Official Case Reports Journal of the Asian Pacific Society of Respirology
Respirology Case Reports
OPEN ACCESS

Endobronchial vasculitis in childhood granulomatosis with polyangiitis

Matthew D. Wong & Leanne M. Gauld

Department of Respiratory and Sleep Medicine, Queensland Children's Hospital, Brisbane, QLD, Australia.

Keywords
Bronchial diseases, bronchoscopy, granulomatosis with polyangiitis, paediatrics, vasculitis.

Correspondence
Matthew D. Wong, Department of Respiratory and Sleep Medicine, Queensland Children's Hospital, PO Box 3474, Brisbane, QLD 4101, Australia.
E-mail: mdchwong@gmail.com

Received: 3 December 2020; Revised: 14 February 2021; Accepted: 15 February 2021; Associate Editor: Daniel Ng.

***Respirology Case Reports*, 9 (4), 2021, e00729**

doi: 10.1002/rcr2.729

Key message

Laryngo-tracheo-bronchial disease in childhood granulomatosis with polyangiitis may acutely present with endobronchial small vessel vasculitis without airway stenosis. Treatment should not be delayed in the presence of haemoptysis as it may indicate acute pulmonary capillaritis which can lead to fatal respiratory failure from diffuse alveolar haemorrhage.

Clinical Image

A 13-year-old male presented with two months of migratory polyarthritis and two weeks of constitutional symptoms, haemoptysis, and exertional dyspnoea. Initial physical examination revealed ulceration of the lip and nasal cavity, oropharyngeal and hard palate erythematous lesions, bilateral auscultatory fine inspiratory crackles, and a purpuric rash. Laboratory investigations included a haemoglobin nadir of 60 g/L, erythrocyte sedimentation rate of 95 mm/h, cytoplasmic anti-neutrophil cytoplasmic antibodies >2560, proteinase 3 anti-neutrophil cytoplasmic antibodies >1000, peak urine protein:creatinine ratio of 63 g/mol, and microscopic haematuria. Chest computed tomography revealed diffuse ground-glass and nodular disease. Flexible bronchoscopy identified several endobronchial lesions without evidence of airway stenosis (Fig. 1) and a subglottic biopsy revealed small vessel vasculitis. Bronchoalveolar lavage was positive for haemosiderin-laden macrophages. Leukocytoclastic vasculitis was identified from skin biopsies and fibrinoid necrosis seen on lip and nasal septum biopsies. His treatment included monthly intravenous methylprednisolone (five pulses) and cyclophosphamide (six infusions) with plans to transition to rituximab following a renal biopsy.

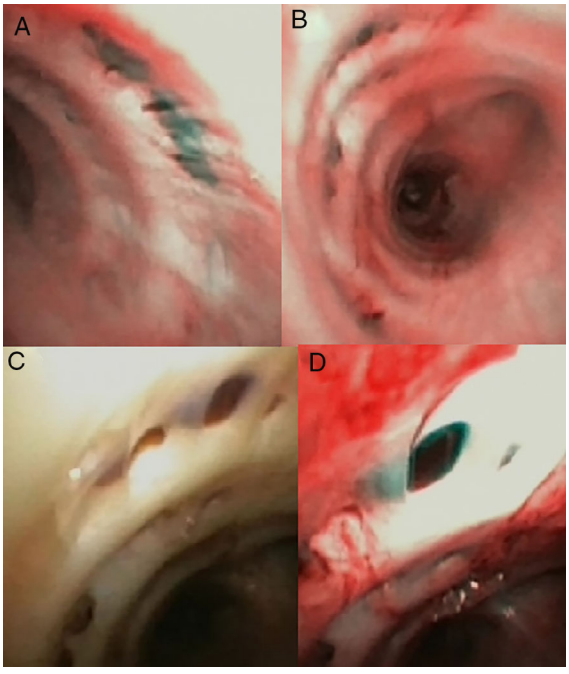

Figure 1. Left (A) and right (B–D) mainstem endobronchial ulcerations and petechiae in childhood granulomatosis with polyangiitis seen on flexible bronchoscopy. Submucosal lesions are seen with narrow-band imaging (A, B, D).

© 2021 The Authors. *Respirology Case Reports* published by John Wiley & Sons Australia, Ltd on behalf of The Asian Pacific Society of Respirology.

This is an open access article under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

Childhood granulomatosis with polyangiitis (GPA) often presents with lung parenchymal and airway disease [1,2]. Endobronchial ulceration and petechiae have rarely been described in childhood GPA and narrow-band imaging confirms the submucosal microvascular lesions that are distinct from pulmonary haemorrhage (Fig. 1). Airway disease in childhood GPA may involve endobronchial vasculitis before the development of laryngo-tracheo-bronchial stenosis.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

Author Contribution Statement

Matthew D. Wong drafted the original manuscript. Matthew D. Wong and Leanne M. Gauld contributed to the study conception, acquired the flexible bronchoscopy images, and approved the final manuscript.

References

1. Filocamo G, Torreggiani S, Agostoni C, et al. 2017. Lung involvement in childhood onset granulomatosis with polyangiitis. Pediatr. Rheumatol. Online J. 15(1):28.
2. Jariwala MP, and Laxer RM. 2018. Primary vasculitis in childhood: GPA and MPA in childhood. Front. Pediatr. 6:226.

© 2021 The Authors. *Respirology Case Reports* published by John Wiley & Sons Australia, Ltd on behalf of The Asian Pacific Society of Respirology.