The Journal of Emergency Medicine, Vol. 57, No. 3, pp. e65–e67, 2019

0736-4679/$ - see front matter

https://doi.org/10.1016/j.jemermed.2019.05.032

Clinical Communications: Adult

EXTRAMEDULLARY PLASMACYTOMA INVOLVING THE TRACHEA: A CASE REPORT AND LITERATURE REVIEW

Mazen Nizar Jizzini, MD,[1] Mohsin Shah, MD,[1] and Sai-Ching Jim Yeung, MD, PHD

Department of Emergency Medicine, The University of Texas MD Anderson Cancer Center, Houston, Texas
Reprint Address: Sai-Ching Jim Yeung, MD, PHD, Department of Emergency Medicine, The University of Texas MD Anderson Cancer Center, 1515 Holcombe Boulevard, Unit 1468, Houston, TX 77030

☐ **Abstract—Background: Extramedullary plasmacytoma is an uncommon type of plasma cell neoplasm that occurs outside of the bone marrow. Very rarely, extramedullary plasmacytomas can involve the trachea, causing significant respiratory distress. Case Report: We describe a patient with a history of multiple myeloma who presented with voice hoarseness and dyspnea and was found to have airway obstruction due to an extramedullary plasmacytoma near the larynx. Why Should an Emergency Physician Be Aware of This?: It is important to investigate the possibility of upper airway obstruction in cancer patients presenting with hoarseness and dyspnea to prevent incorrect management, which can lead to fatal results. In particular, wheezing and dyspnea in patients with a history of asthma may not always be due to asthma exacerbation. Computed tomography scans and emergency laryngoscopy have been shown to be useful in aiding with correct diagnosis of upper airway obstruction, ensuring appropriate treatment.** © **2019 Elsevier Inc. All rights reserved.**

☐ **Keywords—multiple myeloma; extramedullary plasmacytoma; tracheal tumors; cricoid cartilage**

INTRODUCTION

Plasma cell neoplasms comprise several chronic diseases, such as multiple myeloma (MM), solitary plasmacytoma, and extramedullary plasmacytoma (EMP) (1). EMP tumors, which occur outside the bone marrow, account for ~4% of all plasma cell neoplasms (2). Rarely, patients with MM have concomitant EMP. EMP can involve any organ system, but it is most commonly found in the head and neck region. The larynx is the most common site of EMP in this region; very rarely, the trachea and cricoid cartilage can be involved (1,3).

We describe a rare case of a patient with MM who presented to an emergency department (ED) with subacute hoarseness, wheezing, and dyspnea and was found to have EMP with near-to-complete tracheal obstruction.

CASE REPORT

A 63-year-old man with a history of asthma and MM, treated with the fourth cycle of daratumumab, pomalidomide, and dexamethasone regimen 2 weeks prior, presented to the ED with wheezing, moderate dyspnea, and hoarseness that had developed 2 days previously (4). The patient had no fever, cough, or other upper-respiratory symptoms Blood pressure was 153/92 mm Hg, heart rate was 72 beats/min, respiratory rate was 18 breaths/min, and pulse oximetry was 96% on room air. Physical examination revealed wheezing that was louder during a prolonged expiratory phase. The oral pharynx was normal, and there were no lymphadenopathy or palpable masses in the neck. Routine laboratory tests were unremarkable, and chest radiography (Figure 1)

[1] Contributed equally and are joint first authors.

Received: 14 February 2019; Final submission received: 28 March 2019;
Accepted: 6 May 2019

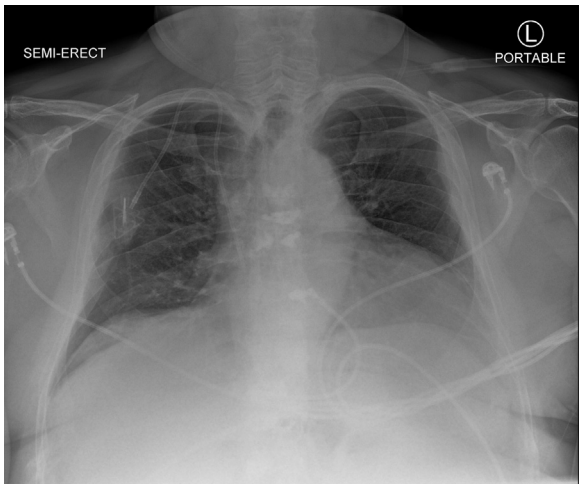

Figure 1. Anteroposterior chest radiograph taken shortly after arrival in the emergency department. A right-sided central line is present and projects over the area of the cavoatrial junction. Electrocardiograph leads and vertebroplasty artifacts also are noted. No focal infiltration or consolidation is identified.

did not show pneumonia. Laryngeal or upper airway edema was suspected due to new hoarseness, and the patient was started on nebulizer treatment with racemic epinephrine 0.25 mg. Arterial blood gas showed a pH of 7.32, $PaCO_2$ of 52 mm Hg, and PaO_2 of 37 mm Hg on room air, which was likely to be a mixed venous sample based on inconsistence with pulse oximetry data. When dyspnea did not improve after 2 h, he received a second nebulized racemic epinephrine treatment and a dose of potent glucocorticoid (dexamethasone 10 mg) administered intravenously.

Given the patient's hoarseness, a computed tomography (CT) scan of the neck was done. The scan identified a destructive expansile mass on the right side of the thyroid cartilage and posterior arch of the cricoid cartilage, with inferior extension medial to the left thyroid lobe that compressed the cervical trachea, causing partial obstruction (Figure 2).

The patient was at high risk for rapid respiratory decompensation and was admitted for awake tracheostomy on advice from our institution's head and neck surgery group. On day 7 postadmission, he underwent percutaneous gastrostomy for enteral nutrition support. The postoperative course was complicated by bilateral pneumonia due to cytomegalovirus and methicillin-resistant *Staphylococcus aureus*. He was on ventilator support and antibiotic and antiviral therapies for 10 days until his pneumonia improved. He underwent fractionated external beam radiotherapy to the tumors obstructing the upper airway. He was discharged to skilled nursing care after 27 days of hospitalization.

DISCUSSION

Plasma cell neoplasms are characterized by neoplastic proliferation of a single clone of plasma cells producing monoclonal immunoglobulins. They can present as either solitary (plasmacytoma) or multiple (MM) lesions. When solitary lesions occur in the soft tissues, it is called EMP. Only a handful of cases of EMP in the cricoid cartilage and trachea have been reported in the literature (5–8). ED presentation with wheezing and shortness of breath in a patient with a medical history of asthma is common. Upper respiratory infection often may precipitate or exacerbate asthma attacks. Patients with MM are usually immunodeficient, and thus, susceptible to respiratory infection (9). Although treatment with thalidomide derivatives increases the risk for venous thromboembolism, hoarseness and wheezing are not

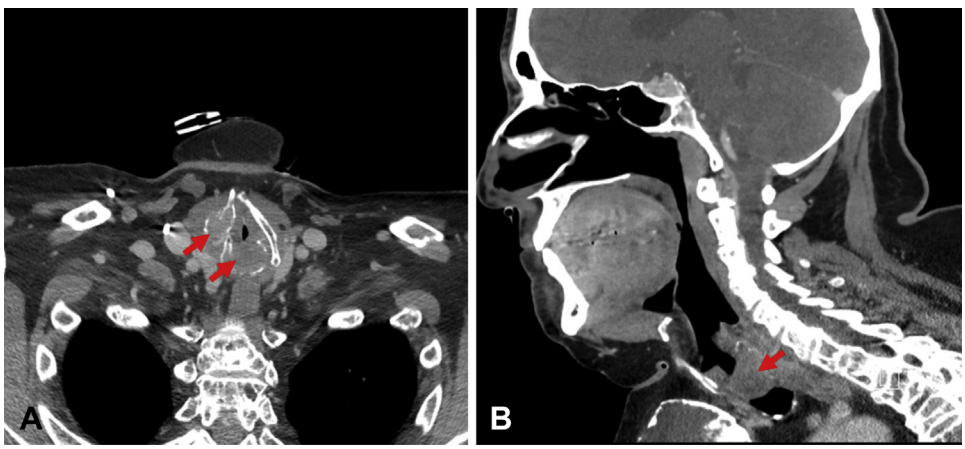

Figure 2. Computed tomography scan of the neck soft tissue with intravenous contrast. Destructive and expansile myeloma involving right side of the thyroid cartilage and posterior lamina of the cricoid cartilage is shown (arrows in Panel A) associated with soft tissue tumor protruding into the laryngeal airway and trachea (arrow in Panel B).

signs of pulmonary embolism (10). This patient's new-onset hoarseness suggested a differential diagnosis of laryngitis, laryngotracheitis, or laryngotracheobronchitis, depending on the extent of airway inflammation (11). Empirical treatment for obstructing laryngitis or laryngitis with asthma exacerbation using bronchodilators, steroids, antibiotics, and observation would have delayed definitive airway management. Despite mild carbon dioxide retention, the patient's lack of severe respiratory distress did not require emergency rapid-sequence intubation, allowing time to evaluate the patency of the upper airway. Without proper awareness of the location and extent of airway obstruction, rapid-sequence intubation without an adequate backup for emergency surgical airway creation could be fatal.

WHY SHOULD AN EMERGENCY PHYSICIAN BE AWARE OF THIS?

For the combination of symptoms of hoarseness, dyspnea, and wheezing, emergency physicians should include upper airway obstruction in the differential diagnosis in addition to acute laryngitis with asthma exacerbation. CT scan of the soft tissue of the neck is a highly useful diagnostic tool for evaluating the upper airway and for disclosing laryngeal pathologies and their relationship to adjacent structures (1,12,13). Multiplanar reconstructions can be used to evaluate the type, degree, and extent of airway narrowing in addition to the location of the tumor (14,15). Although emergency laryngoscopy or anteroposterior/lateral radiography of neck soft tissue would directly visualize a laryngeal airway obstruction, either would require a CT scan to plan a surgical intervention. It is also important to confirm the EMP diagnosis by biopsy under CT or ultrasound evaluation, as EMP and MM differ significantly as to their management; although MM is essentially treated with chemotherapy, irradiation has been shown to be most effective for EMP (1,16–18). If the patient is crashing with respiratory failure, emergency intubation may need to be attempted. Head and Neck Surgery may need to perform emergent tracheostomy or cricothyroid puncture if the endotracheal tube is unable to pass. It is vital for emergency physicians to be aware that not all dyspneas are attributable to asthma and that hoarseness is a tip-off for a laryngeal problem. Careful evaluation of the entire airway and a knowledge of such rare cases would prevent unfavorable outcomes.

Acknowledgment—This work was funded in part by a grant from the US National Cancer Institute, P30 CA016672 (MD Anderson Cancer Center Support Grant). The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Cancer Institute or the National Institutes of Health.

S.-C.J.Y. was a member of an expert panel for Celgene, Inc. He also received funding support for clinical studies from DepoMed, Inc. and Bristol-Myer Squibb.

REFERENCES

1. Oztürk M, Mavili E, Gorkem SB, Cagli S, Yüce I. An unusual cause of dyspnea: myelomatous involvement of cricoid cartilage. A case report. Neuroradiol J 2008;21:584–6.
2. Sukumaran R, Nair RA, Jacob PM, Koshy SM, Mathew AP. Extramedullary plasmacytoma of the trachea. Head Neck Pathol 2014;8:220–4.
3. Wang K, Huang Y, Chen A. A case report of tracheal extramedullary plasmacytoma. Chin J Clin Oncol 2008;5:311–2.
4. Chari A, Suvannasankha A, Fay JW, et al. Daratumumab plus pomalidomide and dexamethasone in relapsed and/or refractory multiple myeloma. Blood 2017;130:974–81.
5. Haser GC, Su HK, Pitman MJ, Khorsandi AS. Extramedullary plasmacytoma of the cricoid cartilage with solitary plasmacytoma of the rib. Am J Otolaryngol 2015;36:598–600.
6. Kalina P, Rykken JB. Multiple myeloma of the thyroid cartilage. Case Rep Hematol 2012;2012:194797.
7. Mitchell HK, Garas G, Mazarakis N, McGlashan J. Extramedullary relapse of multiple myeloma in the thyroid cartilage. BMJ Case Rep 2013;2013. bcr2013200689.
8. Zhang X, Su L, Ran Y-G, et al. Extramedullary plasmacytoma of the trachea: a case report and review of the literature. Medicine (Baltimore) 2018;97:e9594.
9. Anderson KC. Multiple myeloma: a clinical overview. Oncology (Williston Park) 2011;25(suppl 2):3–9.
10. Fotiou D, Gerotziafas G, Kastritis E, Dimopoulos MA, Terpos E. A review of the venous thrombotic issues associated with multiple myeloma. Expert Rev Hematol 2016;9:695–706.
11. Rana RS, Moonis G. Head and neck infection and inflammation. Radiol Clin North Am 2011;49:165–82.
12. Kuo GP, Torok CM, Aygun N, Zinreich SJ. Diagnostic imaging of the upper airway. Proc Am Thorac Soc 2011;8:40–5.
13. Lee KS, Boiselle PM. Update on multidetector computed tomography imaging of the airways. J Thorac Imaging 2010;25:112–24.
14. Ferretti G, Bithigoffer C, Righini C, Arbib F, Lantuejoul S, Jankowski A. Imaging of tumors of the trachea and central bronchi. Radiol Clin North Am 2009;47:227–41.
15. Olmedo G, Rosenberg M, Fonseca R. Primary tumors of the trachea: clinicopathologic features and surgical results. Chest 1982;81:701–6.
16. Maniglia AJ, Xue JW. Plasmacytoma of the larynx. Laryngoscope 1983;93:741–4.
17. Yoskovitch A, al-Abdulhadi K, Wright ED, Watters AK, Chagnon F. Multiple myeloma of the cricoid cartilage. J Otolaryngol 1998;27:168–70.
18. Peretti G, Piazza C, Provenzano L, Giudice M. Multiple myeloma involving the cricoid cartilage. Otolaryngol Head Neck Surg 2005;133:643.