Severe Pulmonary Emphysema in a Young Patient With Vasculitis Associated with Proteinase-3 Anti-Neutrophil Cytoplasmic Antibodies (PR3-ANCA) 

Enfesma pulmonar severo en un paciente joven con una vasculitis asociada a anticuerpos anticitoplasma de neutrófilo tipo proteinasa-3 (ANCA-PR3)

To the Editor,

Pulmonary involvement is common in ANCA-associated vasculitis (AAV), but rarely manifests as pulmonary emphysema (13% of cases).  

We report the case of a 32-year-old man, smoker of 13 pack-years, with no exposure to other toxic substances, no family history, and no significant clinical history, who was diagnosed with anti-proteinase 3 (PR3) c-ANCA vasculitis and severe pulmonary emphysema. At the time of diagnosis, the patient had constitutional symptoms, arthralgia, digital ischemia, and kidney diseases in the form of non-nephrotic proteinuria, and microhematuria with normal glomerular filtration. Clinical laboratory tests revealed hemoglobin 12.2 g/dl and elevated ESR and C-reactive protein. The immunological study was positive for c-ANCA, with an anti-PR3 titer of 79 U/ml (normal value <2 U/ml) and anti-MPO 0 U/ml. Other studies, which included anti-glomerular basement membrane antibodies, ANA, complement, immunoglobulins, cryoglobulins, antiphospholipid antibodies, proteinogram, and hepatitis B, C, and HIV serologies, were normal or negative. Mantoux and Quantiferon were negative. Kidney biopsy showed pauci-immune extracapillary proliferative glomerulonephritis with crescent formation in 46% of the glomeruli (Fig. 1A). ENT computed tomography revealed no significant changes; chest CT showed 3 nodules <5 mm in the right lung, and severe bilateral diffuse mixed centrilobular emphysema with areas of paraseptal involvement and subpleural bullae, mainly in the upper lobes (Fig. 1B–F). No siderophages were found in sputum. Of note on lung function tests were: DLco: 68%; KCO: 66%; FEF 25%–75%: 58%; FEV1: 80%; and FEV1/FVC: 69%. He was treated with glucocorticoids at a starting dose of 1 mg/kg/day p.o. in a tapering schedule, and intravenous cyclophosphamide according to the CYCLOPS scheme. The patient stopped smoking and began treatment with bronchodilators. Alpha-1 antitrypsin levels were determined twice, and were normal on both occasions (140 and 145 mg/dl, respectively). Pi*S and Pi*Z alleles of the AAT gene were also determined qualitatively using PCR-ARMS and were negative.

Six months later, after completing induction therapy, the patient achieved clinical remission and began treatment with azathioprine. Respiratory problems included several infections that were managed with oral antibiotics. No significant changes were found on chest CT, and the 3 nodules previously visualized remained stable. Lung tests performed at that time showed DLco: 46%; KCO: 60%; FEF 25%–75%: 65%; FEV1: 78%; and FEV1/FVC: 76%.

References

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In short, pulmonary emphysema can coexist with ANCA-associated vasculitis, and the pathogenic contribution of this process, in addition to other clearly associated factors, such as tobacco and AATD, cannot be ruled out.

References


Acute Respiratory Failure Due to Chronic Tophaceous Gout With Laryngeal and Bronchial Involvement: An Unusual Complication

Insuficiencia respiratoria aguda secundaria a gota tofácea crónica con afectación laringea y bronquial: una complicación excepcional

To the Editor,

Sustained hyperuricemia (>7 mg/dl), when it manifests as chronic tophaceous gout (CTG), can lead to the formation of granulomas (tophi) around the urate crystals, which have a high capacity for erosion. Laryngeal involvement in CTG is rare, and can cause upper airway obstruction and acute respiratory failure (ARF), and can affect the tracheobronchial tree. We report the case of a patient with CTG who developed ARF after an acute episode of laryngeal gout requiring tracheostomy, and who also presented tophi in the left main bronchus (LMB). The patient ultimately developed squamous cell carcinoma of the left upper lobe (LUL) bronchus. To the best of our knowledge, this is the first description in the literature of a patient with both lesions.

This was a 51-year-old man, smoker of 60 pack-years, with a clinical history of chronic bronchitis, obesity, symptomatic hyperuricemia treated with allopurinol 300 mg/day (although compliance was irregular), arterial hypertension, and metabolic syndrome. One year previously, he had presented in the emergency department on repeated occasions with episodes of dyspnea, even at rest, attributed to COPD exacerbations, treated with bronchodilators and corticosteroids and discharged home with symptomatic treatment. Four months later, he returned to the emergency department with a severe attack of dyspnea. Examination showed increased work of breathing, central and peripheral cyanosis, stridor, and the following arterial blood gases: PaO₂ 55 mmHg, PaCO₂ 60 mmHg, pH 7.20 and HCO₃ 22 mmol. Chest X-ray revealed mild cardiomegaly and no other findings. An examination of the skin showed multiple giant tophi on the elbows, knees and hands, with deformed joints, on the eyelids, and on the abdominal wall, legs, and arms (Fig. 1A and B). He was assessed by the pulmonologist in the emergency department, and urgent evaluation by the

Fig. 1. (A and B) Gouty tophi in the elbow joint and abdominal wall. (C) Bronchoscopy: vocal cord paralysis in adduction with tophaceous deposit on the arytenoids. (D) Bronchoscopy: submucosal tophaceous gout deposits in the left main bronchus.

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