

# Multiple vasculitic pseudotumours masquerading as an obstructing endobronchial malignancy

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## DESCRIPTION

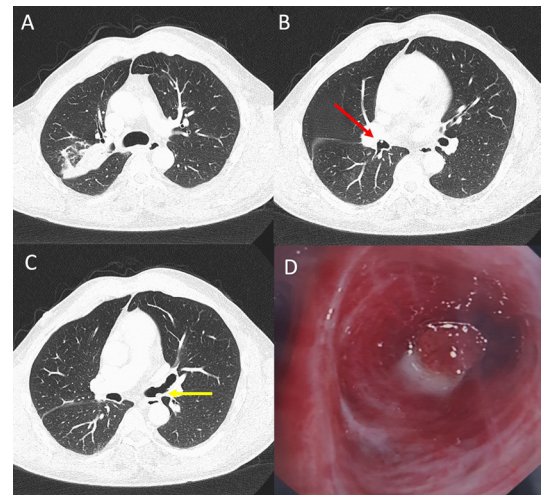
A man in his 50s presented with non-productive cough, which was unresponsive to antibiotics. He was an ex-smoker with a history of p-ANCA vasculitis diagnosed 3 years prior. His vasculitis presented with extensive pulmonary nodules and paucimmune crescentic glomerulonephritis. At presentation, he received induction therapy including pulse steroids and intravenous cyclophosphamide. He was stable on maintenance 200 mg azathioprine.

ANCA titre rose from nadir of 10 to 160 with anti-PR3 IgG level of 125 RU/mL (nadir 18). CT of the thorax revealed consolidation in the posterior right upper lobe with no visible air bronchograms raising suspicion of an endobronchial tumour causing post-obstructive consolidation (**figure 1A**). Endobronchial abnormalities were also noted in the bronchus intermedius and left main bronchus (**figure 1B,C**). Bronchoscopy showed numerous polypoid mass lesions located in the left main bronchus, right bronchus intermedius and a further lesion occluding the right upper lobe posterior segment bronchus (**figure 1D**).

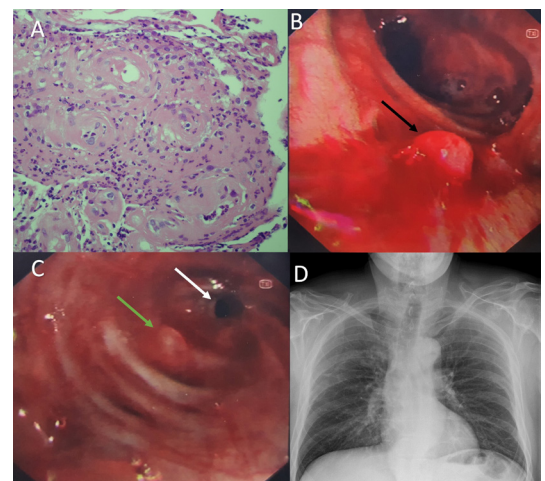
Differential diagnoses considered for multifocal endobronchial masses included: multifocal carcinoma, bronchial papillomatosis, bronchial tuberculosis, multifocal bronchial carcinoid and inflammatory vasculitic pseudotumours.

Endobronchial biopsies revealed markedly inflamed bronchial mucosa, ulceration and granulation tissue. Blood vessels showed extensive inflammatory infiltrate with prominent fibrinoid necrosis (**figure 2A**). The working diagnosis was endobronchial inflammatory pseudotumours due to a flare of vasculitis along with post-obstructive consolidation. He was given rituximab and continued azathioprine 200 mg until treatment response could be assessed. ANCA titres fell to 40 with anti-PR3 IgG falling to 17 RU/mL. Repeat bronchoscopy after 3 months showed marked improvement with complete resolution of the left main bronchus lesion and only minor abnormality persisting in the right bronchus intermedius (**figure 2B**). The right upper lobe posterior segment showed only a minor polypoid lesion remaining but did reveal a new stricture with partial occlusion of the airway (**figure 2C**). Follow-up chest X-ray showed resolution of the consolidation (**figure 2D**) and the patient reported complete resolution of his symptoms. He will continue rituximab and wean off azathioprine.

Pulmonary involvement occurs in 90% of patients with granulomatosis with polyangiitis (GPA) with the most common findings being parenchymal masses and nodules.<sup>1</sup> Airway involvement is found



**Figure 1** (A) CT of the thorax showing consolidation in posterior right upper lobe, no air bronchograms visible. (B) CT of the thorax showing endobronchial abnormality (red arrow) in the bronchus intermedius. (C) CT of the thorax with bronchial wall thickening (yellow arrow) in the left main bronchus. (D) Bronchoscopic image of right upper lobe posterior segment bronchus with a polypoid mass causing complete obstruction.



**Figure 2** (A) Endobronchial biopsy histology showing granulomatous inflammation. (B) Post-treatment bronchoscopic image noting residual bronchus intermedius lesion. (C) Post-treatment bronchoscopic image showing stricture causing partial occlusion (white arrow) and reduction in polypoid lesion size compared with that in **figure 1D**. (D) Post-treatment chest X-ray with resolution of consolidation.



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in 15%–55% of patients.<sup>2</sup> It usually occurs in conjunction with disease manifestations elsewhere, but rarely it may be the only manifestation of the disease.<sup>2</sup> Tracheobronchial involvement in GPA has a variety of manifestations but subglottic stenosis is the most frequent pathology with a reported frequency of 16%–23%.<sup>1</sup>

A comprehensive review on the literature of airway involvement in GPA made reference to polypoid lesions and mucosal projections into the airway lumen mimicking malignancy.<sup>2</sup> A case series on bronchoscopic findings in 77 patients with biopsy-proven GPA presenting with pulmonary involvement found an inflammatory ‘pseudotumour’ or ulceration in seven patients.<sup>3</sup> Despite an extensive review of the literature, there were very few reports of polypoid endobronchial lesions or inflammatory pseudotumours in patients with GPA. This case highlights a rare manifestation of GPA affecting the lower airways. The

accompanying imaging highlights the importance of early recognition to prevent permanent airway obstruction due to stricture formation and the response to immunosuppression.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

### Learning points

- ▶ ANCA vasculitis can cause multifocal endobronchial inflammatory pseudotumours and lead to airway obstruction.
- ▶ Prompt recognition and treatment with immunosuppression are key to reduce the risk of stricture formation and permanent airway obstruction.
- ▶ Bronchoscopy should be considered in patients with ANCA vasculitis with unexplained cough.

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