Nuclear Medicine and Molecular Imaging (2018) 52:401–404
https://doi.org/10.1007/s13139-018-0545-6

INTERESTING IMAGE

ISSN (print) 1869-3482
ISSN (online) 1869-3474

# Fibrosing Mediastinitis: a Rare Cause of Unilateral Absent Lung Perfusion on a V/Q Scan

Alyssa R. Goldbach[1] · Suzanne Pascarella[2] · Simin Dadpravarar[1]

Received: 31 May 2018 / Revised: 5 August 2018 / Accepted: 22 August 2018 / Published online: 3 September 2018


## Abstract

We report a case of a 29-year-old female with a history of asthma, post-partum ARDS, and pulmonary hypertension who presents with severe shortness of breath. The patient describes her shortness of breath as progressive over the past 10 years. Chest radiography and CT angiography of the thorax showed findings consistent with fibrosing mediastinitis with severe stenosis of the left main pulmonary artery. This resulted in appearance of unilateral absent left lung perfusion on quantitative Tc-99-MAA perfusion and Xe-133 ventilation (V/Q) scan.

**Keywords** Ventilation and perfusion scan · Fibrosing mediastinitis · CTPA · Histoplasma · Mediastinal fibrosis

✉ Simin Dadpravarar
sdadparvar@gmail.com

[1] Division of Nuclear Medicine and Molecular Biology, Department of Radiology, Temple University Hospital, 3401 N. Broad Street, Philadelphia, PA 19104, USA

[2] Division of Breast Imaging, Department of Radiology, Temple University Hospital, 3401 N. Broad Street, Philadelphia, PA 19104, USA

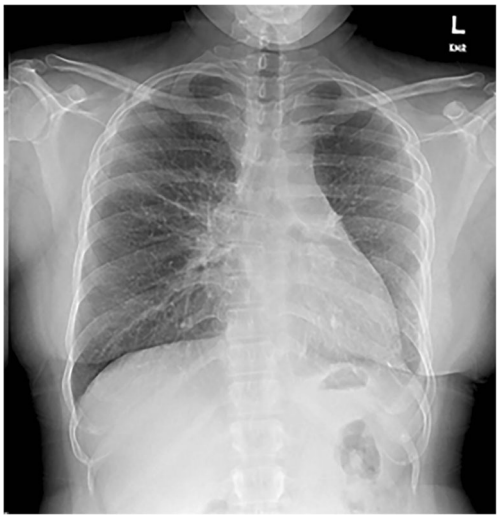


**Fig. 1** A 29-year-old female, originally from the Southeastern USA, status post deployment in Iraq with the military 7 years ago with a history of asthma, post-partum ARDS, and pulmonary hypertension presents with severe shortness of breath. In 2009, the patient reported that she underwent mediastinoscopy for mediastinal adenopathy; however, pathology was negative for monoclonal B-cells. In November 2016, status post normal vaginal delivery, the patient developed shortness of breath and was hospitalized for ARDS. AP radiograph of the chest shows calcified mediastinal and left hilar lymph nodes and a calcified granuloma in the left lower lobe compatible with sequela of chronic granulomatous disease. There are very faint parenchymal opacities seen scattered throughout the left lung and in the right upper lung zone. There is mildly decreased volume of the left hemithorax and mild diffuse left-sided pleural thickening

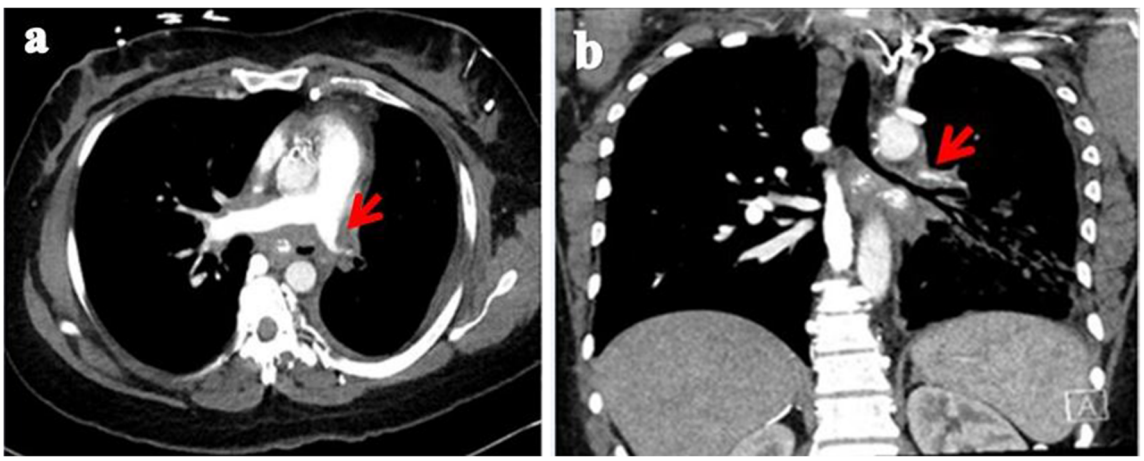


**Fig. 2** Axial (**a**) and coronal (**b**) soft tissue algorithm contrast-enhanced CT of the chest shows calcified lymph nodes and soft tissue encasing a severely narrowed left main pulmonary artery (solid arrow). Also noted are extensive groundglass infiltration of the left lung and milder groundglass infiltrates in the right upper lobe (not pictured). The constellation of imaging findings and reported surgical biopsy in 2009 that was negative for malignancy and non-caseating granulomas are suggestive of fibrosing mediastinitis. These findings are likely sequelae of prior granulomatous disease, most likely histoplasmosis, given the patient's antibodies to Histoplasma Mycelial. The fibrotic reaction characteristic of fibrosing mediastinitis was potentially incited by patient's exposure to dust while deployed with the military in Iraq

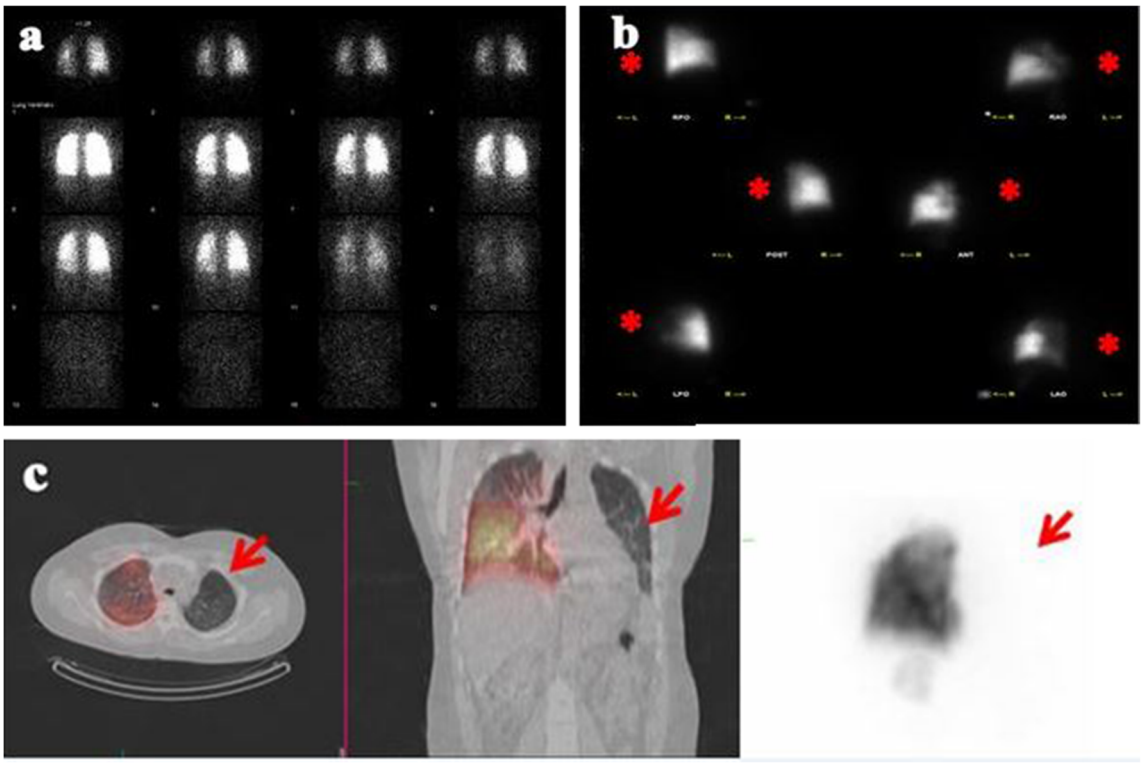


**Fig. 3** Xenon 133 ventilation (**a**) images show symmetric ventilation to the lungs without gas trapping bilaterally. T*echnetium-99m MAA* planar (**b**) and SPECT/CT fusion (**c**) perfusion images show absent perfusion to the left lung (solid arrow). Absent unilateral left lung perfusion is due to severe stenosis of the left main pulmonary artery secondary to encasement by calcified lymph nodes and soft tissue as seen on CT. The left main stem bronchus, however, remains patent, allowing the left lung to be normally ventilated. Quantification values demonstrated that the left lung contributes 3% and the right lung 97% of total lung perfusion. Clinically the patient was diagnosed with fibrosing mediastinitis and is currently listed for lung transplant. Fibrosing mediastinitis was first described by Oulmont in 1855 as an uncommon sequela of prior granulomatous diseases, most commonly Histoplasmosis in the USA; however, other etiologies such as infectious, autoimmune, and idiopathic inflammatory processes have also been proposed [1–5]. It is a rare benign disorder resulting from proliferation of acellular collagen and fibrous tissue within the mediastinum [1, 6]. Fibrosing mediastinitis results in infiltrating mass-like fibrosis and mediastinal lymphadenopathy which often encases and compresses mediastinal structures, including the tracheobronchial tree, esophagus, vena cava, pulmonary veins, and pulmonary arteries [3, 4, 6, 7]. The compression of mediastinal structures can lead to severe cardiopulmonary symptoms [3, 6]. It has been suggested that there are two subtypes of fibrosing mediastinitis, focal and diffuse. The focal variant typically presents as localized calcified mass in the paratracheal or subcarinal region and the diffuse variant as infiltrating, often non-calcified, masses involving multiple mediastinal compartments [1, 3, 8]. Traditionally, medical and surgical treatment for pulmonary arterial disease have been notoriously difficult and ineffective with mortality rates approaching 50% when both pulmonary arteries are involved in symptomatic patients [9, 10]. Most patients succumb to severe pulmonary hypertension and cor pulmonale [11]. The case presented here shows that fibrosing mediastinitis is a possible, although uncommon, cause of unilateral absent lung perfusion on V/Q scan. Additional differential considerations unilateral absent lung perfusion includes obstructing lesions such as carcinoma and mucous plugging, extrinsic compression due to thoracic aortic aneurysm and rare causes such as Sywer-James syndrome [12]

## Compliance with Ethical Standards

**Conflict of Interest** Alyssa Goldbach, Suzanne Pascarella, and Simin Dadpravar declare that they have no conflict of interest.

**Ethical Statement** All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed Consent** Institutional review board of our institute approved this retrospective study, and the requirement to obtain informed consent was waived.

## References

1. Rossi SE, McAdams HP, Rosado-de-Christenson ML, Franks TJ, Galvin JR. Fibrosing mediastinitis. Radiographics. 2001;21:737–57.
2. Loyd JE, Tillman BF, Atkinson JB, Des Prez RM. Mediastinal fibrosis complicating histoplasmosis. Medicine (Baltimore). 1988;67:295–310.
3. Sherrick AD, Brown LR, Harms GF, Myers JL. The radiographic findings of fibrosing mediastinitis. Chest. 1994;106:484–9.
4. Kandzari DE, Warner JJ, O'Laughlin MP, Harrison JK. Percutaneous stenting of right pulmonary artery stenosis in fibrosing mediastinitis. Catheter Cardiovasc Interv. 2000;49:321–4.

5. Oulmont N. Des oblitérations de la veine cave supérieure. Paris: J.B. Ballière; 1855.
6. Thiessen R, Matzinger F, Seely J, Aina R, MacLeod P. Fibrosing mediastinitis: successful stenting of the pulmonary artery. Can Respir J. 2008;15:41–4.
7. Gustafson MR, Moulton MJ. Fibrosing mediastinitis with severe bilateral pulmonary artery narrowing: RV–RPA bypass with a homograft conduit. Tex Heart Inst J. 2012;39:412–5.
8. Dunn EJ, Ulicny KS, Wright CB, Gottesman L. Surgical implications of sclerosing mediastinitis: a report of six cases and review of the literature. Chest. 1990;97:338–46.
9. Kadiev S, Daniels C, Sparks E, Hill S, Cheatham J, Diaz P. Histoplasmosis-induced fibrosing mediastinitis with severe right pulmonary artery stenosis successfully managed with cutting balloon angioplasty and endovascular stent placement. Chest. 2007;132:729A.
10. Doyle TP, Loyd JE, Robbins IM. Percutaneous pulmonary artery and vein stenting. A novel treatment for mediastinal fibrosis. Am J Respir Crit Care Med. 2001;164:657–0.
11. Smith JS, et al. Pulmonary artery stenosis secondary to fibrosing mediastinitis: management with cutting balloon angioplasty and endovascular stenting. Vasc Endovasc Surg. 2011;45:170–3.
12. Chow B, Wittram C, Lee VW. Unilateral absence of pulmonary perfusion mimicking pulmonary embolism. Am J Roentgenol. 2001;176(3):712.