



Airway Management of Myeloid Sarcoma of the Larynx: A Case Report

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ABSTRACT

Objectives: To (1) describe a rare case of laryngeal myeloid sarcoma and the initial management and (2) review the current literature.

Study Design: Case report and review of the literature

Methods: A case of laryngeal myeloid sarcoma will be reported and the current literature will be reviewed. Presenting symptoms, physical exam findings, imaging, pathologic examination of an excised tumor specimen, and serial endoscopic findings prior to and following induction chemotherapy will be included.

Results: A 70-year-old male with an 8-week history of multiple, slowly enlarging subcutaneous nodules presented with 7 days of intermittent hoarseness. CT revealed diffuse homogeneous masses of the subcutaneous tissues, including a left pyriform sinus mass and a circumferential subglottic mass at the level of the cricoid cartilage. Flexible laryngoscopy confirmed a markedly narrowed airway. Biopsy of a subcutaneous nodule was diagnosed as myeloid sarcoma. Pathologic evaluation of peripheral blood and bone marrow showed acute myeloid leukemia with >75% atypical, CD56 positive blast cells. Administration of induction chemotherapy with cytarabine and idarubicin resulted in rapid regression of the lesions, avoiding the need for tracheostomy.

Conclusion: Myeloid sarcoma is a rare manifestation of acute myeloid leukemia that can present with progressive laryngeal involvement. Prompt evaluation and confirmation by an otolaryngologist is a necessity. Timely administration of standard induction chemotherapy can result in rapid regression of these lesions. This can allow for cautious but conservative management of the airway, potentially avoiding the need for tracheostomy.

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INTRODUCTION

Myeloid sarcoma (MS) is a rare extramedullary manifestation of acute myeloid leukemia.¹ While the head and neck is a common extramedullary site for hematologic malignancies, myeloid sarcomas of the larynx represent less than 1% of all hematologic malignancies in this location.² Presentation of laryngeal MS can vary from asymptomatic as an incidental finding, to dysphagia, dyspnea, hoarseness, stridor, and respiratory distress. In these patients, establishing an airway is of foremost importance. Endotracheal intubation is preferable due to its decreased morbidity, although tracheostomy is sometimes necessary.

Preferred treatment is with standard induction chemotherapy with cytarabine and idarubicin, with the expected survival of these patients well represented in the literature. There are no reports demonstrating the timing of resolution of these lesions in response to systemic chemotherapy, nor discussing conservative airway management in patients with laryngeal involvement who receive systemic therapy. Here we present a case of secondary laryngeal myeloid sarcoma in a 70-year-old patient who received timely systemic therapy and avoided tracheostomy.

CASE DESCRIPTION

A 70-year-old man presented to an outside surgery clinic with complaints of multiple slowly enlarging subcutaneous nodules for 8 weeks associated with a 10 pound weight loss and 7 days of intermittent hoarseness. He denied fevers, chills, night sweats, dyspnea, stridor, dysphagia, odynophagia, otalgia, or musculoskeletal pain. On physical examination there were multiple firm, fixed, subcutaneous nodules overlying the abdomen and extremities. Blood work found a white blood cell count (WBC) of 35.8 thousand per microliter (K/ μ l) with 53% blast equivalents. Contrast CT scan incidentally found, in addition to the above masses, a homogenous left supraglottic mass extending to the midline and a homogeneous circumferential subglottic mass at the level of the cricoid cartilage narrowing the airway (**Figure 1**). Excisional biopsy of an abdominal lesion was performed under local anesthesia and confirmed a diagnosis of acute myeloid leukemia with extramedullary manifestation, or myeloid sarcoma.

The patient was airlifted to the Salt Lake City VA Hospital for definitive treatment. A PICC line was placed for induction chemotherapy in the MICU, and otolaryngology was consulted for an airway evaluation. Flexible laryngoscopy found a 0.5cm midline nodule at the base of tongue, a mucosalized left supraglottic fullness that extended from the aryepiglottic fold to the midline obscuring the left true vocal fold except when completely adducted, as well as diffuse intraluminal circumferential swelling at the level of the cricoid cartilage with an airway patency of approximately 4mm (**Figure 2**). Elective intubation and tracheostomy were both considered to stabilize the airway. However, considering the patient was clinically stable and the oncologists were confident the lesions would shrink rapidly with chemotherapy, we elected to observe the patient in the MICU with a low threshold for fiberoptic intubation or awake tracheostomy if the patient showed any sign of airway compromise. The patient was started on a “7+3” chemotherapy regimen with cytarabine and idarubicin.

After 24 hours of chemotherapy the patient’s hoarseness had resolved. Repeat flexible laryngoscopy showed ~10% regression of the laryngeal lesions. Serial endoscopy after 24 hours, 4 days, and 7 days showed progressive and near complete resolution of the lesions, with only minimal residual fullness in the left supraglottis (**Figure 3**).

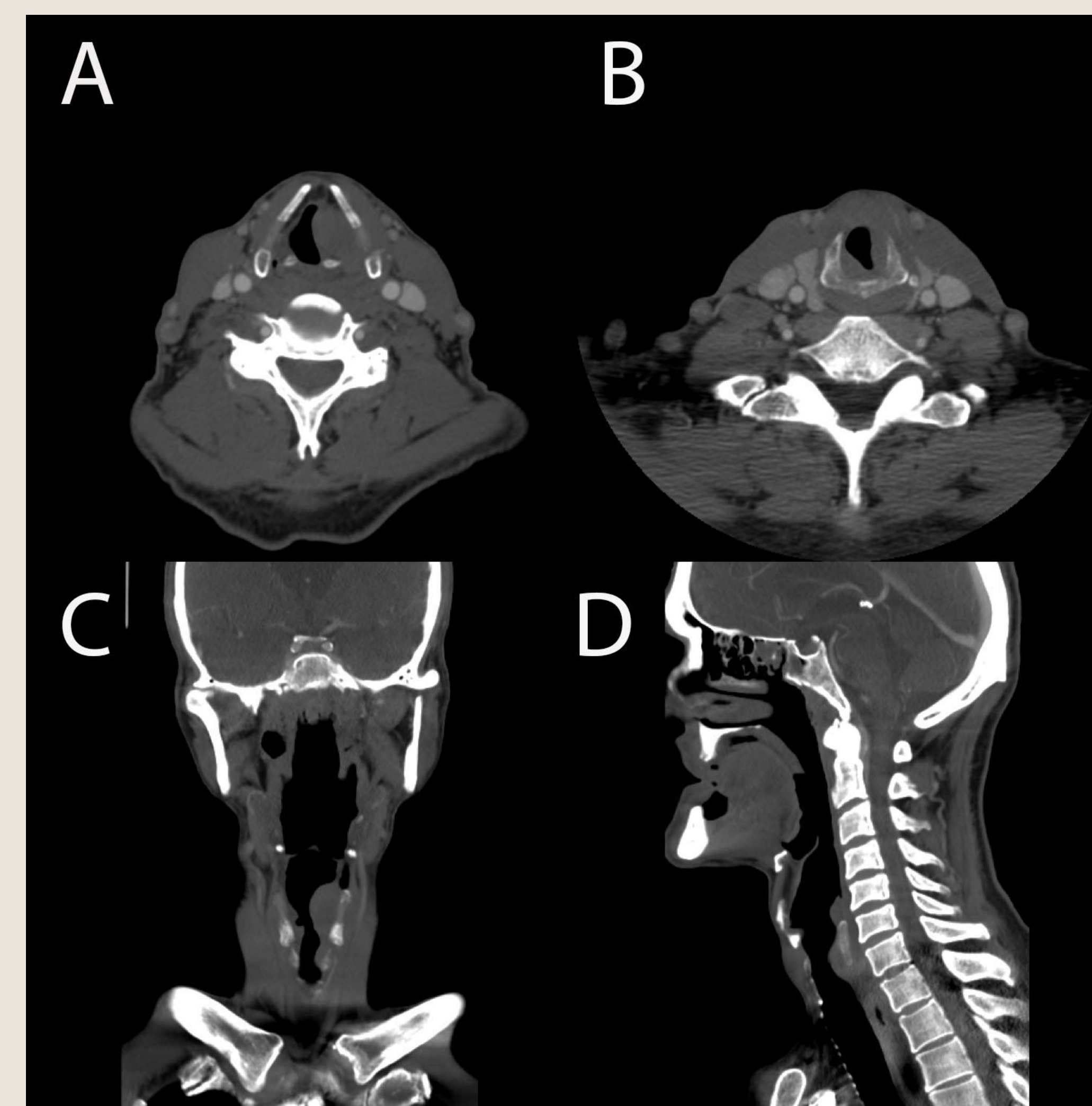


Figure 1. Representative axial (A,B), coronal (C), and sagittal (D) CT scan images from initial presentation.

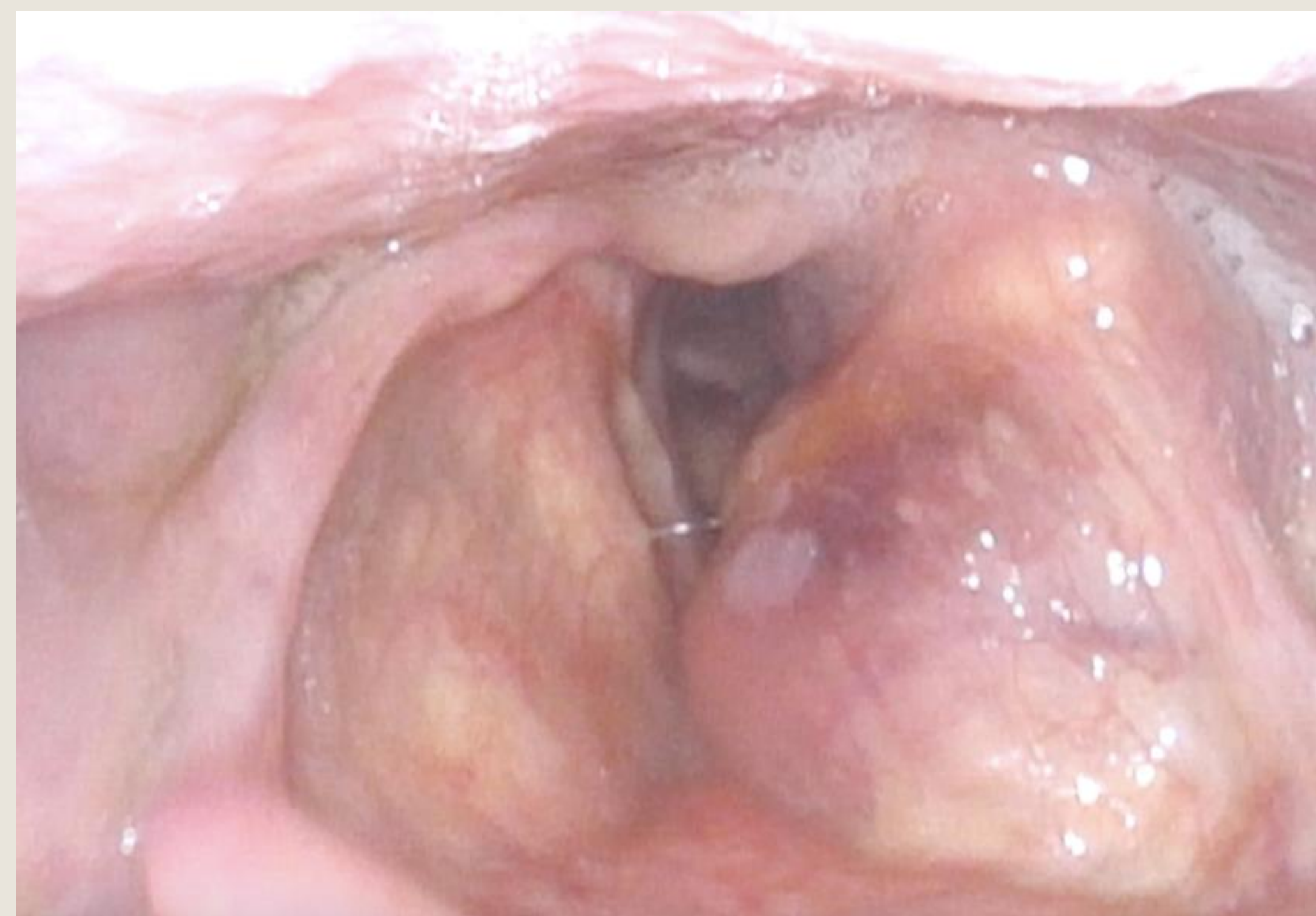


Figure 2. Flexible laryngoscopy at initial presentation.



Figure 3. Flexible laryngoscopy 24 hours after induction chemotherapy.



Figure 4. Flexible laryngoscopy 7 days after induction chemotherapy.

DISCUSSION

Myeloid sarcoma is a rare extramedullary neoplastic proliferation and accumulation of atypical myeloid blasts. While these lesions can manifest anywhere in the body, 12% occur in the head and neck, although only 1% occur in the larynx. Symptoms in patients with laryngeal involvement can range from asymptomatic to respiratory distress. In a recent review of the literature, Kim et al found that of the 20 cases reported, 8 patients presented with no respiratory symptoms, 5 had stridor, 4 had throat pain, 3 had dyspnea, and 2 each had hoarseness and dysphagia.^{2,4,7} Of the 8 cases where initial airway management was discussed, 5 underwent tracheostomy at either initial presentation or subsequent encounters. Only two cases discussed clinical response, with a significant decrease in laryngeal tumor burden on imaging 4 weeks following treatment.^{2,8}

This is the first report in the literature discussing the rate of regression of laryngeal myeloid sarcoma with induction chemotherapy. Yilmaz et al found that the use of systemic chemotherapy with cytarabine and an anthracycline antibiotic or anthracenedione similar to induction treatment protocols for AML yielded complete remission in 65%, decreased the rate of progression to acute leukemia, and prolonged survival (median 20 months) compared to local therapy, though the interval to expect a decrease in tumor burden is not discussed.⁹ Pullarkat discusses the usefulness of interval bone marrow biopsy to assess for blast clearance and assess complete remission in AML; however, extramedullary disease is not addressed.¹⁰

In patients with any etiology of glottic obstruction, establishing and maintaining a secure airway is of utmost importance. While this patient did have hoarseness and objective findings of partial laryngeal obstruction, he was communicative and ventilating without difficulty, and impending complete obstruction was felt to be unlikely. Of paramount importance in the decision to forego tracheostomy or intubation in this case is the known diagnosis at presentation and the immediate administration of systemic therapy by the oncology service.

CONCLUSIONS

We present the first report in the literature discussing the rate of regression of laryngeal myeloid sarcoma with induction chemotherapy and the possibility for cautious but conservative airway management in such a scenario. While a rare diagnosis, otolaryngologists should be aware of its existence and the potential for airway compromise associated. In a scenario of a definite diagnosis of myeloid sarcoma and the ability to rapidly begin induction chemotherapy, patients with a stable airway can potentially avoid tracheostomy and the associated morbidity.

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