Case Report

Central airway obstruction from lymphomatoid granulomatosis treated with an endobronchial stent

Daniel J. Greenberg *, Ariel Jaitovich, Nagendra Madisi

Albany Medical Center, Division of Pulmonary and Critical Care Medicine, Albany, NY, USA

A R T I C L E   I N F O

Keywords:
Central airway obstruction
Endobronchial stent
Endobronchial tumor
Lymphomatoid granulomatosis
Malignant airway obstruction

A B S T R A C T

Lymphomatoid granulomatosis (LG) is an extremely rare disease and is an unusual cause of central airway obstruction (CAO) with no standard of treatment in these conditions. LG is characterized by angioinvasion and angioinfiltration along with lymphohistiocytic cells. We present a 21-year-old female with LG who developed endobronchial lesions causing malignant CAO and acute hypoxic respiratory failure. She was treated with argon plasma coagulation, as well as a self-expandable metallic stent in the left main bronchus. Her stent was removed 4 months later after chemotherapy. Endobronchial stenting may be a useful bridge in patients who are undergoing more definitive treatment.

1. Introduction

Primary lung cancer is the main cause of non-foreign body-related malignant central airway obstructions (CAO). Indeed, about 30% of patients with lung cancer develop at some point in their disease course CAO-induced complications such as atelectasis, pneumonia, and dyspnea, and death from locoregional disease [1]. In these patients, bronchoscopy has a palliative management intent or relief from life-threatening obstruction. Lymphomatoid granulomatosis (LG), a rare Epstein Barr virus (EBV) associated B cell lymphoproliferative disorder, typically causes bilateral lower lobe lung nodules, and only rarely causes central airway obstruction [2]. Very few cases of LG endobronchial involvement have been reported in the English literature, two treated with chemotherapy [3,4] and one with pneumonectomy [5]. There is no data on bronchoscopic interventional options for these patients. Here, we present a patient with lymphomatoid granulomatosis complicated by CAO and acute hypoxic respiratory failure treated with an endobronchial stent, which was later removed after chemotherapy.

2. Case presentation

A 21-year-old woman without previous medical history developed lower extremities skin ulcerations followed by hemoptysis and dyspnea. She was hospitalized and later transferred to a tertiary center for further management. Chest computed tomography (CT) showed pneumomediastinum associated with a circumferential mediastinal mass surrounding the mid-lower trachea, proximal mainstem bronchi, along with scattered parenchymal nodules and masses (Fig. 1). She underwent skin biopsy that was consistent with LG induced by EBV. Hospital course was complicated by stridor and worsening acute hypoxic respiratory failure requiring urgent airway protection with endotracheal intubation. Emergency bronchoscopy showed severe tracheobronchitis with inspissated necrotic tissue in the mid to lower trachea and near-complete obstruction of the left mainstem bronchus and ~60% narrowing of the right

Abbreviations: CAO, central airway obstruction; LG, lymphomatoid granulomatosis; EBV, Epstein Barr virus; CT, computed tomography.

* Corresponding author. 43 New Scotland Ave, Albany, NY, 12208, USA.
E-mail addresses: greenb1@amc.edu (D.J. Greenberg), jaitova@amc.edu (A. Jaitovich), madisin@amc.edu (N. Madisi).

https://doi.org/10.1016/j.rmcr.2022.101770
Received 25 October 2022; Accepted 5 November 2022
Available online 8 November 2022
2213-0071/© 2022 The Authors. Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
main bronchus (Fig. 2). Multiple endobronchial ulcerated lesions were biopsied with histopathology results consistent with LG. Tumor debridement at the left main bronchus was done using argon plasma coagulation and a bipolar gold probe. A self-expandable metallic stent measuring 12 × 20 mm was deployed in the left main bronchus. The right main bronchus was then dilated with a controlled radial expansion balloon to more than 90% diameter. The patient was later extubated successfully, started on chemotherapy with R-EPOC regimen and discharged home. A new CT chest around 4 months later showed a decreased mediastinal mass size, improved patency in the trachea, and main bronchi with partial occlusion of the left main stent concerning for mucous impaction. A repeat bronchoscopy was performed to assess stent patency and possible stent removal. The airway mucosal wall was smooth without evidence of cancer tissue into the mucosa. The left main bronchus stent was in appropriate position and was found to be partially occluded with mucous that was cleared with suctioning. Given significant improvement in tumor burden with patency of both bronchi the left main stent was removed, and the airways remained patent (Fig. 3). The patient was eventually discharged home.

3. Discussion

LG is an EBV-associated B cell lymphoproliferative disorder, with a significant reactive T cell infiltration and angioinvasion involving the tracheobronchial tree, with pulmonary nodules and ulcers. Cutaneous involvement is seen in approximately 34% of cases, as erythematous papules and subcutaneous nodules with or without ulcerations. LG causes lung involvement in almost all cases, with
nodules distributed along the bronchovascular structures and/or interlobular septa being the predominant feature, usually sparing the lymph nodes and bone marrow [2,4,6].

Differential diagnosis of nodular opacities with lymphocytic infiltration includes granulomatous diseases such as sarcoidosis, tuberculosis, fungal infections, and vasculitides. The diagnosis is made histopathologically by dermal infiltration of lymphohistiocytic cells with angioinvasion and angioinfiltration, and no caseation [2].

Current treatment options include immunomodulation for low grade disease and immunochemothrapy for high grade disease. Only around one third of patients achieve remission with a high risk of relapse or refractoriness to treatment [2]. Median survival for pulmonary LG is only 14 months, with a mortality 65–90% [4]. Additionally, central airway obstruction from LG is not documented in the literature and therefore the benefits of bronchoscopic interventional procedures have never been reported.

Only 3–4% of patients with extra-nodal non-Hodgkin lymphoma develop tracheal-bronchial involvement [7]. Five-year survival rates of malignant tracheal tumors in non-surgically and surgically treated patients are 10% and 50%, respectively. A retrospective study focused on multiple primary tracheal tumors showed that 90% of cases were recanalized with the use of multiple bronchoscopy procedures leading to significant dyspnea improvement [8].

Bronchoscopic interventions for primary tracheal lymphomas are limited to case reports, and include stenting or laser photoresection followed by local ethanol injection [9,10]. One patient with tracheal mucosa-associated lymphoid tissue lymphoma received an endobronchial stent followed by chemotherapy, and had the stent removed 35 days after chemotheray [11]. There are 3 case reports to date describing endobronchial involveiment in LG [3–5], and no reported case of central airway obstruction leading to respiratory failure from underlying LG. Our patient's stent was removed 4 months after chemotherapy initiation and thus it appears that stent placement in conjunction with chemotherapy treatment, and eventural stent removal is a reasonable treatment strategy in patients with CAO from LG.

**Funding**

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

**Declaration of competing interest**

None.

**References**

[1] A. V Ernst, D. Feller-Kopman, H.D. Becker, A.C. Mehta, Central airway obstruction, Am. J. Respir. Crit. Care Med. 169 (12) (2004 Jun 15) 1278–1297, https://doi.org/10.1164/rccm.200210-11185O.

[2] C. Melani, E. Jaffe, W. Wilson, Pathobiology and treatment of lymphomatoid granulomatosis, a rare EBV-driven disorder, Blood 135 (16) (2020) 1344–1352, https://doi.org/10.1182/blood.2019009933.

[3] K. Kim, J. Park, J. Yoo, et al., Low grade pulmonary lymphomatoid granulomatosis with an endobronchial mass, Tuberc. Respir. Dis. 78 (2) (2015) 137, https://doi.org/10.4046/tub.2015.78.2.137.

[4] G.R. Mohyuddin, F. Sultan, G. Khaleeq, A rare presentation of a rare disease: pulmonary lymphomatoid granulomatosis, Case Rep. Pulmonol. 2012 (2012) 371490, https://doi.org/10.1155/2012/371490.

[5] W. Bartosik, A. Raza, S. Kalimuthu, A. Fabre, Pulmonary lymphomatoid granulomatosis mimicking lung cancer, Interact. Cardiovasc. Thorac. Surg. 14 (5) (2012 May) 662–664, https://doi.org/10.1093/icvts/ivr083.

[6] M.W. Beatty, J. Toro, L. Sorbara, J.B. Stern, S. Pittaluga, M. Raffeld, W.H. Wilson, E.S. Jaffe, Cutaneous lymphomatoid granulomatosis: correlation of clinical and biologic features, Am. J. Surg. Pathol. 25 (2001) 1111–1120, https://doi.org/10.1097/00000478-200109000-00001.

[7] M. Luick, E. Hansen, M. Greenberg, et al., Primary tracheal non-hodgkin’s lymphoma, J. Clin. Oncol. 29 (8) (2011) e193–e195, https://doi.org/10.1200/JCO.2010.32.0309.

[8] Z. Hao, Z. Yao, J. Zhao, et al., Clinical efficacy of treatment for primary tracheal tumors by flexible bronchoscopy: airway stenosis recanalization and quality of life, Exp. Ther. Med. (2020), https://doi.org/10.3892/etm.2020.8900.

[9] S. Huang, T. Ng, X. Xu, H. Chen, A case report of primary anaplastic large cell lymphoma arising from the trachea, Transl. Cancer Res. 8 (2) (2019) 699–704, https://doi.org/10.21037/tcr.2019.02.05.
[10] J. Tsurutani, A. Kinosita, H. Kaida, F. Narasaki, M. Fukuda, M. Oka, et al., Bronchoscopic therapy for mucosa-associated lymphoid tissue lymphoma of the trachea, Intern. Med. 38 (3) (1999) 276–278, https://doi.org/10.2169/internalmedicine.38.276.

[11] J. Ding, Z. Chen, M. Shi, Tracheal stenting for primary tracheal mucosa-associated lymphoid tissue lymphoma, Eur. J. Med. Res. 18 (1) (2013), https://doi.org/10.1186/2047-783X-18-1.