Case Report

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Antineutrophil cytoplasmic antibody-related vasculitis with a unique imaging presentation of organizing pneumonia: The key role of lung ultrasound

Fabiola Pugliese[1*], Francesco Tursi[2*], Valeria Frassani[3], Ottavia Nori[3], Chiara Oliveri[1], Alessandro Marinetti[5], Carlo Cosimo Quattrocchi[4,5], Romano Nardelli[3], Susanna Cozzio[1]

[1]Internal Medicine Unit of "Santa Maria del Carmine" Hospital, Azienda Provinciale per i Servizi Sanitari, Rovereto, Italy; [2]Cardiac and Pneumological Rehabilitation Medicine, Codogno Hospital, Lodi, Italy; [3]Pneumology Multizonal Unit of Arco Hospital, Azienda Provinciale per i Servizi Sanitari, Arco, Italy; [4]Centre for Medical Sciences - CISMed, University of Trento, Italy; [5]Radiology "Santa Maria del Carmine" Hospital, Azienda Provinciale per i Servizi Sanitari, Rovereto, Italy. *equally contributed to the manuscript.

ABSTRACT

Introduction: Neutrophil cytoplasmic antibody (ANCA)-related vasculitis (AAV) is characterized by necrotizing inflammation of small and medium-sized arteries. This heterogeneous group of vasculitides, including microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA) and eosinophilic granulomatosis with polyangiitis (EGPA), further classified according to two distinct types of ANCA pattern, cytoplasmic ANCA (c) and perinuclear ANCA (p), mainly directed against proteinase 3 (PR3) and myeloperoxidase (MPO), respectively. The most frequent pulmonary imaging finding is honeycombing, typical of the usual interstitial pneumonia (UIP) pattern, particularly in AAV MPO-ANCA positive patients. Another pattern, although rare, is represented by organizing pneumonia (OP). The efficacy and reliability of lung ultrasound (LUS) is very good in connective tissue diseases, especially interstitial pulmonary fibrosis (ILD). Subpleural infiltrates on ultrasound are visualized as round or oval hypoechoic consolidations, without visible central flow in color Doppler and power Doppler modes.

Case presentation: We report a case of a 25-year-old woman who admitted to our hospital with fever (38°C), dyspnea and pleuritic chest pain. The medical history was positive for Hashimoto's thyroiditis. Laboratory tests showed elevated inflammatory markers, no evidence of respiratory failure. LUS revealed bilateral and multiple pulmonary consolidations with a rounded and anechoic appearance. We performed computed tomography (CT) which showed multiple bilateral peripheral and non-segmental peri-bronchovascular consolidations with air bronchogram, which corresponds to the OP pattern. Over the next three days, LUS monitoring revealed a rapid expansion in the size and number of consolidations. Transthoracic biopsy revealed a histopathological picture attributable to vasculitis. ANCA antibody determinations were positive for anti-PR3 ANCA antibodies (213 AU/ml), MPO-ANCA antibodies were negative. The consolidations showed a clear improvement after the start of cortisone therapy 1 mg/kg i.v., subsequently followed by Rituximab (RTX) 1 g i.v.

Conclusion: There is emerging evidence to support that PR3-ANCA and MPO-ANCA antibodies have the potential to stratify patients into unique phenotypic subgroups. LUS allows also a multiple reassessments to monitor the response to therapy. In this case report in particular, LUS played a decisive role in the diagnostic steps, accelerating the achievement of the definitive diagnosis, thanks to the OP-like pattern which extended to involve the pleura.

Key words: LUS, anti-PR3 ANCA antibodies, AAV, organizing pneumonia

Correspondence: Fabiola Pugliese, MD, Corso Angelo Bettini, 1 - Rovereto (TN), Italy - E-mail: fabiola.pugliese@apss.tn.it; fabiola.pugliese@gmail.com

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Introduction

Antineutrophil cytoplasmic antibody (ANCA)-related vasculitis (AAV) is characterized by necrotizing inflammation of small- and medium-sized arteries. It is a heterogeneous group of vasculitides including microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA), and eosinophilic granulomatosis with polyangiitis (EGPA), further classified according to two distinct types of ANCA patterns [1-3]. The two main antibodies include cytoplasmic ANCA (c) and perinuclear ANCA (p) directed mainly against proteinase 3 (PR3) and myeloperoxidase (MPO), respectively [4,5]. Patients with GPA presentation are predominantly positive to PR3-ANCA, while those with MPA predominantly show MPO-ANCA. Emerging evidence suggests that PR3-ANCA and MPO-ANCA antibodies have greater specificity to clinical diagnosis for stratifying patients into unique phenotypic subgroups [6-12]. The pulmonary radiological presentation pattern of these diseases is the "honeycomb" pattern typical of interstitial pneumonia (UIP), particularly in AAV MPO-ANCA positive patients. Another pattern, although rare, is represented by organizing pneumonia (OP) [11-16].

The efficacy of lung ultrasound (LUS) has been well documented in many pulmonary diseases, such as pneumonia, atelectasis, pulmonary edema, and pneumothorax [15,16]. Reports concerning LUS applicability for the assessment of pulmonary changes secondary to connective tissue disease focus primarily on fibrosis in interstitial lung disease (ILD). Single publications indicate its applicability also in diagnostics of other, less common complications secondary to systemic connective tissue disease [17,22].

Subpleural infiltrates in ultrasound are visualized as hypoechoic round or oval consolidations, without central flow visible in color Doppler (CD) and power Doppler (PD) modes.

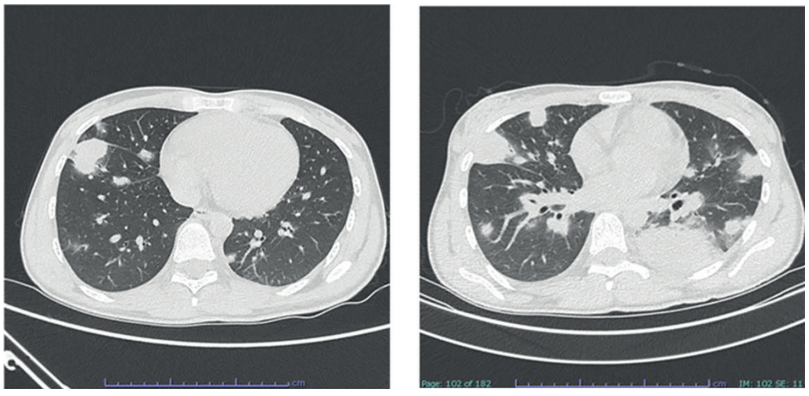

Figure 1. Chest CT: Showing subpleural round consolidations with features of cavitation. Follow-up CT after three days revealed rapid expansion in both size and number of consolidations.

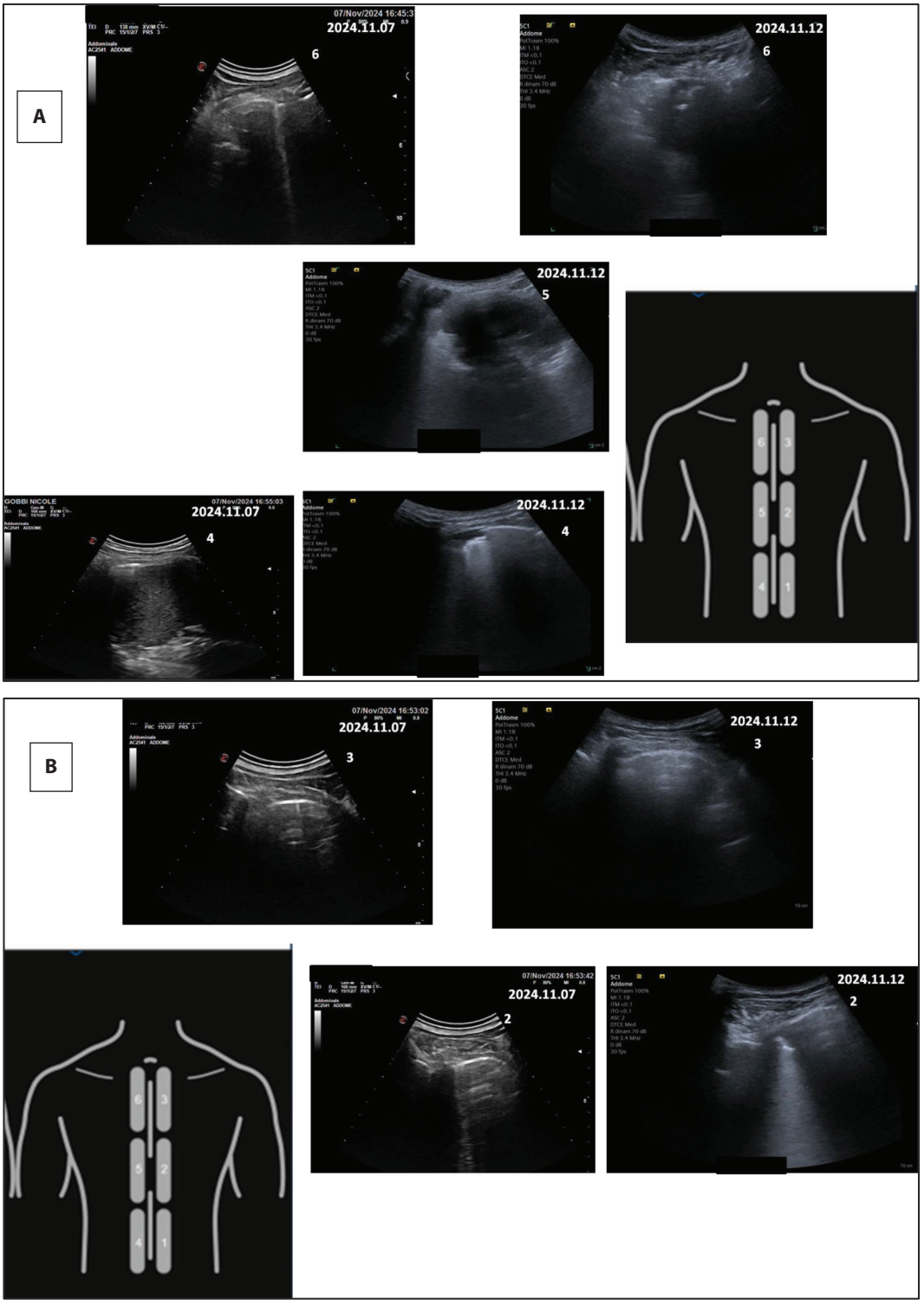

Figure 2. (A-D) LUS: Lung ultrasound images showing round hypoechoic consolidations (infiltrates) with hypoechoic pleura. Serial monitoring demonstrated progressive enlargement and increasing number of consolidations over three days.

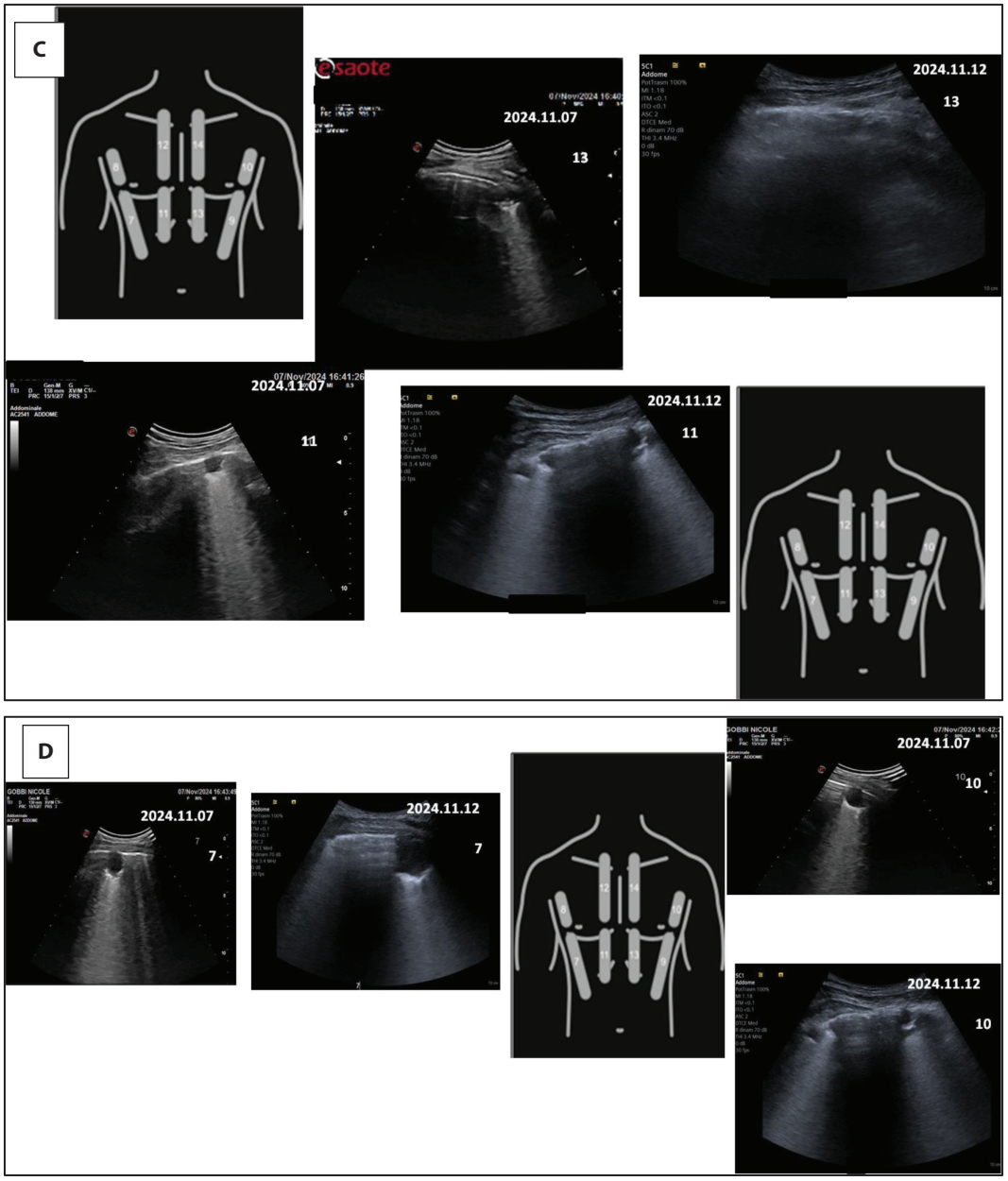

Figure 2. (*Continued*).

Case Presentation

We present the case of a 25-year-old woman who presented to the emergency department with fever (38°C), dyspnea, pleuritic chest pain and dry cough. The patient's medical history was positive for Hashimoto's thyroiditis. Chest auscultation revealed decreased breath sounds in the left mid basal lung fields. Laboratory tests showed elevated inflammatory markers but no evidence of respiratory failure. LUS revealed bilateral and multiple pulmonary consolidations with a rounded and anechoic appearance. On admission, a chest computed tomography (CT) scan was performed (Figure 1) which showed multiple bilateral peripheral and non-segmental peri-bronchovascular consolidations with air bronchogram in the middle lobe of the

left lung and upper lobe of the right lung, which was consistent with the OP pattern. The patient started antibiotic therapy with intravenous amoxicillin-clavulanate, however the fever persisted and the general conditions worsened. Furthermore, in the following three days, serial LUS monitoring showed a rapid increase in both the size and number of consolidations (Figure 2). Transthoracic biopsy yielded a histopathological picture attributable to vasculitis. ANCA antibody determinations were positive with a pattern for anti-PR3 ANCA antibodies (213 AU/ml), while MPO-ANCA antibodies were negative. The consolidations showed a clear improvement after the start of cortisone therapy 1 mg/kg i.v., subsequently followed by Rituximab (RTX) 1 g i.v.

Conclusion

AAV is a systemic disease that frequently involves the lungs, and pulmonary manifestations can sometimes be the initial clinical presentation. Although rare, organizing pneumonia (OP) should be considered in the differential diagnosis of AAV, and testing for serum ANCA is recommended. PR3-ANCA and MPO-ANCA antibodies may further help stratify patients into distinct phenotypic subgroups.

LUS represents a valuable, non-invasive tool for repeated assessments, allowing close monitoring of disease evolution and therapeutic response. In this case, LUS was pivotal in guiding the diagnostic process, detecting the OP pattern with pleural involvement, and expediting the definitive diagnosis due to its high sensitivity for pleural and subpleural abnormalities. Importantly, an extensive review of the literature did not identify previous reports specifically describing OP-like presentations in AAV evaluated by LUS. While LUS has been applied in AAV and OP has been reported in association with MPO-ANCA, publications integrating all three elements (OP + AAV + LUS) are essentially absent. Therefore, our case represents a novel contribution, providing unique insights into the sonographic appearance of atypical pulmonary involvement in AAV and highlighting the potential role of LUS in supporting early diagnosis and management of these patients.

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