

Unusual presentation of a mass and “cobble-stone like” changes in the bronchus of a patient with granulomatosis with polyangiitis

Ping Fan¹, Zhiming Hao¹, Peilong Cao², Lan He^{1,*}

¹Department of Rheumatism and Immunology, The First Affiliated Hospital of Xi'an Jiaotong University, Xi'an 710061, Shaanxi Province, China

²Department of Pathology, The First Affiliated Hospital of Xi'an Jiaotong University, Xi'an 710061, Shaanxi Province, China

Received February 19, 2023 accepted March 06, 2023

A 44-year-old male was diagnosed with granulomatosis with polyangiitis (GPA) 1 year ago because of intermittent cough, short of breath, epistaxis, hearing loss and a positive proteinase-3 anti-neutrophilic cytoplasmic antibody (PR3-ANCA) test. After being treated with oral prednisone, cyclophosphamide for a year, he was hospitalized due to reappearance of cough and epistaxis. The nasopharyngoscopy showed extensive necrosis of the left nasal cavity. The biopsy of nasal cavity mucosa revealed fibrinoid necrotizing vasculitis (Figure 1A). His chest computed tomography (CT) showed multiple nodules and cavities (Figure 1B). Bronchoscopy showed diffuse edematous hyperemic mucosal changes and cobble-stone like lesions (Figure 1C, 1D). A narrowed tracheobronchus could be seen and a whitish mass which moved back and forth with breathing could be observed in the bronchi (Figure 1E). Bronchoscopic biopsy was performed. The pathology showed nonspecific inflammation (Figure 1F). Bronchial lavage fluid were negative for bacterial and fungal cultures both. After being treated with 500 mg methylprednisolone intravenously for 3 days, the patient's cough and epistaxis disappeared.

Bronchoscopic examination is thought to be useful in detecting the abnormal bronchial findings of GPA.^[1] The most common airway abnormalities in GPA are mucosal edema, erythema, thickening, and granularity of mucosal surface could be seen. Persistent mucosal inflammation followed by fibrosis is responsible for bronchial stenosis. GPA can also cause tracheal or bronchial mass lesions, called “inflammatory

pseudotumor”. In general, “cobble-stone like” mucosa or inflammatory pseudotumor in airway indicate active disease.^[2] The quick treatment response of this patient suggested that the changes of this patient was acute inflammation and could be reversed.

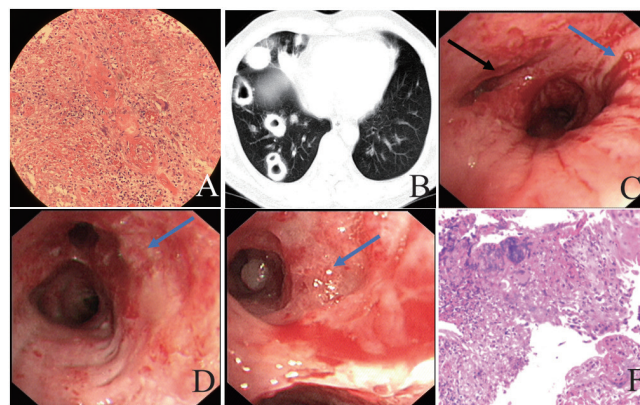


Figure 1: (A) Pathological changes of the nasal cavity mucosa biopsy: inflammatory exudates, granulomas formation with necrosis, and fibrinoid necrosis of a small vascular walls. (B) Chest CT: multiple nodules and cavities. (C, D and E) Bronchoscopic observations: the mucosa of bronchus was edema, hyperemia (blue arrow in Figure C), narrowing of the middle lobe of the right lung (black arrow in Figure C), “cobble-stone” changes of the mucosa (blue arrow in Figure D), whitish mass in the lumen of upper lobe of the left lung (blue arrow in Figure E). (F) HE staining with transbronchoscopic biopsy of the “cobble-stone like” changes of the bronchial mucosa: chronic mucosal inflammation with necrosis, granulation formation with squamous metaplasia (X 200). CT, computed tomography; HE, hematoxylin-eosin.

Address for correspondence:

*Lan He, Department of Rheumatism and Immunology, The First Affiliated Hospital of Xi'an Jiaotong University, Xi'an 710061, Shaanxi Province, China.
E-mail: xajdh187@mail.xjtu.edu.cn

Funding

This article received no external funding.

Author Contributions

Not applicable.

Informed Consent

The patient has given the consent for his images and other clinical information to be reported in the journal.

Ethical Statement

Not applicable.

Conflict of Interest

All authors declare no conflict of interest.

Reference

- [1] Nishiuma T, Ohnishi H, Yoshimura S, *et al.* A case of granulomatosis with polyangiitis (Wegener's granulomatosis) presenting with marked inflamed tracheobronchial mucosa. *Case Rep Med.* 2013;2013: 208194.
- [2] Polychronopoulos VS, Prakash UB, Golbin JM, *et al.* Airway involvement in Wegener's granulomatosis. *Rheum Dis Clin North Am.* 2007;33:755-775.