A Spontaneous Subcutaneous Emphysema As a Life Threatening Complication of Rheumatoid Arthritis Associated Interstitial Lung Disease

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Background

Pulmonary leak syndrome with spontaneous subcutaneous emphysema (SSE) in the absence of pneumothorax has rarely been reported in patients with rheumatoid arthritis (RA) associated interstitial lung disease (ILD) [1]. We present the case of a patient diagnosed with RA associated fibrosing ILD who presented with life threatening SSE.

Case Description

A 68-year-old female known to have RA and associated ILD presented with progressively worsening shortness of breath, cough and worsening facial swelling extending to the neck and upper chest wall for two days. The patient denied any associated chest pain, fever, chills, sputum, lightheadedness, trauma or syncopal episodes. She had been diagnosed with RA associated fibrosing ILD two years prior. The patient had been on home oxygen, pirfenidone and was offered biopsy in the past, which she had refused. She had also been on chronic prednisone therapy five milligrams per day (mg/day). She had not been on any other immunosuppressants. On presentation the patient was not in any distress but was desaturating, requiring six liters supplemental oxygen to maintain acceptable saturation. She had palpable crepitations extending from the mid-chest and axillary area to lower eye lids suggestive of extensive subcutaneous emphysema. Laboratory data was unremarkable. Imaging studies (Figure 1A,B,C) did confirm subcutaneous emphysema and pneumomediastinum (PM) without any pneumothorax.

Case Description (continued)

The patient was managed conservatively with supplemental oxygen and steroid therapy. Her condition continued to deteriorate with worsening subcutaneous emphysema without any evident pneumothorax on imaging studies. She required intubation and mechanical ventilation secondary to worsening hypoxia. Eventually, given no clinical improvement, the patient was transitioned to comfort care and passed.

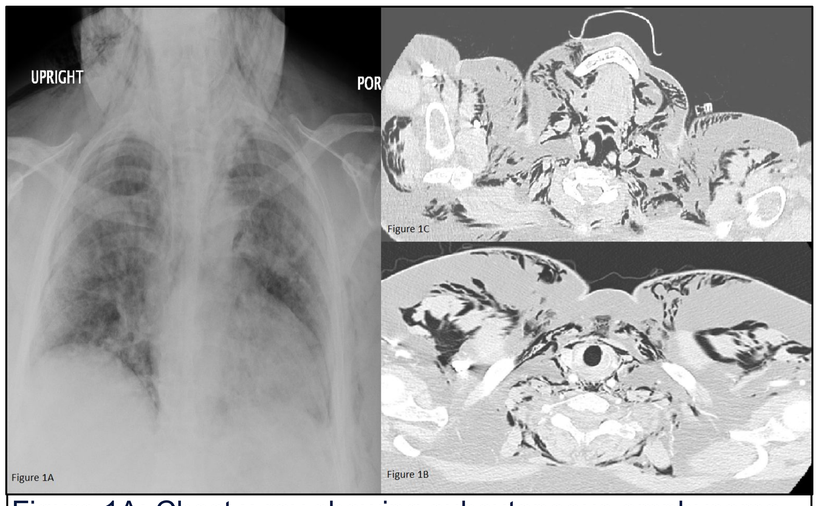

Figure 1A: Chest x-ray showing subcutaneous emphysema (SSE). Figure 1B: Chest computer tomography (CT) scan showing extend of SSE. Figure 1C: Worsening of chest CT scan showing extend of SSE.

Discussion

SSE and PM are commonly associated with any condition that creates a gradient between intra-alveolar and perivascular interstitial pressure [2]. They are extremely rare in patients with RA associated fibrosing ILD. Most cases of SSE without pneumothorax will not have any associated chest pain, as reported in our case. Management can be extremely challenging. Common known risk factors for SSE in patients with connective tissue disease include the presence of extensive ILD, prolonged steroid and/or immunosuppressive therapy, vasculopathy and rheumatoid nodules. Despite the size and rapidity of accumulation of subcutaneous emphysema, conservative management has been described to be effective in the absence of any significant pneumothorax. Unfortunately, conservative therapy failed to meaningfully recover our patient. It is important to recognize SSE and pulmonary leak syndrome as an important, but rare, life threatening manifestation of RA associated fibrosing ILD.

References

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This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

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