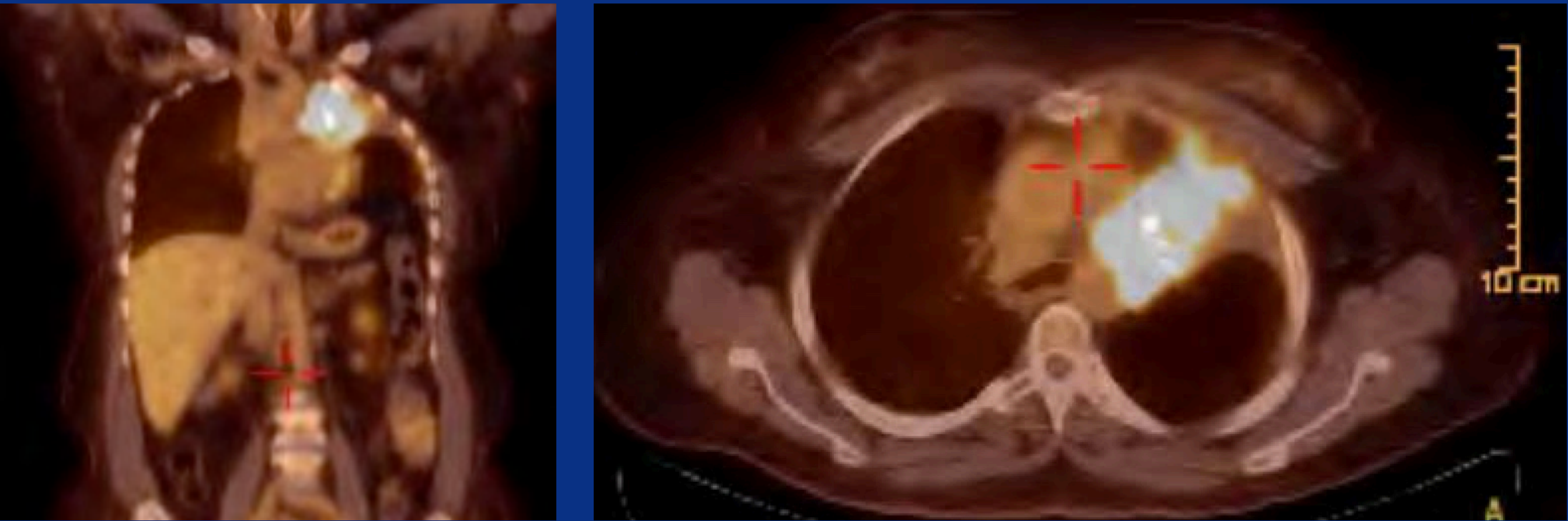


Figure 1: PET CT demonstrated a 5.3 x 5.0 cm lung NET with encasement of thoracic aorta, pulmonary trunk, and left pulmonary artery.

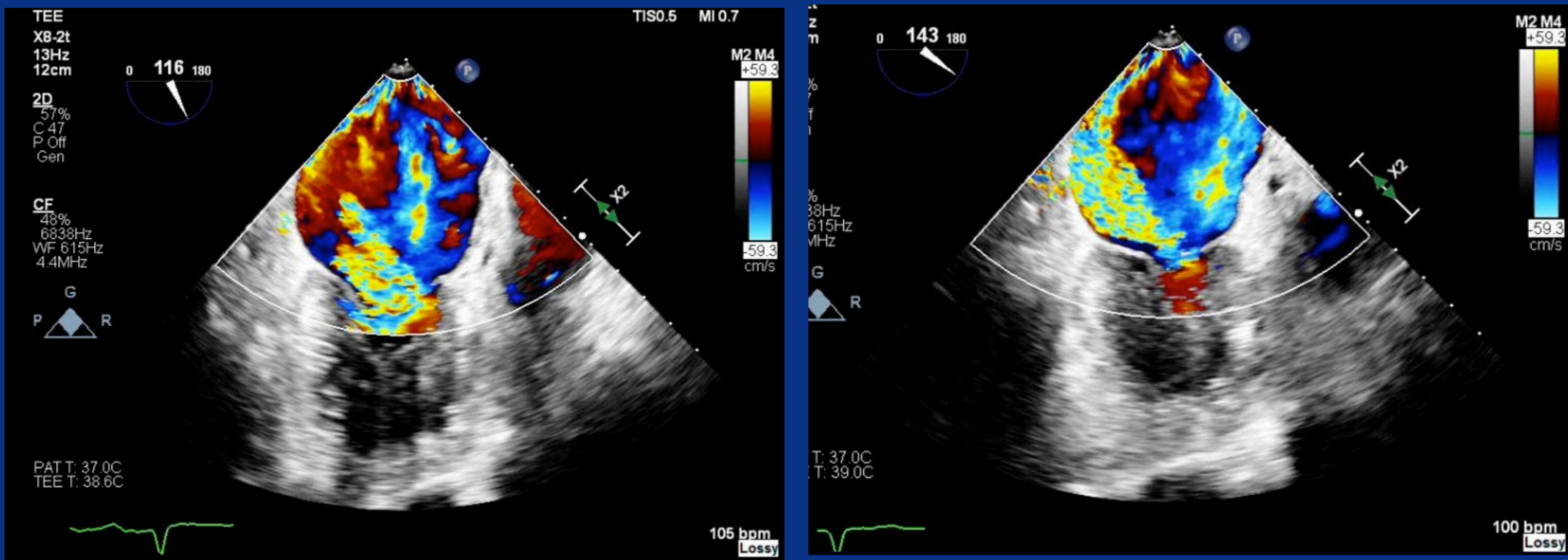


Figure 2: Echocardiographic appearance of left-sided carcinoid disease: A two-dimensional color-flow transesophageal echocardiogram showed thickened and retracted mitral valve leaflets with poor coaptation, leading to severe mitral regurgitation (EROA 0.48cm², RVol > 60 mL.)

FOLLOW-UP

The patient underwent minimally invasive, high risk mitral valve replacement with a 27 mm St. Jude porcine valve. She tolerated the procedure well. Unfortunately, though her valvular issues had resolved, her tumor was then deemed non-resectable.

DISCUSSION

- Lung NETs rarely produce carcinoid syndrome, and carcinoid syndrome characteristically produces right-sided valvular lesions.
- Left-sided valvular involvement has been described in patients with lung NET or PFO, and occurs especially when tumor size is greater than five centimeters.
- Characteristic echocardiographic features of carcinoid heart disease include:
 1. Lack of valvular calcification, and
 2. Thickened, shortened and retracted valve leaflets leading to malcoaptation and moderate to severe regurgitation.
- In this case, hormonal secretion from the patient's large lung neuroendocrine tumor contributed to severe left-sided valvular disease.
- Further, this unique case illustrates the utility of minimally invasive valvular replacement in patients who are not candidates for open surgery.

CONCLUSIONS

- Cardiac surgery has been demonstrated to significantly improve symptoms in patients with carcinoid syndrome and valvular involvement. However, invasive surgery is associated with high perioperative mortality.⁴
- New and evolving therapies have improved survival in patients with NET, and cardiac involvement has become a major cause of morbidity and mortality.
- Going forward, it is imperative to consider minimally invasive techniques for valvular repair in patients with NET, especially if the patient is high risk for open surgery.

REFERENCES

1. Hendifar, AE, et al. "Neuroendocrine tumors of the lung." *Journal of Thoracic Oncology* 12.3 (2017): 425-436.
2. Möller, Jacob E., et al. "Factors associated with progression of carcinoid heart disease." *New England Journal of Medicine* 348.11 (2003): 1005-1015.
3. Pellikka, PA, et al. "Carcinoid heart disease. Clinical and echocardiographic spectrum." *Circulation* 87.4 (1993): 1188-1196.
4. Connolly, Heidi M., et al. "Outcome of cardiac surgery for carcinoid heart disease." *Journal of the American College of Cardiology* 25.2 (1995): 410-416.

DISCLOSURES:

AUTHORS FOR THIS CASE REPORT HAVE NO DISCLOSURES TO REPORT