



Case Report

Limited Cutaneous Systemic Sclerosis Presenting as a Solitary Lung Mass

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Case Report

We present the case of a 70-year-old female with history of hypertension, gastro esophageal reflux disease, and irritable bowel syndrome (IBS) who had presented with gradually worsening fatigue and exertional dyspnea for 9 months. Initial evaluation by her primary care physician revealed a large left upper lobe lung mass on chest x-ray. CT chest revealed a large bilobed mass lesion located in the left upper lobe of the lung measuring 9.4 x 6.3 x 9.6 cm, with internal foci of calcification and mildly enlarged mediastinal and hilar adenopathy. Calcified right lower lobe granulomas and diffuse bilateral ground glass opacification was also found in addition to mild lower lobe predominant bronchiectasis and mosaic attenuation. CT-guided core needle biopsy of the mass revealed thickening of the intima and media of pulmonary arteries by hyalinised fibrous tissue along with fibroblast proliferation. Congo red stain was negative for amyloid deposition and there was no evidence of malignancy. The findings were reported as being consistent with systemic sclerosis. Pulmonary function test and echocardiography was performed. PFTs revealed moderate restrictive pulmonary disease with a severe defect in diffusion with FEV1/ FVC – 85, FVC- 1.73L (59%), TLC- 3.42L (70%) and DLCO – 36%. The echocardiogram was suggestive of severe pulmonary hypertension with pulmonary artery pressure of 80-90 mmHg, which was confirmed by right heart catheterization showing mean pulmonary artery pressure of 57mm Hg and pulmonary artery occlusive pressure of 18mm Hg with partial vasodilatory response. A simultaneous rheumatologic evaluation revealed a

history suggestive of Raynaud's phenomenon for the last 8 years. She denied morning stiffness, skin thickening, arthralgia or joint swelling. Physical exam was notable for multiple telangiectasias on the hard palate and hands. No rashes or skin thickening was identified. Laboratory evaluation was significant for strongly positive for anti-nuclear antibody (ANA) titer >1280, anti SSA antibody, anti-centromere antibody (>8); anti-cyclic citrullinated peptide antibody (CCP) + (18.9) and rheumatoid factor (RF) (188) which was suggestive of limited diffuse systemic sclerosis. Other tests including anti-double stranded DNA, anti-Smith, anti-RNP (ribonucleotide protein), anti-SSB, anti Scl 70 antibody, HIV and hepatitis screen were negative. Joint survey revealed mild to moderate osteoarthritis arthritis (OA) involving bilateral carpocarpal, distal interphalangeal joints and metatarsophalangeal joints. The initial concern was for a malignancy with paraneoplastic systemic sclerosis, but the biopsy was not suggestive of such. She was diagnosed with limited cutaneous systemic sclerosis. She was treated with continuous IV treprostinil with pulmonary follow up showing improvement in 6-minute walk distance. Hydroxychloroquine was started with a short course of oral prednisone for inflammatory arthropathy and amlodipine for Raynaud's disease with improvement in symptoms. Repeat CT scan 18 months after the initial study showed stability of the lung mass (Figures 1,2&3).

Keywords: Lung Mass; Pulmonary Artery Hypertension; Systemic Sclerosis; Interstitial Lung Disease.

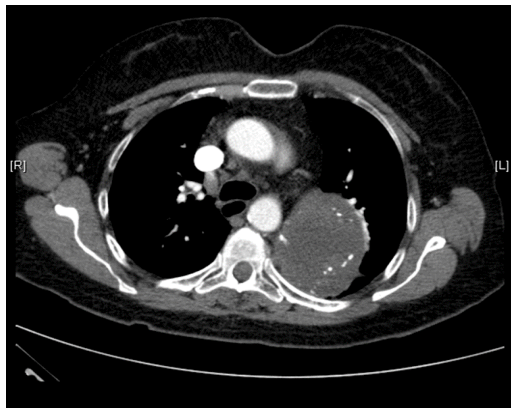


Figure 1: CT chest with and without contrast showing the left upper lobe mass. Note discrete margins of the lesion with no invasion into adjacent tissues and scattered areas of calcification.



Figure 2: CT chest with and without contrast showing the same left upper lobe mass lesion in the coronal section. Note truncated appearance of pulmonary arteries consistent with the diagnosis of pulmonary artery hypertension.

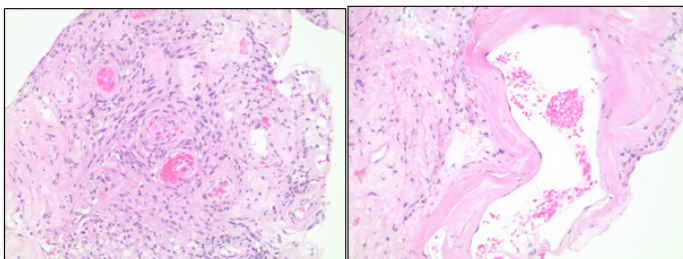


Figure 3: Pathology-Hematoxylin and Eosin stain showing hyalinised arteries and fibrous tissue.

Discussion

Systemic sclerosis (SSc) is a multisystem disease characterized by activation of the immune system resulting in fibroblast proliferation and excessive collagen formation with

development of progressive fibrosis of the skin and internal organs [1] along with vasculopathy characterized by arterial intimal proliferation capillary dilatation and destruction [2]. The estimated incidence in the United States is 20 per million per year with a prevalence rate of 240 per million [3]. There are multiple ways in which the disease presents itself but fatigue, stiff joints, loss of strength and pain was seen more commonly. Less frequently reported symptoms included dyspnea, GERD like symptoms, depression, and weight loss [4]. In our case, the patient presented with dyspnea and the initial evaluation led to the surprising discovery of a large lung mass with an ultimate diagnosis of systemic sclerosis. A localized area of sclerosis in the lung due to systemic sclerosis presenting as a lung mass has not been previously reported in the literature. It is extremely important to consider malignancy in the differential as there have been sporadic cases reported of lung cancer in the setting of systemic sclerosis [5,6]. There are rare reports of mass lesions in the setting of systemic sclerosis with alternative diagnoses such as tumoral calcinosis (deposition of dense, calcified masses, often lobulated in appearance, usually around large joints) [7,8] and in one case due to an infection from actinomycosis [9] (which is a branching gram-positive bacteria). The underlying mechanism for the development of a large solitary lung mass is unclear but can be attributed to T cell infiltration, excessive fibroblast proliferation and exaggerated type I collagen deposition, which is the hallmark of systemic sclerosis [1].

Conclusion

The evaluation of a lung mass can present a diagnostic challenge and it is extremely important to rule out an underlying malignant cause. In rare instances, patients can present with a lung mass due to a non-malignant cause from systemic diseases, as was the case in our patient with systemic sclerosis.

References

1. Systemic sclerosis: current pathogenetic concepts and future prospects for targeted therapy. *Lancet*. 347:1453-1458.
2. LeRoy EC. (1992) A brief overview of the pathogenesis of scleroderma (systemic sclerosis). *Annals of the rheumatic diseases*. 51:286-288.
3. Mayes MD. (2003) Scleroderma epidemiology. *Rheumatic diseases clinics of North America*. 29:239-254.
4. Sandusky SB, McGuire L, Smith MT, Wigley FM, Haythornthwaite JA. (2009) Fatigue: an overlooked determinant of physical function in scleroderma. *Rheumatology*. 48:165-169.
5. Rosenthal AK, McLaughlin JK, Linet MS, Persson I. (1993) Scleroderma and malignancy: an epidemiological study. *Annals of the rheumatic diseases*. 52:531-533.
6. Talbot JH, Barocas M. (1979) Progressive systemic sclerosis (PSS) and malignancy, pulmonary and non-pulmonary. *Medicine*. 58:182-207.

Citation: Iftqar S, Schmidt P (2023) Limited Cutaneous Systemic Sclerosis Presenting as a Solitary Lung Mass. Ann Case Report. 8: 1484. DOI:10.29011/2574-7754.101484

7. Olsen KM, Pike EJ, Chew FS. (2004) Progressive systemic sclerosis with massive paraspinal soft-tissue calcinosis. AJR. American journal of roentgenology. 183:634.
8. Tuy BE, John TK, Ugliarolo AD, Beebe KS, Vives MJ, et al (2008) Tumoral calcinosis presenting as neck pain and mass lesion of the cervical spine. American journal of orthopedics. 37:E191-195.
9. AR A, JH R, W W, SW W, JF F. (2011) An incidental finding of a lung lesion in a patient with systemic sclerosis. Respiratory Medicine CME 4:178-180.