SUNTEXT REVIEWS

SunText Review of Medical & Clinical Research
ISSN: 2766-4813

Open Access
Case Report
Volume 6:4

Pulmonary Sweet's Syndrome Presenting as Organising Pneumonia with Cytopenias: A Rare Manifestation of a Neutrophilic Dermatosis

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Received date: 22 August 2025; **Accepted date:** 08 September 2025; **Published date:** 14 September 2025

Citation: Syed A, Rawther AK, Kumar P (2025) Pulmonary Sweet's Syndrome Presenting as Organising Pneumonia with Cytopenias: A Rare Manifestation of a Neutrophilic Dermatosis. SunText Rev Med Clin Res 6(4): 235.

DOI: https://doi.org/10.51737/2766-4813.2025.135

Abstract

Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, is an uncommon inflammatory condition characterised by the sudden onset of fever, painful cutaneous lesions, and neutrophilia. Although it usually affects the skin, extracutaneous manifestations can occur and often present significant diagnostic challenges. Pulmonary involvement is exceptionally rare and can mimic infection, malignancy, or autoimmune interstitial lung disease. We describe the case of a middle-aged woman with biopsy-proven cutaneous Sweet's syndrome who presented with progressive dyspnoea, pancytopenia, and systemic inflammation. Computed tomography of the chest revealed an organising pneumonia pattern. A comprehensive workup excluded infectious, autoimmune, and malignant causes. The patient responded rapidly to systemic corticosteroid therapy, with complete clinical and haematological recovery. This case emphasises the importance of recognising pulmonary Sweet's syndrome in patients with neutrophilic dermatoses and unexplained pulmonary infiltrates, and it reinforces corticosteroids as the mainstay of treatment.

Keywords: Pulmonary Sweet's Syndrome; Neutrophilic Dermatosis

Introduction

Sweet's syndrome was first described by Robert Douglas Sweet in 1964 as an acute febrile neutrophilic dermatosis characterised by fever, painful erythematous skin lesions, and neutrophilic infiltration of the dermis in the absence of vasculitis [1]. The reported incidence is approximately 2.7–3.0 cases per million population annually, with a predilection for middle-aged women [2]. Although Sweet's syndrome most commonly involves the skin, extracutaneous manifestations occur in up to 20% of cases, with involvement of the musculoskeletal, ocular, hepatic, renal, and pulmonary systems [2,3].

Pulmonary Sweet's syndrome is rare, with fewer than fifty cases reported worldwide [3-5]. The most frequent radiological pattern is that of organising pneumonia, although interstitial pneumonitis, diffuse alveolar haemorrhage, and nodular infiltrates have also been documented [5-7]. These manifestations can easily be misdiagnosed as infectious pneumonia, vasculitis, or cryptogenic organising pneumonia. Delay in recognition has been associated with progression to acute respiratory distress syndrome (ARDS) and, in some cases, death [6-8]. Prompt recognition and timely initiation of corticosteroid therapy are therefore critical.

Case Presentation

A middle-aged woman with a five-year history of biopsy-proven Sweet's syndrome presented with a two-week history of progressive exertional dyspnoea, malaise, and fatigue. She did not report fever, cough, sputum production, haemoptysis, or new cutaneous lesions.

Her past medical history included asthma–chronic obstructive pulmonary disease overlap, chronic back pain, migraine headaches, and depression. She was a lifelong non-smoker and resided in a remote community in Northern Australia. In the preceding 18 months, she had experienced multiple admissions for presumed pneumonia. On one occasion she required intensive care unit admission for septic shock due to left lower lobe

consolidation. On another admission she developed right lower lobe pneumonia complicated by haemoptysis and a type 2 non-ST elevation myocardial infarction; coronary angiography at that time demonstrated no obstructive coronary artery disease (Figure 1).

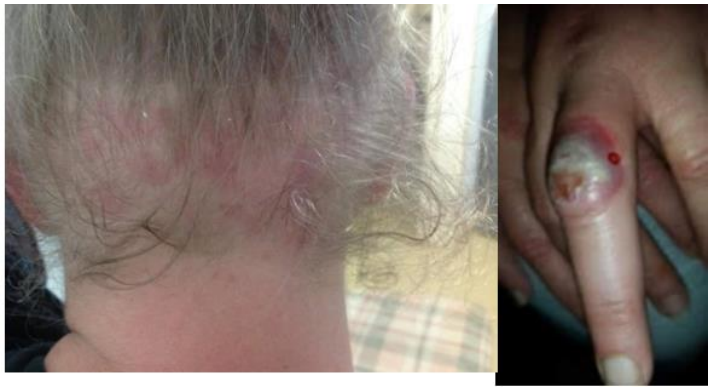

Figure 1: *Cutaneous lesions at time of initial Sweet's syndrome diagnosis (2018).*

On examination she was afebrile, tachypnoeic, and hypoxic on room air, with an oxygen saturation of 89%. Lung auscultation revealed bilateral basal crackles and an expiratory wheeze. No active Sweet's lesions were present on the skin. Cardiovascular and abdominal examinations were unremarkable (Figure 2).

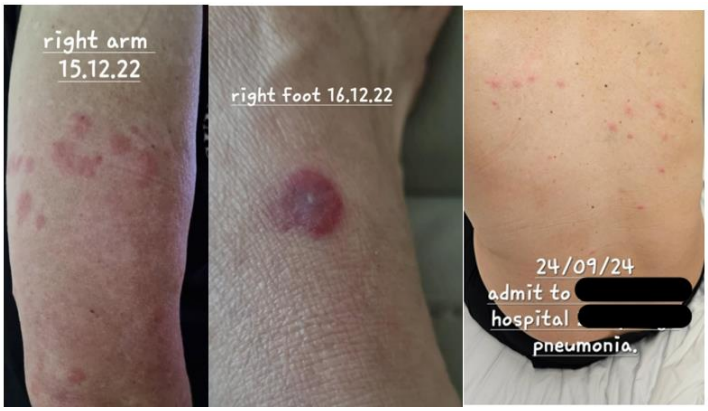

Figure 2: *Recurrent cutaneous lesions documented across subsequent presentations, affecting multiple body sites.*

Investigations

A computed tomography pulmonary angiogram demonstrated bilateral, predominantly subpleural and peribronchovascular consolidations, peribronchial nodularity, interstitial thickening, and small bilateral pleural effusions. No pulmonary embolism was detected. The findings were consistent with an organising pneumonia pattern [5,9].

Laboratory investigations showed normocytic anaemia (Hb 101 g/L, MCV 100 fL), neutropenia (ANC 1.26 × 10^9/L), and thrombocytopenia (95 × 10^9/L). C-reactive protein was elevated at 61 mg/L, and fibrinogen was raised at 4.6 g/L. Additional tests: haptoglobin 3.19 g/L, albumin 24 g/L, GGT 106 U/L, ACE 47 U/L, ferritin and vitamin B12 within normal limits. Autoimmune screening (ANA, dsDNA, ENA, RF, anti-CCP, ANCA) was negative. Peripheral smear revealed no blasts, dysplasia, or leucoerythroblastic changes.

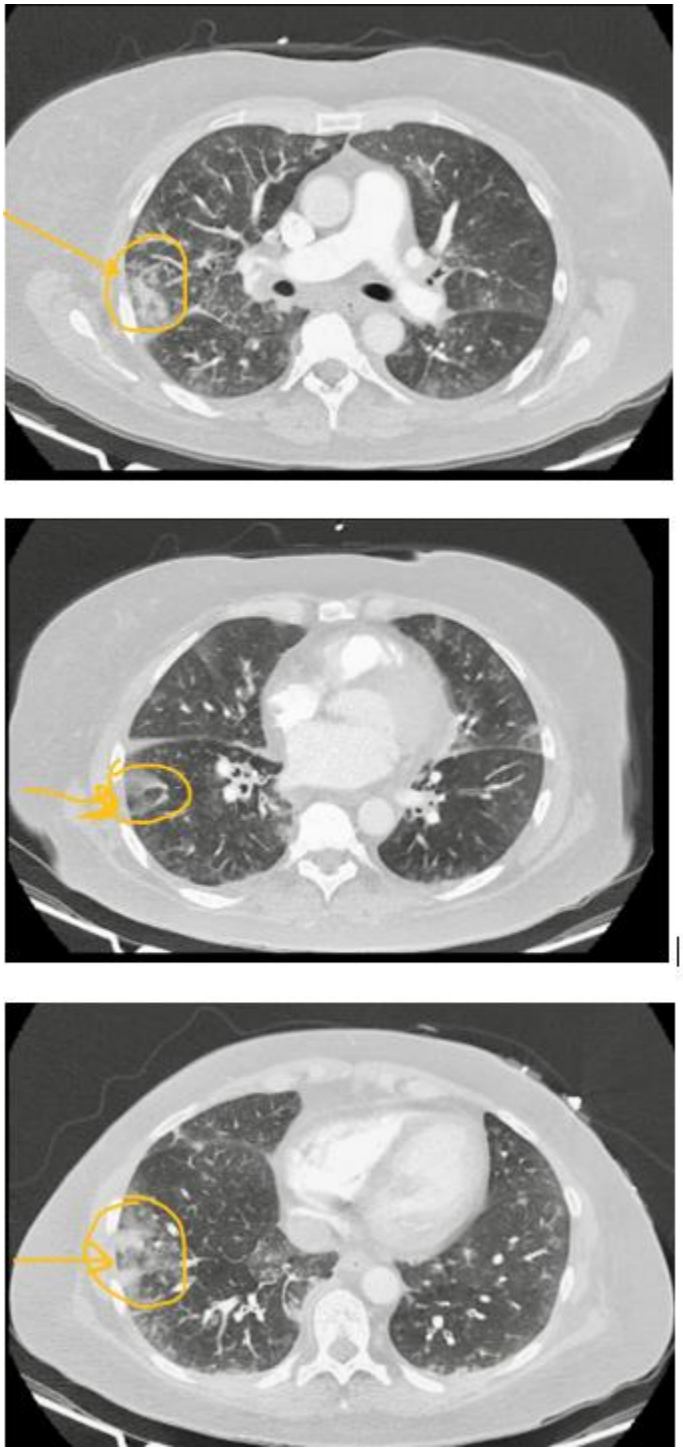

Figure 3: *Representative HRCT images. Arrows indicate peribronchial consolidations and masses. The perilobular "arcade-like" pattern is most clearly visible in (right lung).*

During a previous admission, bronchoscopy with bronchoalveolar lavage and transbronchial biopsies had been performed. Cultures for bacterial, fungal, and mycobacterial organisms were negative. Cytology excluded malignancy. Histopathology demonstrated non-specific chronic inflammation without granulomas or vasculitis.

Diagnostic Considerations

The differential diagnosis of bilateral pulmonary infiltrates in this patient included:

- Infectious pneumonia or atypical infection.
- Cryptogenic or post-infectious organising pneumonia.
- Vasculitis or sarcoidosis.
- Connective tissue disease-associated interstitial lung disease.
- Haematological malignancy.
- Drug-induced pneumonitis.
- Pulmonary Sweet's syndrome.

Infection was excluded by negative cultures, absence of fever, and lack of response to prior antibiotics. Vasculitis and connective tissue disease were ruled out by negative serology and absence of granulomatous or vasculitic changes. Haematological malignancy was considered unlikely given the absence of blasts or dysplasia. There were no recent medication exposures to suggest drug-induced pneumonitis.

Given the patient's history of cutaneous Sweet's syndrome, the presence of cytopenias, the organising pneumonia pattern on imaging, the exclusion of other causes, and the prompt corticosteroid response, a diagnosis of pulmonary Sweet's syndrome presenting as organising pneumonia with cytopenias was made.

Management and Outcome

The patient was commenced on oral prednisolone at 0.75 mg/kg/day. Empirical intravenous piperacillin–tazobactam was initiated given her sepsis history but discontinued once infection was excluded. Probiotics and gastric protection were prescribed.

Within 72 hours, she demonstrated rapid clinical improvement: dyspnoea resolved, oxygenation normalised, inflammatory markers declined, and blood counts improved. A bone marrow biopsy was not performed as cytopenias resolved and no dysplastic features were present. She was discharged on a tapering corticosteroid regimen. At follow-up, she remained clinically stable with no relapse of pulmonary or cutaneous disease.

Discussion

Pulmonary involvement in Sweet's syndrome is rare, with <50 cases described [3-5]. The pathogenesis involves cytokine-mediated neutrophilic infiltration, particularly IL-1β, IL-6, TNF-α, and G-CSF [6]. These cytokines can also impair haematopoiesis, explaining the reversible cytopenias observed in systemic disease [10,11].

Radiologically, pulmonary Sweet's often resembles cryptogenic organising pneumonia (COP). High-resolution CT typically demonstrates subpleural or peribronchovascular consolidation, perilobular arcades, and ground-glass opacities [9,12]. The "reverse halo" or atoll sign is specific but present in only ~20% of cases [13]. Pleural effusions, as seen in our patient, are also reported [14].

Systemic manifestations of Sweet's syndrome include musculoskeletal symptoms (asymmetric arthralgia of shoulders, hands, wrists, ankles, knees) and ocular involvement (conjunctivitis, uveitis, inflammatory glaucoma) [8].

Multiple reports support corticosteroids as the mainstay of pulmonary Sweet's management. Ito et al [5] described biopsy-proven organising pneumonia resolving with steroids. Astudillo et al [14] documented a severe case improving only after steroids were commenced. Takimoto et al [7] first described pulmonary involvement, showing prednisone responsiveness. By contrast, Thurnheer and Aparicio reported fulminant, fatal pulmonary Sweet's with ARDS despite supportive care [15,8].

Relapse occurs in up to 40% of systemic cases. Steroid-sparing agents such as colchicine, dapsone, or cyclosporine are used in recurrent or refractory disease. Biologics targeting IL-1 and IL-6 are emerging options, though evidence is limited to case reports and small series [6,16].

This case underscores the diagnostic challenge posed by pulmonary Sweet's syndrome, especially when mimicking infection or ILD. Early corticosteroid initiation is diagnostic and therapeutic, avoiding unnecessary antimicrobial use or invasive testing, and may prevent life-threatening ARDS.

Conclusion

Pulmonary Sweet's syndrome is a rare but serious extracutaneous manifestation of neutrophilic dermatoses. It should be considered in patients with cutaneous Sweet's syndrome who present with unexplained pulmonary infiltrates and cytopenias. Organising pneumonia is the most common radiological pattern. Cytopenias likely reflect cytokine-mediated marrow suppression. Prompt corticosteroid therapy typically results in rapid resolution. Early recognition is essential to avoid misdiagnosis, prevent inappropriate treatment, and reduce the risk of progression to acute respiratory distress syndrome.

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