Received: 1 June 2023 | Revised: 26 August 2023 | Accepted: 14 September 2023

DOI: 10.1002/ccr3.7962

CASE REPORT

Clinical Case Reports Open Access WILEY

Multiple airway plasmacytomas: A rare cause of proximal airway obstruction requiring tumor debulking

Christopher Ghiathi[1] | **Ashley N. Barlev**[2] | **Gabriel C. Caponetti**[3] | **John P. Plastaras**[4] | **Devraj Basu**[5] | **Adam D. Cohen**[6] | **Kevin C. Ma**[1]

[1]Division of Pulmonary Allergy and Critical Care Medicine, Department of Medicine, Perelman School of Medicine of the University of Pennsylvania, Philadelphia, Pennsylvania, USA

[2]Department of Pathology and Laboratory Medicine, Perelman School of Medicine of the University of Pennsylvania, Philadelphia, Pennsylvania, USA

[3]Division of Hematopathology, Department of Pathology and Laboratory Medicine, Perelman School of Medicine of the University of Pennsylvania, Philadelphia, Pennsylvania, USA

[4]Department of Radiation Oncology, Perelman School of Medicine of the University of Pennsylvania, Philadelphia, Pennsylvania, USA

[5]Department of Head and Neck Surgery, Perelman School of Medicine of the University of Pennsylvania, Philadelphia, Pennsylvania, USA

[6]Division of Hematology/Oncology, Department of Medicine, Perelman School of Medicine of the University of Pennsylvania, Philadelphia, Pennsylvania, USA

Correspondence
Christopher Ghiathi, Division of Pulmonary Allergy and Critical Care Medicine, Department of Medicine, Perelman School of Medicine of the University of Pennsylvania, Philadelphia, PA, USA.
Email: cghiathi@gmail.com

Key Clinical Message

Extramedullary plasmacytomas (EMP) can present as airway lesions causing central airway obstruction. Though typically solitary, EMPs should be considered in the evaluation of multifocal tracheobronchial tumors. Bronchoscopic tumor debulking and radiation therapy can be used for symptomatic relief.

Keywords: airway obstruction, hematology, interventional pulmonary, multiple myeloma, plasmacytoma, respiratory medicine

1 | INTRODUCTION

Extramedullary plasmacytomas (EMP) are plasma cell neoplasms that arise outside the bone marrow, frequently involving mouth and nasopharynx.[1] In the head and neck, solitary and dyad lesions have been reported to involve laryngeal or tracheobronchial structures resulting in central airway obstruction (Table 1).[2–7] However, multiple EMPs with extensive involvement of larynx and tracheobronchial tree has rarely been described. Herein we discuss a rare case of multiple EMPs resulting in upper airway and bronchial obstruction treated with endoscopic tumor debulking.

2 | CASE SUMMARY

A 40-year-old male with a history of obstructive sleep apnea and obesity was transferred to our institution from an outside hospital for management of airway obstruction. He developed shortness of breath and globus sensation starting 1 year prior to presentation. Over the following months, he developed progressive shortness of breath, exercise intolerance, and hoarseness. On presentation, he was afebrile, hemodynamically stable, and without respiratory distress. Physical exam was significant for inspiratory stridor. Computed tomography (CT) scan of the chest demonstrated a 4 cm lobulated

mass on the right lateral wall of the trachea, a 1.2 cm paratracheal soft tissue density concerning for extraluminal tumor invasion, and multiple other airway lesions including the left hilum and right lower lobe orifice (Figure 1). No lung parenchymal infiltrates or consolidations were noted. Laboratory assessment revealed a normal complete blood count and differential, negative SARS-COV-2 and HIV testing, and normal quantitative immunoglobulins. Bronchoscopic examination demonstrated a mass on the anterior commissure, multiple partially obstructive pedunculated nodules at the proximal trachea involving the conus and thoracic inlet, and large extraluminal tumor with extrinsic compression (Figure 2). Obstructing lesions at the left hilum and right lower lobe orifice were also identified (Figure 2). No secretions were noted. Cultures of the bronchoalveolar lavage did not reveal any pathogenic organisms. Tumor debulking was performed using a combination

Authors	Findings
Jizzini et al.	Plasmacytoma near the larynx causing hoarseness and airway obstruction
Yacoub et al.	Plasmacytoma in the upper mediastinum causing airway compression
Uppal et al.	Plasmacytoma of the larynx requiring tracheostomy
Dammad et al.	Subglottic and upper tracheal plasmacytomas causing obstruction requiring rigid bronchoscopy
O'Neal et al.	Plasmacytoma of the cricoid cartilage causing airway obstruction
Stevic et al.	Tracheal plasmacytoma causing airway obstruction requiring surgical resection
Bujoreanu et al.	Chest wall and subglottic plasmacytomas causing stridor and airway compromise

TABLE 1 Aggregation of published cases showing plasmacytoma lesions causing upper or central airway compromise.

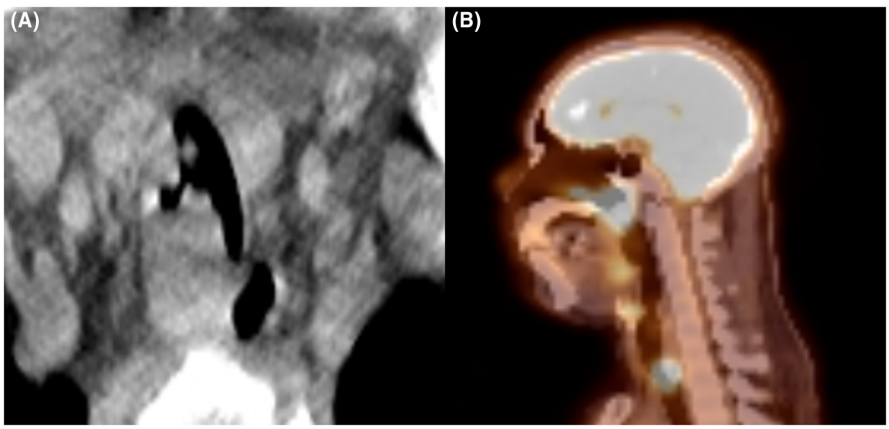

FIGURE 1 Axial computed tomography (CT) scan of 4 cm lobulated tracheal mass (A). Sagittal positron emission tomography (PET)/CT demonstrating multiple FDG-avid airway lesions (B).

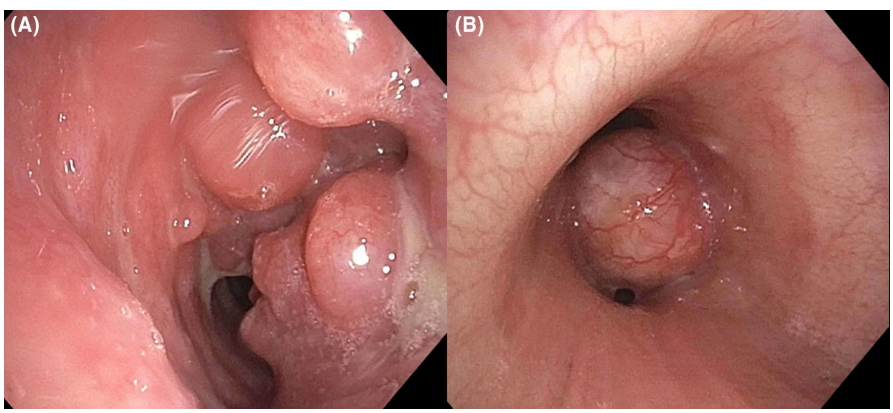

FIGURE 2 Bronchoscopic view of multiple lesions in the proximal trachea (A). Obstructing lesion in the left hilum (B).

of cautery snare (SnareMaster, Olympus America, State College, PA) and cryoablation (ERBE USA, Marietta, GA). Histopathologic evaluation of lung biopsies demonstrated sheets of kappa-restricted EBER (in situ hybridization) negative plasma cells compatible with the diagnosis of plasmacytoma (Figure 3).

A bone marrow biopsy showed no overt morphologic or immunophenotypic evidence of plasma cell neoplasm. Serum protein electrophoresis revealed a 1.3 g/dL peak in the gamma region, while immunofixation identified this peak as an IgG kappa band. Basic chemistries showed no evidence of renal dysfunction, anemia, or hypercalcemia to suggest multiple myeloma (MM). Positron emission tomography–CT (PET–CT) demonstrated multiple ^{18}F-deoxyglucose (FDG) avid lesions including the previously seen lesions in the right paratracheal space and right lower lobe as well as lesions within the right nasopharynx, anterior glottis, and left femur (Figure 1). No areas of abnormal uptake were identified in the abdomen or alimentary tract. Biopsies of anterior supraglottic and right posterior nasal cavity lesions also demonstrated involvement by plasmacytomas with histopathologic features identical to those in the lung biopsies. Radiation therapy to the tracheobronchial tree was considered but deferred in light of his multifocal disease and improved clinical status after tumor debulking. He was treated with daratumumab plus lenalidomide/bortezomib/dexamethasone (D-RVd) induction therapy.

3 | DISCUSSION

Plasma cell neoplasms are a group of hematolymphoid malignancies that include MM, plasma cell leukemia, solitary plasmacytoma of the bone, and solitary EMP. Solitary EMP lacks the hypercalcemia, renal dysfunction, anemia, bone lesions, or bone marrow involvement that characterizes MM. EMPs are typically diagnosed later in life and most frequently present in the upper aerodigestive tract.[8] Definitive radiotherapy can be curative for the majority of solitary EMPs. EMPs can also arise as a manifestation of MM.[9] While this patient did not have evidence of bone marrow involvement, he did have radiographic evidence of bone involvement that was clinically diagnosed as macrofocal myeloma.

EMPs in the upper airways can present with sinus pain, nasal drainage, and epistaxis if involving the nasal cavity, or hoarseness, stridor, and dyspnea if involving the larynx, trachea, or central bronchi. FDG-PET/CT is useful for identifying all sites for multifocal disease.[5] While EMPs as a cause of proximal airway obstruction have been described, multiple lesions involving extensive portions of the larynx and tracheobronchial tree is a rare presentation. Mechanical debulking/excision and targeted radiation therapy have been used for lesions causing obstruction, as they are exquisitely radiosensitive. Given the multifocal nature of the patient's disease, and bone involvement consistent with myeloma, induction chemotherapy with D-RVd was used after palliative endoscopic debulking.

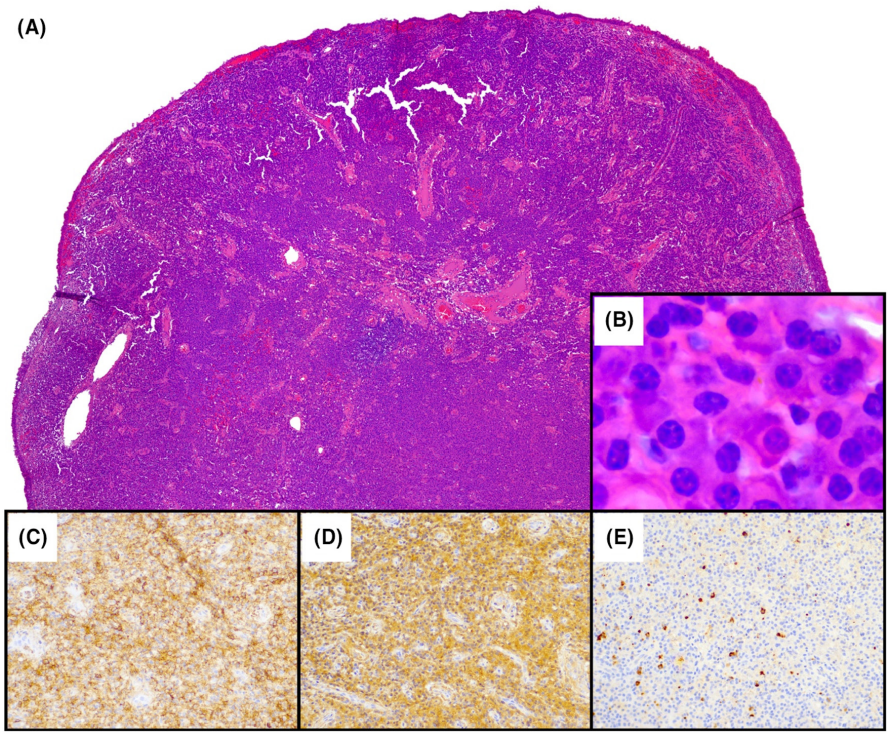

FIGURE 3 Composite microphotograph (10x) of the H&E-stained section of lung tracheal tumor shows respiratory type epithelium with an extensive diffuse infiltrate (A). A high magnification microphotograph (100×, oil) reveals that the diffuse infiltrate is almost exclusively composed of variably sized plasma cells (B). By immunohistochemistry, these plasma cells are positive for CD138 (C) and kappa (D), and almost completely negative for lambda (E) (20×).

4 | CONCLUSION

EMPs, both solitary and secondary to MM, can present with airway lesions resulting in central airway obstruction. A full work-up, including bone marrow biopsy is needed to help distinguish solitary EMPs from EMPs secondary to MM. Though typically solitary in nature, EMPs should be considered in the evaluation of multifocal tracheobronchial tumors. Bronchoscopic tumor debulking and radiation therapy can be used for symptomatic relief. FDG-PET/CT should be performed to evaluate disease burden.

AUTHOR CONTRIBUTIONS

Christopher Ghiathi: Visualization; writing – original draft; writing – review and editing. **Ashley N. Barlev:** Visualization; writing – original draft. **Gabriel C. Caponetti:** Supervision; visualization; writing – review and editing. **John P. Plastaras:** Writing – review and editing. **Devraj Basu:** Writing – review and editing. **Adam D. Cohen:** Writing – review and editing. **Kevin C. Ma:** Supervision; writing – review and editing.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

ORCID

Christopher Ghiathi https://orcid.org/0000-0001-7688-439X

REFERENCES

1. Gerry D, Lentsch EJ. Epidemiologic evidence of superior outcomes for extramedullary plasmacytoma of the head and neck. *Otolaryngol Head Neck Surg*. 2013;148(6):974-981. doi:10.1177/0194599813481334
2. Jizzini MN, Shah M, Yeung SJ. Extramedullary plasmacytoma involving the trachea: a case report and literature review. *J Emerg Med*. 2019;57(3):e65-e67. doi:10.1016/j.jemermed.2019.05.032
3. Yacoub GS, Dubaybo BA. Plasmacytoma and upper airway obstruction. *Am J Otolaryngol*. 1999;20(4):257-259. doi:10.1016/s0196-0709(99)90011-x
4. Uppal HS, Harrison P. Extramedullary plasmacytoma of the larynx presenting with upper airway obstruction in a patient with long-standing IgD myeloma. *J Laryngol Otol*. 2001;115(9):745-746. doi:10.1258/0022215011908829
5. Dammad T, Jalil BA. Extramedullary plasmacytoma presenting with acute airway compromise, treated with emergent rigid Bronchoscopic resection. *J Bronchology Interv Pulmonol*. 2016;23(2):e18-e20. doi:10.1097/LBR.0000000000000266
6. O'Neal ML, Entel RJ. Plasmacytoma of the cricoid cartilage with airway mass effect. *ORL J Otorhinolaryngol Relat Spec*. 2002;64(5):373-376. doi:10.1159/000066232
7. Stevic R, Ercegovac M, Stojšić J, Čolić N. Rare tracheal tumor: solitary plasmacytoma. *J Postgrad Med*. 2018;64(2):115-118. doi:10.4103/jpgm.JPGM_739_16
8. Bujoreanu I, Hogan C, Virk JS. Multiple upper aerodigestive tract masses presenting with stridor-a rare case of aggressive multifocal extramedullary plasmacytoma. *Ear Nose Throat J*. 2022;101(3):186-188. doi:10.1177/0145561320949704
9. Wirk B. Plasma cell myeloma with multiple plasmacytomas. *Clin Adv Hematol Oncol*. 2011;9(8):633-635.

How to cite this article: Ghiathi C, Barlev AN, Caponetti GC, et al. Multiple airway plasmacytomas: A rare cause of proximal airway obstruction requiring tumor debulking. *Clin Case Rep*. 2023;11:e7962. doi:10.1002/ccr3.7962