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 pauci-immune 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. Airway involvement is found in 75% of GPA patients with 30% presenting with respiratory symptoms. These findings are consistent with other case series in GPA and with endobronchial involvement in GPA and Wegener’s granulomatosis. Endobronchial involvement should be considered in all GPA patients with respiratory symptoms.


Accepted 22 January 2023

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Images in...

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

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. 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.3 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.

Acknowledgements The authors would like to acknowledge the contributions of Dr Omer Ahmed for his work on this case.

Contributors All authors have made substantial contributions to the work presented and were involved in the clinical management of the case. NL drafted the case report. CLW and NL completed a literature review and compiled the images. AO’B and LC edited the final draft of the report.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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.

References
1 Ananthakrishnan L, Sharma N, Kanne JP. Wegener’s granulomatosis in the chest: high-resolution CT findings. *Air Am J Roentgenol* 2009;192:676–82.

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