Fibrosing mediastinitis-related pulmonary artery and vein stenosis-limiting chemotherapy

Michael Lawrenz Ferreras Co,¹ Hena N Patel,² Arianne Clare Agdamag,² Tochukwu M Okwuosa²

¹Department of Medicine, Rush University Medical Center, Chicago, Illinois, USA

²Internal Medicine-Cardiology, Rush University Medical Center, Chicago, Illinois, USA

Correspondence to

Dr Michael Lawrenz Ferreras Co, michael_co@rush.edu

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DESCRIPTION

A 74-year-old woman presented for second opinion of dyspnoea management. Her medical history included breast cancer treated with mastectomy, doxorubicin, cyclophosphamide and tamoxifen. She developed recurrent metastatic disease in the mediastinum, managed with 89 six-week cycles of paclitaxel over 10 years that was well tolerated and effective in stabilising the disease initially but eventually discontinued due to new sternal metastasis diagnosed on imaging and increasing dyspnoea. Examination revealed diminished left-sided breath sounds. Routine labs were normal. Echocardiogram showed normal left ventricular function and dilated right chambers. Repeat chest CT demonstrated: (1) extensive calcification and soft tissue mass in the anterior mediastinum consistent with fibrosing mediastinitis (FM); (2) severe left pulmonary artery stenosis (PAS, figure 1); and (3) an infiltrating soft tissue density in the pericardium. Subsequent pericardial tissue biopsy showed non-specific fibrosis. Pulmonary angiography showed bilateral PAS. Angiographic and surgical interventions were deferred due to the extent of the mediastinal fibrosis and overall poor prognosis. Repeat imaging

Figure 1 CT chest showing severe left pulmonary artery stenosis.

Figure 2 Repeat CT several months later showing stenosis of the left pulmonary vein.

showed progression of FM with new left pulmonary vein stenosis (figure 2), leading to a multidisciplinary decision to pursue palliative care. Palliative hormonal therapy was subsequently initiated.

FM is a rare disorder characterised by fibrous tissue proliferation within the mediastinum leading to disabling symptoms or death with compression

Learning points

- ▶ Fibrosing mediastinitis (FM) should be considered in any patient who underwent chemotherapy and has dyspnoea.
- ▶ Paclitaxel, one of the commonly used chemotherapeutic drugs for metastatic breast cancer, may be associated with FM despite its known low pulmonary toxicity profile.
- ▶ Complications of FM, like pulmonary artery stenosis and pulmonary vein stenosis, can be potentially chemotherapy limiting and need to be addressed promptly. Pulmonary artery stenting or surgical repair could be considered; however, early diagnosis and intervention prior to extensive mediastinal fibrosis is paramount.

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of surrounding structures.¹ Paclitaxel and cyclophosphamide are associated with pulmonary fibrosis and may also be associated with FM like other chemotherapeutic agents, including bleomycin (most commonly), busulfan, carmustine and gemcitabine; with incidences ranging from 1% to 10%.^{2,3} A multidisciplinary approach to treatment, which includes screening and early intervention, may be key to management of these cases.

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