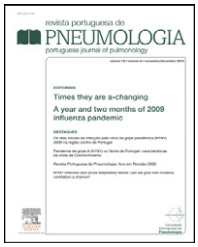
Rev Port Pneumol. 2013;19(1):45–48

revista portuguesa de PNEUMOLOGIA
portuguese journal of pulmonology
www.revportpneumol.org



CASE REPORT

Granulomatosis with Polyangiitis initially misdiagnosed as lung cancer☆

S. Campainha[a,*], M. Gonçalves[b], V. Tavares[b], P. Castelões[b], A. Marinho[c], S. Neves[a]

[a] *Serviço de Pneumologia, Centro Hospitalar de Vila Nova de Gaia/Espinho EPE, Vila Nova de Gaia, Portugal*
[b] *Unidade de Cuidados Intensivos Polivalente, Centro Hospitalar de Vila Nova de Gaia/Espinho EPE, Vila Nova de Gaia, Portugal*
[c] *Departamento de Doenças Auto-imunes do Serviço de Medicina Interna, Centro Hospitalar do Porto, Porto, Portugal*

Received 3 October 2011; accepted 12 March 2012

KEYWORDS
Systemic vasculitis;
Non-small-cell lung carcinoma;
Fine-needle biopsy

Abstract Granulomatosis with Polyangiitis (GPA), which was formerly named Wegener's Granulomatosis (WG) is a systemic disease characterized by necrotizing granulomatous inflammation and vasculitis that primarily involves upper and lower respiratory tract, as well as kidneys. Diagnosing GPA on the basis of transthoracic fine needle aspiration (TFNA) may be problematic, as it can be misdiagnosed as cancer. We describe a patient with a probable GPA which was originally diagnosed as malignancy, but who responded to lung cancer chemotherapy.

PALAVRAS-CHAVE
Vasculite sistémica;
Carcinoma não-pequenas células;
Biópsia por agulha fina

Granulomatose com poliangeíte inicialmente diagnosticada como cancro do pulmão

Resumo A granulomatose com poliangeíte (GPA), previamente denominada granulomatose de Wegener, é uma doença sistémica caracterizada por inflamação granulomatosa necrotizante e vasculite que envolve principalmente o trato respiratório superior e inferior, bem como os rins. O diagnóstico de GPA com base em biópsia por agulha fina (BAF) transtorácica pode ser problemático, levando ao diagnóstico incorreto de cancro. Os autores descrevem o caso de um paciente com provável GPA, inicialmente diagnosticado como cancro do pulmão, que curiosamente respondeu à quimioterapia para cancro do pulmão.

Introduction

Granulomatosis with Polyangiitis (GPA), more frequently called Wegener's Granulomatosis (WG) is a systemic disease characterized by necrotizing granulomatous inflammation and vasculitis that primarily involves upper and lower respiratory tract, as well as kidneys. It can sometimes present

☆ Please cite this article as: S. Campainha, et al., Granulomatose com Poliangeíte inicialmente diagnosticada como Cancro do Pulmão. Rev Port Pneumol. 2012. http://dx.doi.org/10.1016/j.rppneu.2012.04.002.
* Corresponding author.
E-mail address: scampainha@gmail.com (S. Campainha).

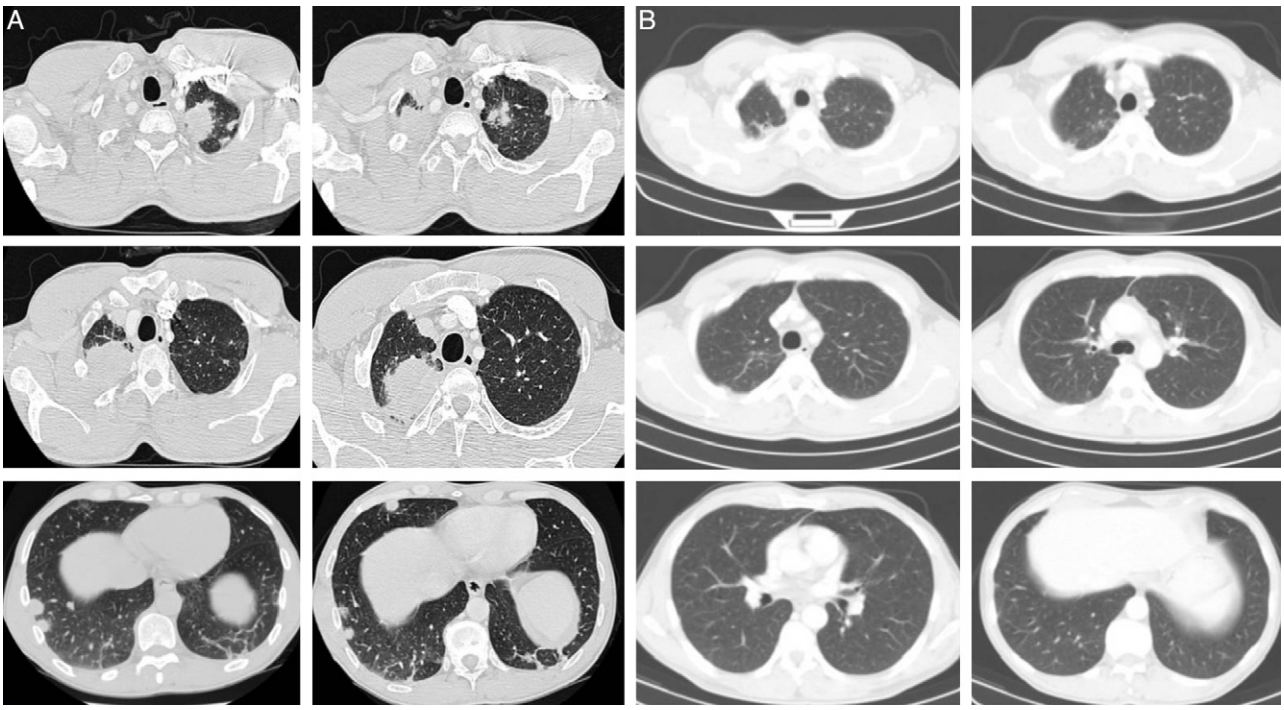


Figure 1 Chest CT at the time of diagnosis (A) and 3 months after chemotherapy (B). (A) Multiple nodular lesions in both lungs. The bigger lesions present in the apices. (B) Resolution of the nodular lesions after 3 cycles of chemotherapy. Some fibrotic sequelar changes can be seen in both apical regions.

as solitary or multifocal pulmonary lesions, without signs of extrapulmonary involvement.

Diagnosing GPA on the basis of transthoracic fine needle aspiration (TFNA) may be problematic, as it can be misdiagnosed as cancer. We describe a patient with GPA originally diagnosed as malignancy.

Case history

A 45-year-old man, 20 pack-year smoker, diagnosed with stage IV non-small cell lung cancer (NSCLC), by TFNA, underwent 6 cycles of chemotherapy with carboplatinum and gemcitabin, and complete remission was observed after 3 cycles (Fig. 1). At that time the cytological studies were reviewed, the diagnosis of NSCLC was maintained and antineutrophil cytoplasmic antibodies (ANCA) performed were negative. He was kept under surveillance by his Pulmonary Oncology team.

After an asymptomatic period of 15 months, with no signs of recurrence, he started having exertional dyspnea, dry cough, anorexia and weight loss (3 kg over a month). Physical examination was unremarkable. Chest X-ray showed multiple nodules and masses in both lungs some of which exhibited cavitation, as confirmed by Chest CT (Fig. 2A and B).

At this time a TFNA of a nodular lesion was repeated, which showed cellular debris, compatible with tumoral necrosis.

One week later, the patient developed oral ulcers and purpuric rash of lower limbs and external ear, as well as petechiae in the soft palate. Immunologic study revealed anti-neutrophilic cytoplasm antibody (cANCA) of 1/300 for proteinase 3 (PR3) > 200. Urinalysis revealed active sediment with proteinuria and eritrocituria. Rigid bronchoscopy showed ulcerated lesions along the tracheobronchial tree (Fig. 2C).

Bronchial biopsies proved extensive necrosis and vasculitis, and there were no features of a neoplastic nature. A skin biopsy was also performed, revealing leucocytoclastic vasculitis.

The patient was started on methylprednisolone 1 g IV od 3 days, followed by cyclofosfamide 100 mg PO od and prednisolone 1 mg/kg/d.

After a period of clinical and radiological improvement, respiratory failure developed requiring invasive mechanical ventilation and transfer to ICU unit. Repeated bronchoalveolar lavage revealed *Klebsiella pneumonia* and *Mycobacterium intracellulare* infection requiring therapy with imipenem and rifampin, clarithromicin and ethambutol, while keeping IV cyclophosphamide. Progressive disease was observed, with posterior development of nephrotic syndrome and renal failure. Implemented rescue therapy with plasmaferesis and rituximab, continuing with cyclophosphamide and IV immunoglobulins. Despite all the different approaches the patient did not survive.

Discussion

GPA is a vasculitis of unknown etiology first recognized as a separate entity by Friedrich Wegener in 1930, it is a granulomatous inflammation involving the respiratory tract, and necrotizing vasculitis affecting predominantly the small-sized vessels.