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SPONTANEOUS PNEUMOMEDIASTINUM IN RHEUMATOID ARTHRITIS ASSOCIATED INTERSTITIAL LUNG DISEASE AND BRONCHIECTASIS: A THREE-COMPONENT PATHOPHYSIOLOGIC FRAMEWORK

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ABSTRACT

Spontaneous pneumomediastinum is a recognized complication of connective tissue disease associated interstitial lung disease, most extensively described in dermatomyositis. Despite rheumatoid arthritis being the most common connective tissue disease with pulmonary involvement, spontaneous pneumomediastinum in rheumatoid arthritis associated lung disease has been reported only once previously. Rheumatoid arthritis uniquely causes two distinct forms of structural lung injury (interstitial lung disease and bronchiectasis) both linked to seropositive autoimmunity.

An 83-year-old woman, a never-smoker with seropositive rheumatoid arthritis, progressive pulmonary fibrosis, left lower lobe bronchiectasis, low body mass index, and iatrogenic lymphopenia developed extensive pneumomediastinum during community-acquired pneumonia. Computed tomography scan demonstrated pneumomediastinum with subcutaneous emphysema, emphysematous lungs with diffuse interstitial scarring, bronchiectasis, and lobar consolidation. Conservative management with antibiotics achieved complete radiographic resolution at 8 weeks with no recurrence at 12 weeks.

We hypothesize a three-component pathophysiologic framework: structural alveolar fragility from interstitial lung disease and emphysema may provide the substrate for alveolar rupture, while infection-driven coughing from true airway bronchiectasis may serve as the mechanical trigger via the Macklin effect. In rheumatoid arthritis, all three components appear driven by a shared seropositive autoimmune process. This mechanistic profile may differ fundamentally from dermatomyositis, where spontaneous pneumomediastinum is thought to reflect rapidly progressive parenchymal destruction, suggesting that dermatomyositis-derived mortality data may not be directly applicable to rheumatoid arthritis.

Spontaneous pneumomediastinum in seropositive rheumatoid arthritis may follow a favourable trajectory when the mechanical trigger is reversible. High titer seropositivity, low body mass index, and iatrogenic immunosuppression may identify a convergent risk phenotype.

KEYWORDS

Rheumatoid arthritis, pneumomediastinum, interstitial lung disease, bronchiectasis, Macklin effect

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LEARNING POINTS

- Seropositive rheumatoid arthritis uniquely drives two distinct forms of structural lung injury (interstitial lung disease [ILD] and bronchiectasis) that may converge to create a substrate for spontaneous pneumomediastinum via the Macklin effect. Internists should recognize that rheumatoid arthritis (RA) patients with high-titer rheumatoid factor and anti- cyclic citrullinated peptide are at increased risk for both manifestations, and their coexistence may lower the threshold for alveolar rupture during infection-driven coughing.
- Not all pneumomediastinum in connective tissue diseases carries the same prognosis. When spontaneous pneumomediastinum (SPM) occurs in the setting of a reversible mechanical trigger such as acute pneumonia, conservative management with targeted antibiotics may achieve complete resolution, in contrast to the progressive-parenchymal phenotype seen in dermatomyositis, where SPM independently predicts mortality.
- Consider low-threshold computed tomography imaging in seropositive RA patients with known ILD and bronchiectasis who present with acute respiratory infections and chest or neck discomfort, even without the Hamman sign. Transient pneumomediastinum may be underrecognized in this population because symptoms are attributed to infection alone, and repeat imaging is not routinely performed.

INTRODUCTION

Spontaneous pneumomediastinum (SPM) occurs when alveolar rupture allows air to dissect along bronchovascular sheaths into the mediastinum; the Macklin effect[1]. Primary SPM in young healthy adults is typically benign, while secondary SPM in patients with structural lung disease carries a significantly worse prognosis[2]. Among connective tissue diseases (CTDs), SPM has been most extensively described in dermatomyositis- interstitial lung disease (ILD), where Jung et al. found SPM in 12/70 (17.1%) patients with myositis-associated ILD, independently predicting mortality (HR 2.83, 95% CI 1.1–7.27)[3].

Per our search, we identify only one prior rheumatoid arthritis (RA)-specific case, despite RA being the most common CTD with pulmonary involvement[4]. This underrepresentation may reflect underrecognition rather than true rarity, as RA-ILD patients with concurrent bronchiectasis may not undergo repeat imaging sufficient to detect transient pneumomediastinum, particularly if symptoms are attributed to infection alone. This disparity is notable because RA uniquely causes two distinct forms of structural lung injury (ILD and bronchiectasis) both linked to seropositive autoimmunity. RA-ILD and RA-bronchiectasis each have a pooled prevalence of approximately 18.7%, with rheumatoid factor (RF) and anti- cyclic citrullinated peptide (CCP) positivity independently associated with both manifestations[5-7]. We present a case of secondary SPM in RA and hypothesize a three-component pathophysiologic framework that may help explain the development of SPM in this clinical context.

CASE DESCRIPTION

An 83-year-old woman, a never-smoker with seropositive RA of approximately 20 years' duration, presented to an outside emergency department after a ground-level fall with head trauma, then was transferred to a receiving facility. Her RA was managed with hydroxychloroquine 200 mg daily, mycophenolate mofetil 1,000 mg twice daily, and prednisone 5 mg daily, after prior methotrexate discontinuation due to pulmonary fibrosis concerns. Mycophenolate had been initiated for progressive fibrosis refractory to hydroxychloroquine monotherapy. Pulmonary fibrosis had been diagnosed in November 2019 with serial computed tomography (CT) scan progression (2019, 2021, 2023). Nintedanib 150 mg twice daily had been ordered but not been initiated. She had left lower lobe bronchiectasis with true airway disease features: recurrent infections requiring escalating antibiotics, chronic productive cough, and a prior emergency department visit for respiratory infection. Serial forced expiratory volume in one second (FEV1) had declined from 2.07 l to 1.60 l. Forced vital capacity and diffusing capacity of the lung for carbon monoxide were unavailable. Body mass index (BMI) was 14.85 kg/m^2.

Vital signs on presentation: blood pressure 118/72 mmHg, heart rate 71 bpm, respiratory rate 18 breaths per minute, T 36.9°C, oxygen saturation 100% on room air. Examination revealed decreased breath sounds bilaterally. The patient was disoriented; complete history was unobtainable. RF 280 IU/ml, anti-CCP >250 U/ml, Myositis-specific antibody panel was negative. Key laboratory findings are presented in *Table 1*.

CT chest demonstrated extensive pneumomediastinum and subcutaneous emphysema without pneumothorax; emphysematous lungs with diffuse interstitial scarring; left lower lobe bronchiectasis; and dense peripheral left lower lobe consolidation with air bronchograms consistent with pneumonia *(Fig. 1)*. No acute rib fractures were identified. Sputum culture grew rare yeast with normal flora; no bacterial pathogen was isolated. Oesophageal rupture was excluded based on the absence of oesophageal wall abnormality, pleural effusion, or oral contrast extravasation on CT scan. Echocardiography demonstrated preserved biventricular function (ejection fraction 55–60%) with normal right ventricular systolic pressure (26 mmHg), excluding significant pulmonary hypertension.

The clinical presentation, acute lobar consolidation, neutrophilic leucocytosis, C-reactive protein 108.7 mg/l, and rapid antibiotic response, was most consistent with bacterial community-acquired pneumonia. Traumatic pneumomediastinum was considered unlikely given the low-energy mechanism, absence of rib fractures or pneumothorax, and concurrent pneumonia with marked inflammation.

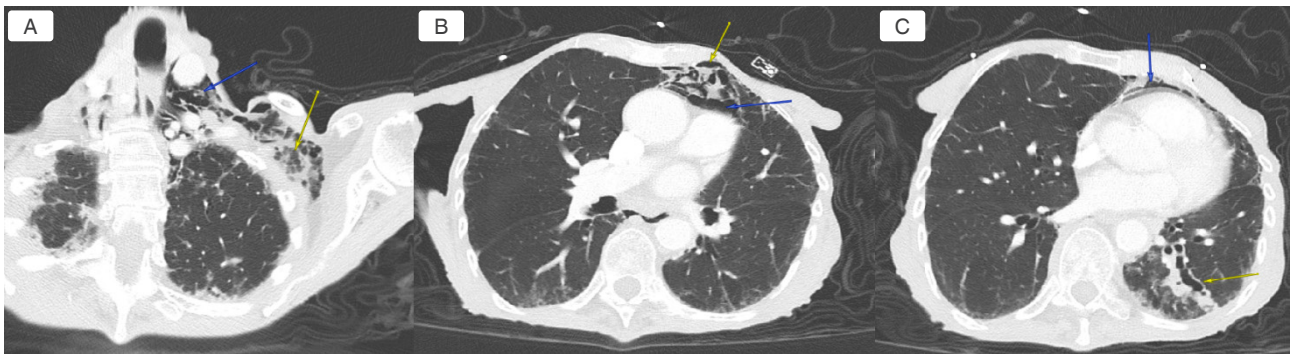


Figure 1. Axial contrast-enhanced computed tomography scan of the chest at presentation. A) Thoracic inlet: pneumomediastinum (blue arrow) and subcutaneous emphysema (yellow arrow) with bilateral emphysema and interstitial scarring. B) Upper chest: subcutaneous emphysema (yellow arrow) and pneumomediastinum (blue arrow). C) Lower chest: pneumomediastinum (blue arrow) and left lower lobe bronchiectasis within dense consolidation consistent with pneumonia (yellow arrow). No pneumothorax.

Parameter	Value	Reference range	Clinical significance
Inflammatory markers			
CRP (mg/l)	108.7	10	Marked acute inflammation consistent with bacterial pneumonia
ESR (mm/hr)	67	0–30	Elevated; nonspecific
Procalcitonin (ng/ml)	0.17	0.10	Mildly elevated; low sensitivity in immunosuppressed patients
Immunologic	-	-	-
Rheumatoid factor (IU/ml)	280	14	High-titer; independently predicts RA-ILD (HR 1.68 for RF >90 IU/ml) and bronchiectasis (2.34-fold risk, seropositive)
Anti-CCP (U/ml)	>250	20	High-titer; independently associated with both RA-ILD and RA-bronchiectasis
Myositis-specific antibody panel	Negative	Negative	Excludes overlap myositis as contributor to ILD or SPM
Hematologic	-	-	-
WBC ($\times 10^3/\mu l$)	12.2	4.5–11	Neutrophilic leukocytosis supporting bacterial infection
Absolute neutrophil count ($\times 10^3/\mu l$)	10.9	1.8–7.7	Elevated; consistent with acute bacterial process
Absolute lymphocyte count ($\times 10^3/\mu l$)	0.55	1–4.8	Iatrogenic lymphopenia (mycophenolate + prednisone); infection risk factor
Haemoglobin (g/dl)	11.2	12–16	Mild anaemia; chronic disease
Metabolic/organ function			
LDH (U/l)	146	120–246	Normal; contrasts with elevated LDH threshold (378 U/l) predictive of SPM in DM-dominated cohorts
Lactate (mmol/l)	1.2	0.5–2	Normal; excludes hemodynamic compromise or tissue hypoperfusion
Creatinine (mg/dl)	0.6	0.6–1.2	Normal renal function
Albumin (g/dl)	2.8	3.5–5	Low; consistent with chronic disease, malnutrition (BMI 14.85 kg/m^2)
Microbiology	-	-	-
Sputum culture	Rare yeast, normal flora	No pathogen	Negative bacterial culture; common in hospitalized CAP (~38% pathogen detection)

Table 1. Key laboratory findings at presentation.

Abbreviations: CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; CCP, cyclic citrullinated peptide; WBC, white blood cells; LDH, lactate dehydrogenase; RA, rheumatoid arthritis; ILD, interstitial lung disease; HR, hazard ratio; RF, rheumatoid factor; SPM, spontaneous pneumomediastinum; DM: dermatomyositis; BMI, body mass index; CAP, community-acquired pneumonia.

Conservative management was initiated: ceftriaxone 1,000 mg intravenously (IV) daily plus azithromycin 500 mg IV daily, bronchodilators, home prednisone continued, and mycophenolate held. Oxygen was weaned to room air by day 2. The patient was discharged home on day 5. CT at 8 weeks demonstrated complete SPM resolution. At 12-week follow-up, there was no recurrence, and mycophenolate was successfully restarted. The clinical timeline is presented in *Table 2*.

DISCUSSION

We hypothesize that SPM in RA may result from the convergence of three components: 1) structural substrate from ILD; fibrosis creates architectural distortion susceptible to alveolar rupture; 2) structural substrate from emphysema; alveolar wall destruction lowers the rupture threshold; and 3) a dynamic trigger from infection-driven cough; true airway bronchiectasis generates recurrent infections and violent cough, providing the acute intrapulmonary pressure elevation needed for alveolar rupture via the Macklin effect[1]. At the fibrotic-emphysematous interface, differential alveolar compliance between stiff fibrotic tissue and hyperinflated emphysematous tissue may create stress concentration during cough-induced pressure transients, potentially further lowering the threshold for alveolar rupture. While this biomechanical rationale is plausible, it remains speculative in the absence of direct physiologic measurement, and alternative explanations for alveolar rupture in this setting cannot be excluded *(Fig. 2)*.

What may distinguish RA from other CTDs is that the same seropositive autoimmune process appears to drive all three components. Seropositive RA confers 2.34-fold increased bronchiectasis risk (95% CI 2.20–2.49) versus 1.56-fold in seronegative RA (6). RF titers >90 IU/ml independently predict incident RA-ILD (HR 1.68, 95% CI 1.02–2.77)[7]. Emphysema occurs in 27% of RA-ILD never-smokers and is independently associated with mortality[8]. Our patient's RF 280 IU/ml and anti-CCP >250 U/ml place her in the highest risk category for all three manifestations.

Evidence supporting pneumonia as the proximate trigger includes temporal association with acute pneumonia,

Time Point	Event	Key findings/interventions
Longitudinal history		
~20 years prior	RA diagnosis	Seropositive (RF 280 IU/ml, anti-CCP >250 U/ml); treatment initiated
November 2019	Pulmonary fibrosis diagnosed	CT: interstitial lung disease; methotrexate discontinued due to fibrosis concerns
2019–2023	Serial CT progression	Progressive fibrosis on CT (2019, 2021, 2023); mycophenolate initiated for fibrosis refractory to hydroxychloroquine monotherapy
Prior to admission	Bronchiectasis established	Left lower lobe bronchiectasis with recurrent infections, chronic productive cough, prior ED visit; serial FEV1 decline 2.07 → 1.60l
Prior to admission	Nintedanib ordered	150 mg twice daily ordered but not yet initiated
Acute Presentation	-	-
Day 0	ED presentation	Ground-level fall with head trauma → transfer to receiving facility
Day 0	Initial assessment	BP 118/72 mmHg, HR 71 bpm, RR 18 breaths per minute, T 36.9°C, SpO_2 100% RA; BMI 14.85 kg/m²; disoriented
Day 0	CT chest (contrast-enhanced)	Extensive pneumomediastinum with subcutaneous emphysema; emphysematous lungs with diffuse interstitial scarring; LLL bronchiectasis; LLL consolidation with air bronchograms; no pneumothorax; no rib fractures
Day 0	Laboratory findings	CRP 108.7, WBC 12.2, ALC 0.55, LDH 146, lactate 1.2; see *Table 1*
Day 0	Echocardiography	EF 55–60%, RVSP 26 mmHg; no pulmonary hypertension
Day 0	Esophageal rupture excluded	No esophageal wall abnormality, pleural effusion, or contrast extravasation
Day 0	Treatment initiated	Ceftriaxone 1,000 mg IV daily + azithromycin 500 mg IV daily; bronchodilators; home prednisone 5 mg continued; mycophenolate held
Day 2	Clinical improvement	Oxygen weaned to room air
Day 5	Discharge	Discharged home; clinical improvement
Follow-Up		
Week 8	CT chest	Complete resolution of pneumomediastinum and subcutaneous emphysema
Week 12	Outpatient follow-up	No SPM recurrence: mycophenolate successfully restarted

Table 2. *Clinical timeline.*

Abbreviations: RA, rheumatoid arthritis; RF, rheumatoid factor; CCP, cyclic citrullinated peptide; CT, computed tomography; ED, emergency department; FEV1, forced expiratory volume in one second; LLL, left lower lobe; CRP, C-reactive protein; WBC, white blood cells; ALC, absolute lymphocyte count; LDH, lactate dehydrogenase; EF, ejection fraction; RVSP, right ventricular systolic pressure IV, intravenously; SPM, spontaneous pneumomediastinum.

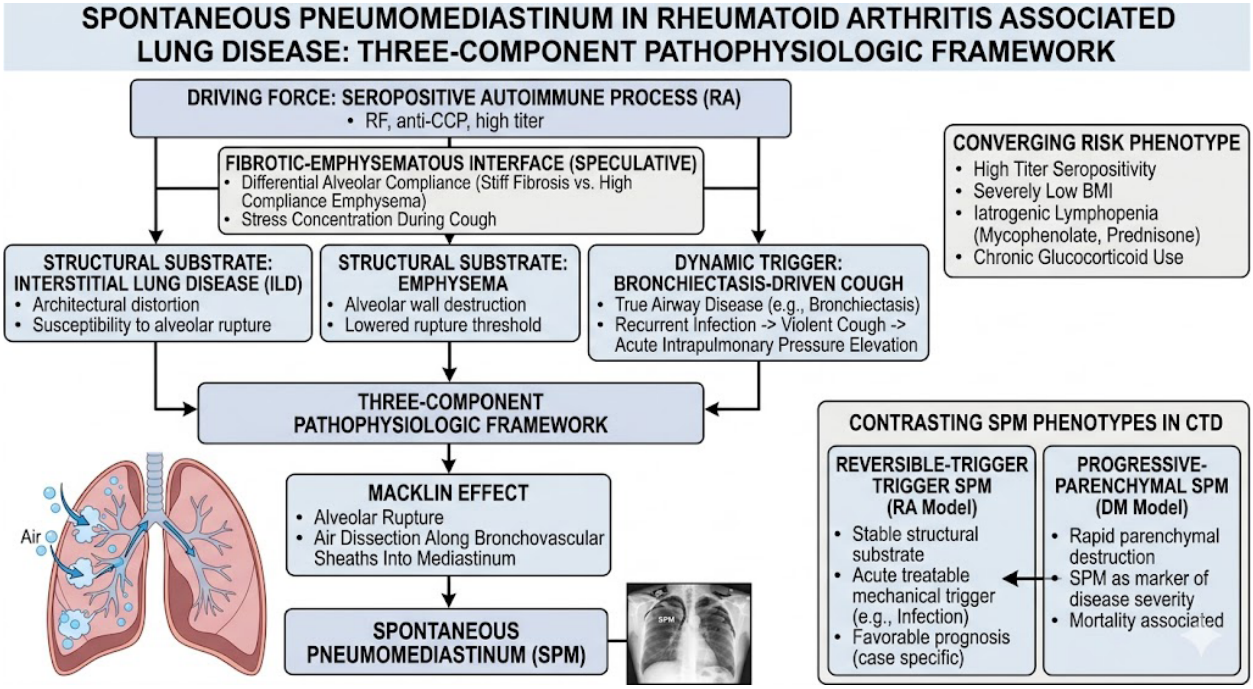


Figure 2. *A schematic diagram illustrating the three-component pathophysiologic framework (structural substrate from ILD + structural substrate from emphysema + dynamic trigger from bronchiectasis-driven cough → Macklin effect → spontaneous pneumomediastinum SPM).*

Abbreviations: ILD, interstitial lung disease; SPM, spontaneous pneumomediastinum; RF, rheumatoid factor; CCP, cyclic citrullinated peptide; BMI, body mass index; RA, rheumatoid arthritis; DM, dermatomyositis.

complete resolution at 8 weeks after antibiotics despite unchanged structural lung disease, and no recurrence at 12 weeks. However, SPM can resolve spontaneously, and causation cannot be established from a single observation. It is also possible that the ground-level fall contributed to the pneumomediastinum, although the absence of rib fractures, pneumothorax, and the concurrent infectious presentation make this less likely.

This case highlights a potentially important distinction between two SPM phenotypes in CTD-associated lung disease. In what we term “reversible-trigger SPM,” as observed in our patient, the structural substrate is chronic and relatively stable, and SPM appears to be precipitated by an acute, treatable mechanical event such as infection-driven cough. In contrast, “progressive-parenchymal SPM,” characteristic of dermatomyositis, likely reflects ongoing rapid parenchymal destruction where SPM serves as a marker of disease severity rather than a discrete mechanical event. If validated in larger cohorts, this distinction could have direct prognostic implications and would argue against applying dermatomyositis-derived mortality data to RA-SPM. However, this phenotypic classification is based on limited data and requires confirmation through multicentre case series or registry-based studies.

Okamoto et al. identified low BMI (cutoff 20.2 kg/m^2) and elevated lactate dehydrogenase (LDH, cutoff 378 U/l) as independent SPM predictors in CTD-ILD[9]. Our patient’s BMI (14.85 kg/m^2) was well below threshold, but LDH was normal (146 U/l), suggesting that the dual ILD/bronchiectasis pathology may lower the SPM threshold even without elevated LDH, or that thresholds derived from dermatomyositis-dominated cohorts may not be generalizable to RA. Lymphopenia (absolute lymphocyte count 0.55 ×10^3/μl) reflects the immunosuppressive burden of combined prednisone and mycophenolate. Even at 5 mg/day, prednisone carries significant infection risk, with a dose-dependent increase in hospitalized infection (1-year cumulative incidence 11.0% at ≤5 mg/day versus 8.6% with no glucocorticoids, p <0.001)[10]. Okamoto et al. found all 13 CTD-ILD patients who developed SPM did so after corticosteroid initiation[9]. The convergence of chronic glucocorticoids and immunosuppressant-induced lymphopenia underscores the need for steroid-sparing strategies and regular complete blood count monitoring[10].

Our patient’s course, hemodynamic stability, no pneumothorax, normal lactate, rapid oxygen weaning, complete 8-week resolution, no 12-week recurrence, appears consistent with the reversible-trigger phenotype and may suggest a favourable prognosis in this context. The only prior RA-SPM case demonstrated persistent SPM over two months with concomitant pneumothorax and no reported bronchiectasis, suggesting a single-mechanism model (ILD alone)[4]. The contrasting outcomes between these two cases, though limited by their small number, raise the possibility that the presence or absence of a reversible trigger may influence the clinical trajectory of SPM in RA-associated lung disease.

SPM in RA should prompt evaluation for both ILD and bronchiectasis. Current guidelines conditionally

recommend pulmonary function tests and high-resolution CT scan for people with RA who have ILD risk factors, including RA-related autoantibody elevation. Clinicians should consider low-threshold CT imaging in seropositive RA patients with known ILD and bronchiectasis who present with acute respiratory infections and chest or neck discomfort, even without classic Hamman sign. Future prospective studies and multicentre registries are needed to determine the true incidence of SPM in RA-associated lung disease, to validate the proposed phenotypic classification, and to establish whether the reversible-trigger phenotype consistently carries a more favourable prognosis than progressive-parenchymal SPM.

CONCLUSION

This case, only the second reported SPM in RA-associated lung disease, illustrates a hypothesized three-component pathophysiologic framework in which structural alveolar fragility from ILD and emphysema may provide the substrate, while infection-driven coughing from true airway bronchiectasis may provide the mechanical trigger. In RA, all three components appear linked to a shared seropositive autoimmune process. High titer seropositivity, severely low BMI, iatrogenic lymphopenia, and chronic glucocorticoid use may identify a convergent risk phenotype. Conservative management with targeted antibiotic therapy achieved complete resolution in this case, supporting the concept that SPM associated with a reversible trigger may carry a more favorable prognosis than progressive-parenchymal SPM described in dermatomyositis. Further investigation is warranted to validate these observations.

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