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Spontaneous Pneumomediastinum, Retroperitoneal Air, and Subcutaneous Emphysema in Patient with Connective Tissue Disease Associated Interstitial Lung Disease

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Abstract

Spontaneous pneumomediastinum (SPM) and subcutaneous emphysema are rare complications of various autoimmune conditions, most commonly seen in inflammatory myositis. We report the first case of SPM, subcutaneous emphysema and air in the retroperitoneum and bladder wall in a patient with interstitial lung disease with an overlap of inflammatory myositis and Sjogren's syndrome.

The patient is a 64-year-old Polish female with a recent diagnosis of interstitial lung disease as an incidental finding on imaging, initially thought to be due to dermatomyositis (DM) or Sjogren's disease due to a positive anti-Jo antibody and SSA antibody at an outside hospital. Six months after initial diagnosis, the patient was somnolent and lethargic at home and was found to have extensive subcutaneous emphysema, air in the bladder wall, and pneumomediastinum based on chest x-ray and CT scan findings. The patient was treated with broad spectrum antibiotics for initial concern for infection, but no source was identified. Repeat serologies at our hospital revealed a negative myositis panel including Melanoma Differentiation-Associated protein 5 (MDA5) antibodies and anti-Jo antibodies, but persistently elevated SSA antibodies.

Keywords: Pneumomediastinum, Subcutaneous emphysema, Interstitial lung disease, Sjogren's syndrome, Dermatomyositis

Introduction

Interstitial lung disease (ILD) is a commonly recognized complication of various connective tissue diseases, including inflammatory myopathies, Sjogren's disease, and scleroderma. Occasionally, ILD progresses rapidly and can subsequently lead to subcutaneous emphysema, pneumothorax, and pneumomediastinum [1]. These rare but serious complications are under-recognized in both internal medicine and rheumatology.

Pneumomediastinum occurs either as a primary or secondary event. Primary, or spontaneous pneumomediastinum is thought to occur in the setting of increased intrathoracic pressure, causing alveolar rupture and air leakage. The more commonly seen secondary pneumomediastinum occurs as sequela of blunt or penetrating trauma, recent interventions to the esophagus or tracheobronchial tree, tissue dissection, rupture of a hollow viscus, or by gas forming organisms infecting the lungs or mediastinum [2].

The purpose of this report is to recognize spontaneous pneumomediastinum as a catastrophic complication of connective tissue disease, and to elucidate the clinical findings, laboratory findings, and risk factors for the development of SPM and subcutaneous emphysema. To our knowledge, there are no previously documented cases of SPM with involvement of other visceral structures, or with such broad antibody positivity [2].

Case Report

A 64-year-old female with a recent diagnosis of autoimmune interstitial lung disease was admitted for somnolence, lethargy, weakness, confusion, hallucinations, and new rash. On admission, a chest x-ray and subsequent computerized tomography (CT) scan revealed extensive subcutaneous emphysema in the neck, groin, and buttocks, pneumomediastinum, and air in the bladder wall and retroperitoneum. Burn, dermatology, and infectious disease teams were consulted. The patient was started on broad spectrum antibiotics (vancomycin/piperacillin-tazobactam/clindamycin).

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Examination was notable for somnolence, waxing/waning confusion, and a rash suspicious for leukocytoclastic vasculitis on the forearms and lower extremities. Strength was intact. Labs were significant for hyponatremia, elevated creatinine with nephrotic range proteinuria (3 gm), hyponatremia, elevated erythrocyte sedimentation rate (ESR 114 mm) and C-reactive protein (CRP 34.2 mg/dl), elevated aldolase (15.5 U/L), and positive SSA. Serum immunofluorescence was abnormal and immunofixation showed a restricted band of IgG kappa monoclonal protein. Creatinine kinase (CK), antinuclear antibody (ANA), anti-neutrophil cytoplasmic antibody (ANCA), myositis panel, MDA-5, rheumatoid factor (RF), and blood cultures were negative during this hospitalization. Urine cultured from admission grew colonies of E. coli. Non-contrast CT scan showed extensive subcutaneous emphysema in the neck, anterior abdominal and pelvic walls, pneumomediastinum, retroperitoneal air in the region of the iliopsoas muscle, air in the urinary bladder and in the bladder wall. Extensive lung disease was evident on CT, with lower lobe predominant chronic interstitial infiltration and mild bronchiectasis. Two separate punch biopsies of the rash showed minimal perivascular inflammation and hemorrhagic crust not thought to be consistent with vasculitis. Hospital course had several major complications including diverticular perforation and later Cytomegalovirus (CMV) re-activation with a CMV titer of 15829 IU/ml (Figure 1, Figure 2 and Figure 3).

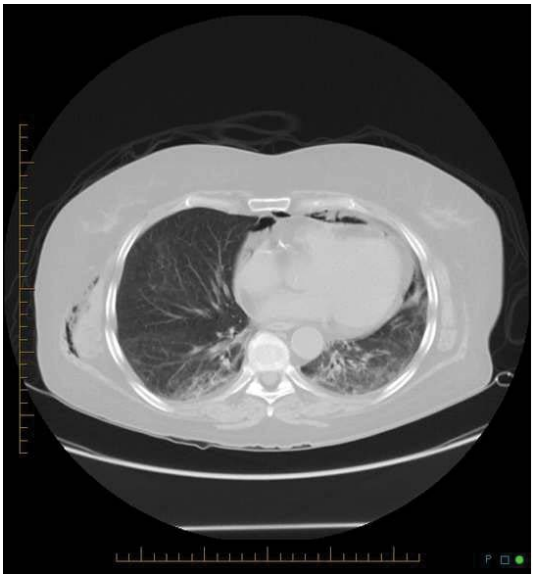

Figure 1: Non-contrast computed tomography scan of chest. Coronal view demonstrates linear interstitial opacities more prominent in the left lung. Spontaneous pneumomediastinum seen between the anterior chest wall and cardia. Subcutaneous emphysema visualized lateral to the right thorax.

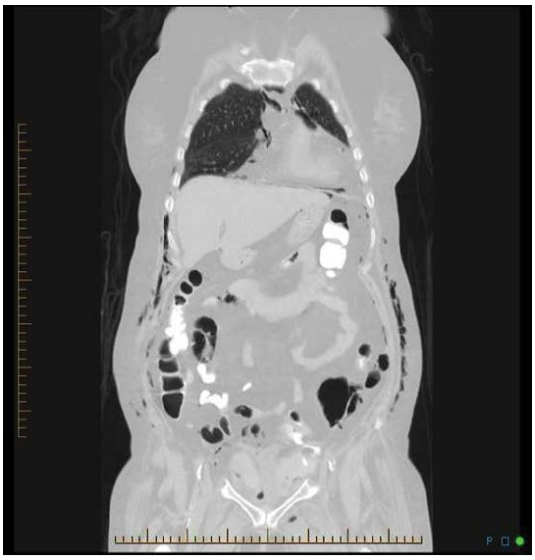

Figure 2: Non-contrast computed tomography scan of chest, abdomen, and pelvis. Axial view demonstrating pneumomediastinum, pneumoperitoneum, and diffuse subcutaneous emphysema.

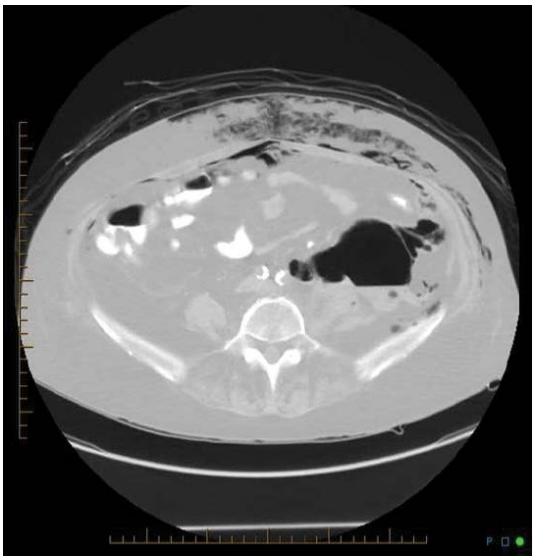

Figure 3: Non-contrast computed tomography scan of the abdomen. Coronal view showing significant subcutaneous emphysema with focal areas of pneumoperitoneum.

Historical review determined the patient was first diagnosed with pulmonary fibrosis at an outside hospital as an incidental finding on CT scan six months prior. Autoimmune workup revealed a positive anti-Jo antibody, SSA antibody, MPO antibody, slightly positive RF, and patient was being treated by the pulmonologist as an anti-synthetase syndrome. Prior pulmonary function testing showed an FEV1 2.0 L, TLC 75% predicted, DLCO 53%. Prior documentation revealed no evidence of weakness or muscle disease and patient was started on azathioprine 50 mg daily and prednisone 40 mg daily with

a taper for presumed non-specific interstitial pneumonia (NSIP) from inflammatory myopathy or Sjogren's disease. During hospitalization, the patient endorsed dry eyes and dry mouth, but denied rashes, fevers, chills, arthralgias, oral/nasal ulcerations, abdominal pain, Raynaud's, miscarriages, clots, or shortness of breath.

Discussion

Spontaneous pneumomediastinum and subcutaneous edema are rare and the differential diagnosis is narrow. It was first described in 1939 by Louis Hamman. Pneumomediastinum has been described more commonly and is usually secondary to either traumatic or iatrogenic cause. SPM can be a result of interstitial lung disease, asthma, chronic obstructive pulmonary disease (COPD), or tobacco use [3]. Patient was noted to have an E. coli urinary tract infection (UTI) which has been largely implicated in causing emphysematous cystitis and intraluminal gas in the bladder, however the localized infection would not explain the extensive involvement. Risk factors for our patient included a long-standing prior history of smoking and corticosteroid use.

Within the context of connective tissue diseases, SPM has been most commonly reported in dermatomyositis, followed by polymyositis, and sclerodermatomyositis. The prevalence of SPM in myositis is reported at 2.2% to 8.6% [4-9]. Furthermore, it is more commonly present in amyopathic dermatomyositis. Normal CKs and occurrence of SPM in patients with DM portends a poor prognosis. Our patient was initially diagnosed and treated as an ILD secondary to dermatomyositis/antisynthetase syndrome due to an initially positive anti-Jo antibody at an outside hospital. She had no other features of DM such as proximal muscle weakness or typical skin findings of Gottron's papules, shawl sign, or heliotrope rash. While her myositis panel and MDA5 antibodies were negative on repeat testing at our hospital and she had no cutaneous or myopathic findings consistent with DM, we would still favor the diagnosis of amyopathic dermatomyositis with a potential overlap with Sjogren's syndrome in the light of her antibody spectrum. The anti-Jo level has been reported to correlate with disease activity and given our patient had been on high doses of prednisone for an extended period of time, it is plausible that the anti-Jo was negative on repeat testing due to treatment. The enzyme-linked immunosorbent assay (ELISA), counter immunoelectrophoresis (CIE) and immunoblotting tests used for anti-Jo-1 testing are highly sensitive and specific which supports a low likelihood of a false positive result [10].

To date, there have been no clinical cases of bladder and retroperitoneal free air associated with dermatomyositis or autoimmune disease. The pathogenesis of SPM is thought to be related to increased intrathoracic pressure leading to an increased alveolar pressure and therefore pressure gradient, subsequently causing alveolar rupture into the interstitium and bronchovascular tissue, and therefore causing pneumomediastinum. Architectural distortion of the lung tissue secondary to ILD leads to rupture of the subpleural blebs especially in the setting of corticosteroid use that leads to weakening of the lung tissue. Vasculopathic lesions can also cause injury of the alveolar wall and lead to air leakage. Risk factors for SPM include ILD, younger age, a cutaneous vasculopathy, lack of CK elevation, history of smoking, and corticosteroid treatment. In this case, the patient had air in the bladder wall as well as in the retroperitoneum, which would be best explained by vasculopathic lesions leading to injury of the bladder wall or an infectious etiology.

Key Points

- Spontaneous pneumomediastinum, subcutaneous emphysema, and pneumoperitoneum are associated with autoimmune conditions, particularly inflammatory myositis and therefore autoimmune workup should be considered in patients with these findings.
- Development of spontaneous pneumomediastinum in a patient with known autoimmune disease portends a poor prognosis, particularly in those patients without CK elevation.

Conflict of Interests

No conflict of interests to disclose.

Statement of Authorship

This is an original manuscript that has not been submitted or published previously.

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