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CLINICAL IMAGE

Unilateral diffuse alveolar hemorrhage in granulomatosis with polyangiitis

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Presentation

A 72-year-old woman was admitted to the emergency department with a one-week history of cough, dyspnea, and hemoptysis. Past medical history revealed a recent onset of ANA-positive oligoarthritis and that she started on methotrexate and low-dose glucocorticoids. On general examination, she was pale and auscultation revealed right-sided coarse crackles. Blood tests showed a low hemoglobin count (8.6 g/dL) and a raised C-reactive protein (129 mg/L). Urine analysis was unremarkable. Chest X-ray showed unilateral alveolar infiltrates (a), and high-resolution computed tomography revealed unilateral diffuse consolidation opacities and ground-glass pattern (b, c) (Fig. 1). Diffuse alveolar hemorrhage was suspected, so bronchoscopy and bronchoalveolar lavage were undertaken. Samples came out to be hemorrhagic and positive for hemosiderin macrophages, leading the diagnosis of diffuse alveolar hemorrhage. Further analysis to rule out any systemic vasculitis showed strongly positive anti-proteinase-3 antibodies

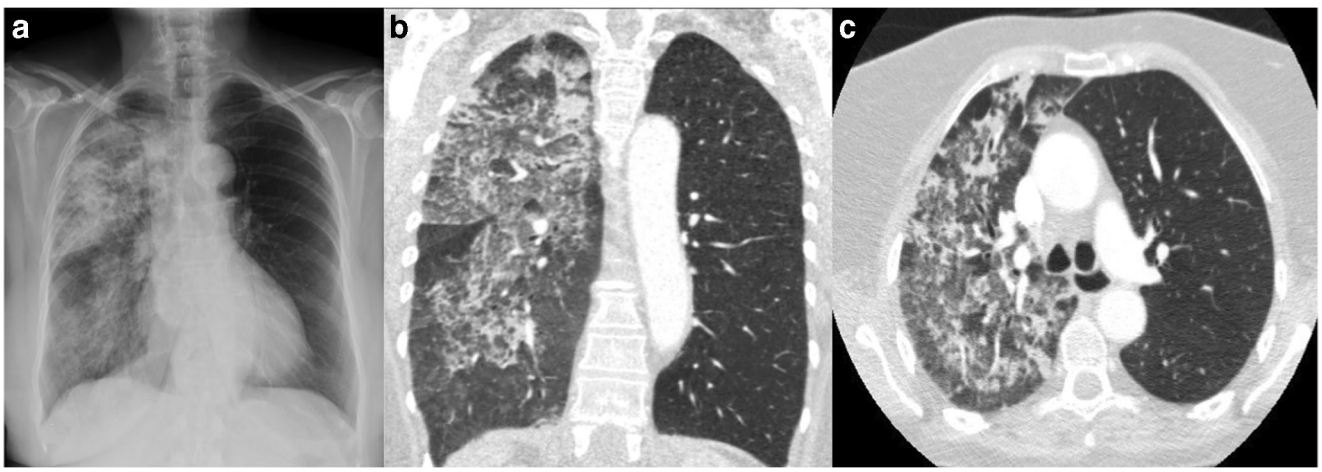

Fig. 1 Alveolar infiltrates on chest X-ray (**a**) and ground-glass opacities on high-resolution computed tomography (**b**, **c**) sparing the left hemithorax due to a complete unilateral right-sided diffuse alveolar hemorrhage in granulomatosis with polyangiitis

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(85.1 UI/mL, normal range < 2.0 IU/ml) so the diagnosis of granulomatosis with polyangiitis was confirmed.

Discussion

Diffuse alveolar hemorrhage is a rare and potentially fatal manifestation of several disorders, including ANCA-associated vasculitis. The clinical presentation of diffuse alveolar hemorrhage is nonspecific, ranging from acute respiratory failure to a more insidious course, and occurs as a consequence of pulmonary capillaritis. Hemoptysis is present in two-thirds of patients, and cough, dyspnea, and fever are other frequent symptoms. Hemoglobin drop is common, and more than 50% of patients require mechanical ventilation [1]. Although mortality varies between different series, with reported survival ranging from 50 to 82% at one year [2, 3], diffuse alveolar hemorrhage is the strongest predictor of early mortality in ANCA-associated vasculitis. Thus, prompt diagnosis and early treatment are always required. The initial imaging evaluation of patients suspected to have diffused alveolar hemorrhage should be a plain chest X-ray; although nonspecific, it is a high sensitive investigation and 94% of patients have radiographic evidence of diffuse alveolar hemorrhage on presentation [4]. The typical radiological pattern of acute diffuse alveolar hemorrhage is characterized by consolidation or ground-glass opacities due to alveolar filling, predominantly distributed in the perihilar regions and sparing of the apices and costophrenic angles. These findings are usually diffuse and bilateral, although they may rarely be unilateral, mainly in the context of pulmonary congestion due to heart failure. High-resolution computed tomography is superior in detecting ground-glass opacities and is always required in cases of suspected diffuse alveolar hemorrhage with normal chest X-ray findings. On high-resolution computed tomography, these abnormalities correspond to patchy ground-glass opacities without significant interlobular septal thickening. Within 48 h, septal thickening may occur and ground-glass opacities persist, resulting in a crazy-paving pattern. Pulmonary fibrosis may develop when hemorrhage recurs [5]. The high-resolution computed tomography findings, although not entirely specific, can guide further investigations such as bronchoscopy and biopsy. To our knowledge, this is the first description of a complete unilateral diffuse alveolar hemorrhage due to granulomatosis with polyangiitis.

Compliance with ethical standards

Disclosures None.

Ethical approval Informed consent for publication of the case and related images was obtained from the patient.

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