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Case Report

Migrant cavitation as primary involvement in a particular case of granulomatosis with polyangiitis [☆]

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ABSTRACT

Granulomatosis with polyangiitis (GPA), previously known as Wegener’s granulomatosis, is a necrotizing granulomatous vasculitis of the small and medium vessels involving the upper respiratory tract, lungs, and kidneys. In this case report, we will describe the case of a 60-year-old man who presented to our observation with recurrent episodes of hemoptoe, fever, and mucopurulent sputum. The diagnosis was made by radiological and laboratory tests.

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Background

Vasculitis is a heterogeneous group of disorders characterized by inflammation and necrosis of the blood vessel wall. The clinical and radiological manifestations are very variable and depend on the anatomical localization, the size of the vessels involved and the inflammation’s characteristics [1]. The Chapel Hill Consensus Conference classified them according to the caliber of the affected vessels, distinguishing in vasculitis of large vessels, vasculitis of medium vessels, and vasculitis of small vessels [2]. Pulmonary capillaries are the most commonly involved [3]. In particular, the GPA

belongs to the vasculitis group of small and medium vessels that classically affects the upper respiratory tract, the lung parenchyma, and the kidneys. The positivity of Neutrophil anticytoplasm antibodies (C-ANCA) is a diagnostic marker in this disease and is present in up to 90% of patients [4]. The pathogenesis is still unknown. GPA typically signs are hemoptoe, fever, and productive cough. Computer tomography (CT) is the most sensitive exam in detecting GPA pulmonary nodules, many of which are cavitated and have a random distribution [5]. The diagnostic criteria used for diagnosis are ACR/EULAR [6]. Therapy is based on the use of corticosteroids and immunosuppressants [7].

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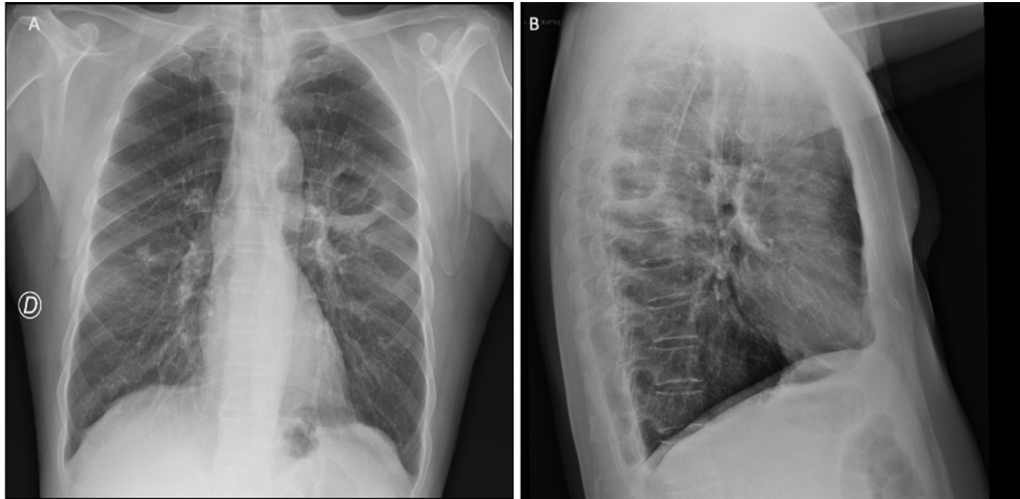


Fig. 1 – PA (A) and lateral (B) chest radiographs showed cavitation on the left and a focal nodular opacity on the right.

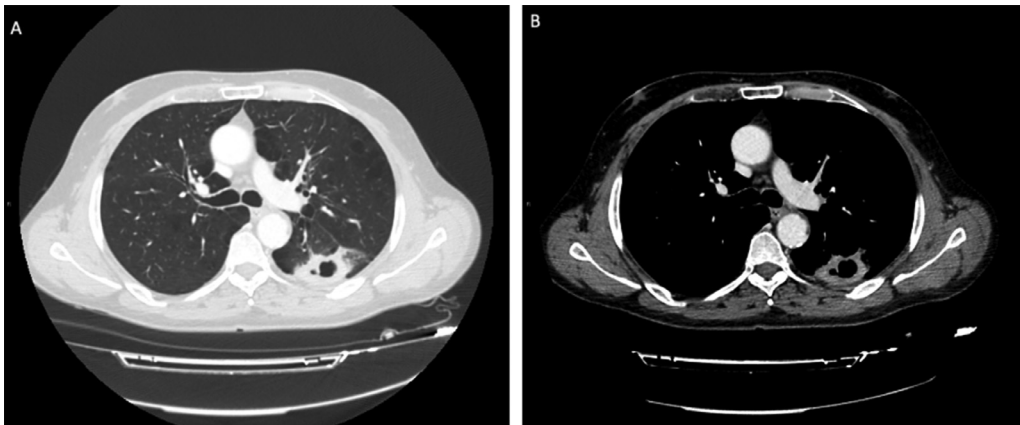


Fig. 2 – CT of the chest axial images: lung (A) and soft tissue (B) reconstruction algorithm showed the cavitated area in correspondence with the apical segment of the lower left lobe.

Case presentation

A 60-year-old Caucasian male presented to the hospital because of recurrent episodes of hemoptoe. The patient was a smoker, averaging about 40 cigarettes per day, asthmatic and cardiopathic, and had a family history of tuberculosis. Two months before the admission, after an episode of fever, productive cough with greenish sputum, hemoptoe, and dyspnea, he went to another hospital where he was diagnosed with tuberculosis after cavitation in the apical-dorsal region of the right lobe was detected by a chest CT scan. Given the persistence of the symptoms, he was admitted to the Respiratory Unit of “Mater-Domini” Hospital in Catanzaro, Italy. The patient was conscious, oriented in time and space during the visit, and pale. The body temperature was 37.5°C, the heart rate was 107 bpm, the peripheral oxygen saturation was 97% in ambient air, and the blood pressure was 120/80 mmHg. The thoracic examination revealed a cylindrical thorax with symmetric hemithorax, while by auscultation, a diffusely reduced

breath sound was observed throughout the entire pulmonary area. No lymph adenomegalies were palpable. An X-ray film of the chest (Fig. 1) showed cavitation with hydro-aerial level in the interclavicular-hilar region on the left and a small nodular formation in the right perihilar area.

Dynamic contrast-enhanced CT of the chest was performed. It showed a 2 × 1 cm consolidation area with cavitation in correspondence to the apical segment of the lower left lobe (Figs. 2 and 3) and the resolution of the cavitation found at the previous CT scan on the right lobe. Nodular formations have also been detected near the oblique fissure and in correspondence with the middle lobe.

The patient underwent video fiber bronchoscopy with microbiological and cytological examination on bronchial aspirate (BAS). The microbial investigation on BAS was negative for BK (PCR, bacterioscopist and cultural), common germs, and fungi. In contrast, the cytological examination showed a rich inflammatory granulocyte and macrophage component. The Quantiferon test results (0.16 IU/mL, positive \geq 0.35 IU/mL) in an immunocompetent patient allowed us to exclude the

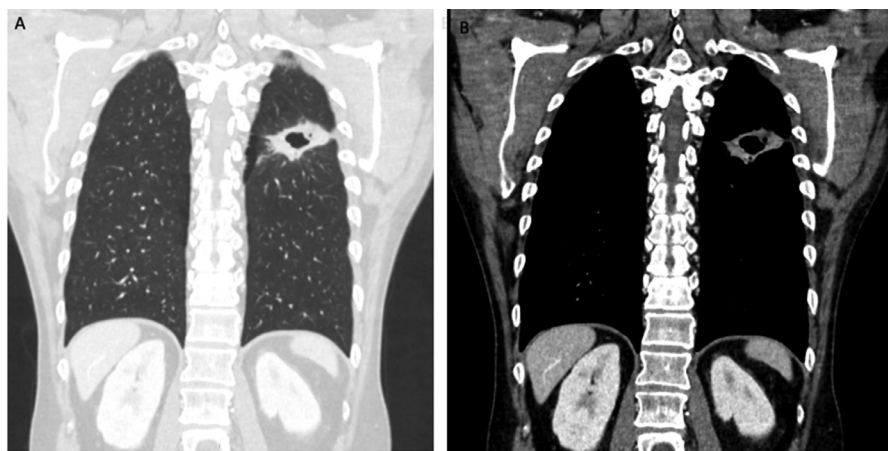


Fig. 3 – CT of the coronal chest images: lung (A) and soft tissue (B) reconstruction algorithm showed the cavitated area in correspondence of the apical segment of the lower left lobe.

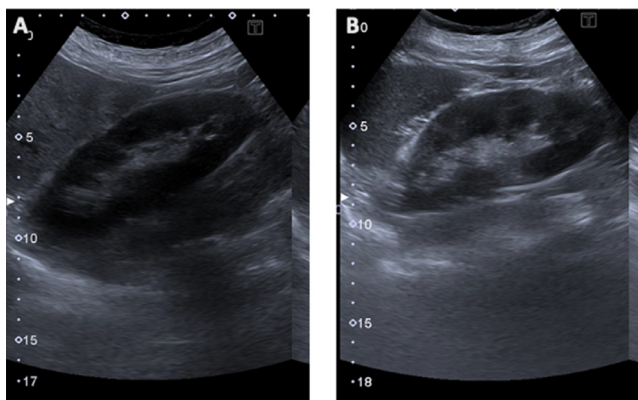


Fig. 4 – Normal sonogram of the right (A) and left (B) kidney.

suspicion of tuberculosis. The C-reactive protein value of 127 md/L (normal values 0-5 mg/L) suggested the presence of a severe inflammatory disease. C-ANCA antibodies were ex-

tremely high with a value of 126 U (standard <20 U). Further evaluation of the kidney by ultrasound revealed normal findings (Fig. 4).

We made diagnosis of GPA according to the latest ACR/EULAR criteria (at least 5 points) [6]: CANCA positivity (+5); pulmonary nodules and cavitation on chest imaging (+2). Treatment with corticosteroids and immunosuppressants was started. Symptoms improvement was observed. Before discharging the patient, the chest X-ray showed the resolution of the cavitation (Fig. 5).

Discussion

GPA, previously known as Wegener's granulomatosis, is a multisystemic necrotizing noncaseous granulomatous vasculitis that affects arteries, capillaries, and small to medium veins. It prefers the male sex, and the onset is typically around the age of 50. Antineutrophil anticytoplasm antibodies

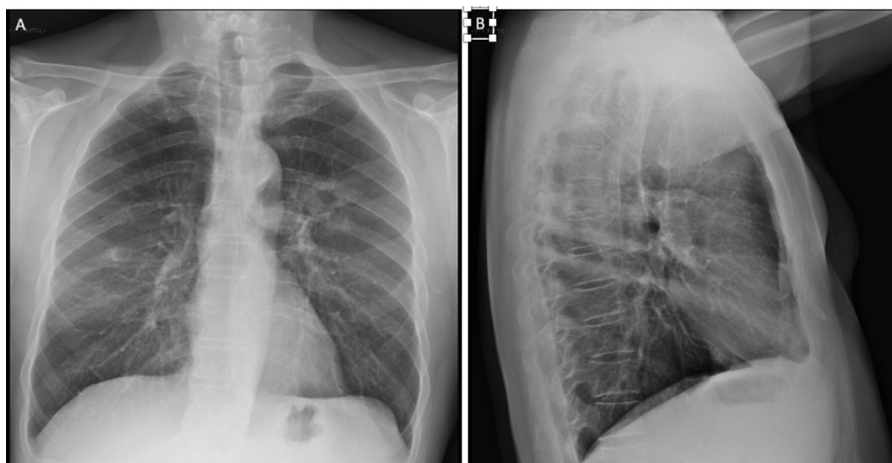


Fig. 5 – PA (A) and lateral (B) chest radiographs showed the resolution of the radiographic panel.

(ANCA) associated with vasculitis (AAVs) are related to granulomatosis with polyangiitis (GPA) [8], microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome), in which lung involvement is most frequently observed [9]. The patients usually present cough, hemoptysis, hemoptoe, chronic nasal obstruction/sinus symptoms, and proteinuria/hematuria. As several studies showed, 90% of cases show positive cancer (PR3), which correlates with disease activity. The classical triad of organ involvement includes the lung (involved in 95% of cases), the upper respiratory tract/sinuses (involved in 75%–90% of cases), and the kidneys (involved in 80% of cases). Based on systemic involvement, granulomatosis with polyangiitis can be further classified as: classical (comprising all 3 organs), limited (usually involving only the respiratory tract), and widespread, that also includes the skin (in 50% of cases), the eyes (in 45% of cases) and the peripheral nervous system (in 35% of cases) [10–13]. To the study of Lee et al. [14] the most common pattern in CT is the presence of multiple nodules, including some cavitates and migrants. According to Guggenberger et al., while pulmonary nodules are primarily asymptomatic, may evolve into pulmonary hemorrhages and, in some cases, cause a more severe, potentially fatal, lung-kidney syndrome [1]. Therapy is based on immunosuppressants (cyclophosphamide, methotrexate, and steroids). As demonstrated by Weiner et al., GPA is rapidly progressive, with a 10% 2-year survival rate without treatment. Appropriate medical therapy has dramatically increased long-term survival [11]. In our case, the clinical picture presented was not specific; for this reason, the diagnostic hypothesis was tuberculosis or bacterial infection. Primarily pulmonary involvement of GPA is, in fact, a rare occurrence, and this often results in a diagnostic delay. However, knowledge of the main clinical and radiological manifestations associated with laboratory tests allows us to formulate a diagnosis without proceeding with invasive investigations, which are not well accepted by the patient, to improve the patient's prognosis and quality of life.

Patient consent

Written informed consent was obtained from the patient.

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