



Pulmonary fibrosis indicative of granulomatosis with polyangiitis initially believed to be sjögren's syndrome

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Abstract

Wegener's disease, currently called granulomatosis with polyangiitis, is a systemic necrotizing vasculitis of small vessels. It is typically associated with nodular opacity type pulmonary lesions of varying size developing towards excavation. Pulmonary fibrosis is more rarely observed. We report a case initially believed to be a Sjögren's syndrome in a 32-year old female Senegalese patient. The diagnosis of Wegener's granulomatosis was given on the basis of pulmonary fibrosis, oral ulcers, and the positivity of anti-PR3 type ANCA. The response to treatment remains favorable to this day, despite the occurrence of a secondary intercostal herpes zoster upon the use of cyclophosphamide and corticosteroids. The treatment of Sjögren's syndrome, for which the criteria were not met as it happens, may have bastardized the clinical expression of Wegener's disease.

Keywords: Sjögren's syndrome, wegener's disease, granulomatosis, pulmonary fibrosis

Introduction

Wegener's disease, currently called granulomatosis with polyangiitis, is a systemic necrotizing vasculitis of small vessels. Ortho-otolaryngology, lung and kidney manifestations dominate the clinical signs, although other organs may also be affected [1].

It is typically associated with nodular opacity type pulmonary lesions of varying size developing towards excavation. Pulmonary fibrosis is more rarely observed. We report a case revealed by pulmonary fibrosis, and initially believed to be a Sjögren's syndrome.

Case presentation

This case concerns a 32-year old Senegalese woman admitted for a chronic productive cough with whitish viscous sputum without fever, associated with NYHA stage I exertional dyspnea. This clinical presentation, which has been evolving since 2008, had required several hospitalizations and repeated unexpected antibiotic therapy. She was on prednisone 10 mg/day and hydroxychloroquine 400 mg/day for Sjögren's syndrome. Diagnosis of Sjögren's syndrome has been mentioned before admission in Internal Medicine, on the basis of dry eye and the results of salivary glands biopsy: grade II of Chisholm Masson. There was no other clinical argument, and rheumatoid factor was negative.

In her history, she did not report smoking or a profession at risk of lung disease or taking pneumotoxic medication.

The physical examination showed no abnormalities apart from diffuse wheezing in both lungs without orthopnea.

Physical examination on admission had shown no objective sign of Sjögren's syndrome. The blood count showed a white blood cell count of 7,220 cells/ml with a normal formula, a hemoglobin level of 13.5 g/dl, and platelets at 331,000 cells/ml.

We noted a nonspecific biological inflammatory syndrome with an accelerated rate of sedimentation, and an increased rate of C-reactive protein. Antinuclear antibodies, extractable nuclear antigen antibodies, Cyclic citrullinated peptide antibodies and anti-native-DNA auto-antibodies were negative.

Creatinine was normal, as was 24h proteinuria, transaminases and muscle enzymes. The tuberculin skin test was negative.

There was no active urinary sediment at the admission of the patient. The chest radiograph showed reticulonodular infiltrates on both sides (Figure 1). Chest computed tomography objectified a diffuse infiltrative pneumopathy with honeycomb images in favor of a pulmonary fibrosis.

Pulmonary function tests showed a mixed restrictive and obstructive type ventilatory deficit not improved by inhaled beta2. Biopsy samples taken at bronchoscopy showed signs of non-specific inflammation.

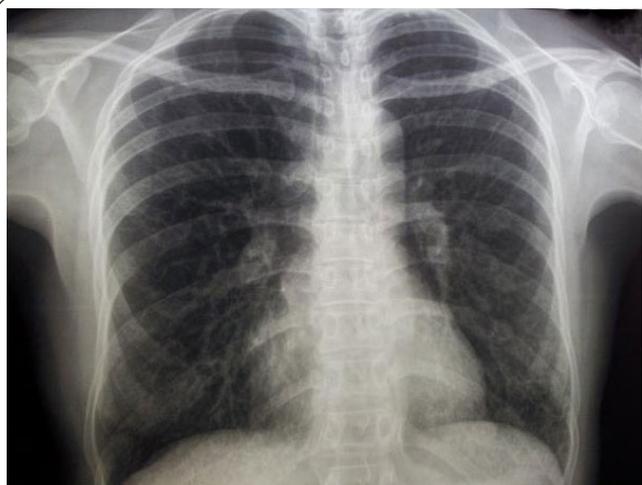


Figure 1. Chest X ray showed reticulonodular infiltrates on both sides.

Additional immunological tests performed upon the onset of lingual ulcers and frontal headaches highlighted positivity of anti-PR3 type ANCA. Brain MRI and CT scans of the facial bones were normal.

The diagnosis of Wegener's granulomatosis was given on the basis of pulmonary fibrosis, oral ulcers, and the positivity of anti-PR3 type ANCA. Biopsy tongue has not been realized. The response to treatment remains favorable to this day, despite the occurrence of a secondary intercostal herpes zoster upon the use of intravenous cyclophosphamide (1 g every month) and corticosteroids (1 mg/kg/day for 4 weeks then dose reduction progressively).

Discussion

The diagnosis of granulomatosis with polyangiitis was given in our patient because of the presence of 2 out of 4 ACR 1990 criteria [2]; and that of pulmonary fibrosis based on CT images [3]. Pulmonary manifestations are variable in Wegener's granulomatosis, and are generally preceded by those of the upper airway.

They are present in 50 to 75% of cases [4,5]. The most characteristic lesion is represented by the nodules observed in half of the cases [6-8]. The other conditions are pulmonary infiltrates in 30-50% of cases, alveolar condensation in 30-50% of cases, and pleural effusion in 9-28% of cases [9]. Pulmonary fibrosis rarely reveals granulomatosis with polyangiitis. Ketata et al., reported one case in 2009 [10]. Most cases of pulmonary fibrosis described are related to p-ANCA vasculitis, unlike granulomatosis with polyangiitis, which is a c-ANCA vasculitis [11,12]. The pathogenesis of this association is still poorly understood, but the direct role of ANCA in the pathogenesis of pulmonary fibrosis is increasingly emphasized [13,14].

The treatment of Sjögren's syndrome, for which the criteria were not met as it happens, may have bastardized the clinical expression of Wegener's disease.

Conclusion

This case once again confirms the multifaceted nature of lung disease in Wegener's granulomatosis with polyangiitis. Although rare, pulmonary fibrosis may reveal this vasculitis. Pathophysiological mechanisms are still poorly understood, but the direct role of ANCA is increasingly cited.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Authors' contributions	SN	AP	EA	ACN	TMD
Research concept and design	✓	✓	--	--	✓
Collection and/or assembly of data	✓	✓	✓	✓	✓
Data analysis and interpretation	--	--	--	--	--
Writing the article	✓	✓	✓	--	--
Critical revision of the article	✓	--	--	--	--
Final approval of article	✓	✓	✓	✓	✓
Statistical analysis	--	--	--	--	--

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