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CASE REPORT

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Laryngeal leukemia cutis: A rare clinical subtype

Jaijeet S. Toor[1*], Fernando Ocampo-Gonzalez[2] and Inna Husain[3]

[1]Department of Surgery, Rush University Medical Center, Chicago, Illinois, USA
[2]Department of Pathology, Rush University Medical Center, Chicago, Illinois, USA
[3]Department of Otolaryngology—Head and Neck Surgery, Rush University Medical Center, Chicago, Illinois, USA

Abstract

Leukemia is a neoplastic growth of hematopoietic cells in the bone marrow that invades blood and other peripheral tissues. Infiltration by neoplastic leukocytes of the epidermis, dermis, or subcutis is known as leukemia cutis. A 64-year-old female with past medical history of hypothyroidism, diabetes, and hypertension presented with progressive dysphonia and dysphagia associated with eruptions of various skin lesions. She was found to have similar lesions in her oropharynx and larynx. A punch biopsy of the skin confirmed leukemia cutis. The patient received chemotherapy and all lesions previously seen resolved.

Leukemia cutis is a rare presenting symptom of acute myeloid leukemia (AML). Oral manifestations of leukemia are rare and often present as lymphadenopathy, ulceration, or bleeding, not as nodular lesions. Laryngeal leukemia cutis should be included in the differential diagnosis of poorly differentiated skin lesions with symptoms of dysphonia, dysphagia, or dyspnea. Patients presenting as such should be referred to an otolaryngologist for evaluation.

Keywords: *larynx, leukemia cutis, pharynx, skin neoplasm*

Main Points:

- *This is the first case report in the literature of leukemia cutis lesions seen in the larynx.*
- *After chemotherapy, the skin, laryngeal, and oropharyngeal lesions initially seen resolved completely.*
- *Patient presenting with poorly differentiated skin lesions and dysphonia should be referred to an otolaryngologist.*

Introduction

Leukemia is defined as a neoplastic growth of hematopoietic cells in the bone marrow that invades blood and other peripheral tissues. Leukemia cutis is the infiltration by neoplastic leukocytes of the epidermis, dermis, or subcutis, and often results in distinguishable cutaneous lesions [1,2]. Depending on the leukemia, the frequency of leukemia cutis ranges from 3-30% [1], but has been most commonly described in acute myeloid leukemia (AML) [3].

Leukemia cutis occurs in approximately 3% of patients with AML and even less often in chronic leukemias [4]. The most common association occurs with acute myelomonocytic and monocytic differentiation with involvement in up to 50% of patients [4,5]. Leukemia cutis may develop concurrently, subsequently, or before the onset of systemic leukemia [5].

The most frequently involved anatomic locations are the lower extremities followed by the upper extremities, back, trunk, and face [6]. Most reports of leukemia cutis in the literature are case reports or case series. There is little to no literature regarding pharyngeal or laryngeal manifestations of leukemia cutis. Herein, we present a patient with leukemia cutis presenting with symptoms of dysphonia and dysphagia.

Case presentation

OA 64-year-old female with past medical history of hypothyroidism, diabetes, and hypertension presented with progressive dysphonia and dysphagia associated with eruptions of various skin lesions on her face, trunk, back, and legs. She was admitted for workup and evaluation of her skin lesions. Otolaryngology was consulted for evaluation of worsening dysphonia and dysphagia.

On physical examination, she was found to have numerous cutaneous lesions of her face and neck. Oropharyngeal examination demonstrated both exophytic and submucosal masses all around the mouth including on the tonsils and uvula (Figure 1). After written informed consent was obtained, flexible nasolaryngoscopy demonstrated bulky submucosal nodules along the soft palate, base of tongue, supraglottis, and arytenoids, and non-obstructing lesions of the true vocal cords (Figure 2).

A punch biopsy of a skin lesion on the patient's neck was performed by dermatology. The biopsy results showed leukemia cutis, acute myeloid, with monocytic features (Figure 3). The pathology was significant for sheets of large monocytoid blasts with a high mitotic rate within the dermis. On immunostaining, it lacked CD3, CD34, and CD117. However, it did show strong expression of

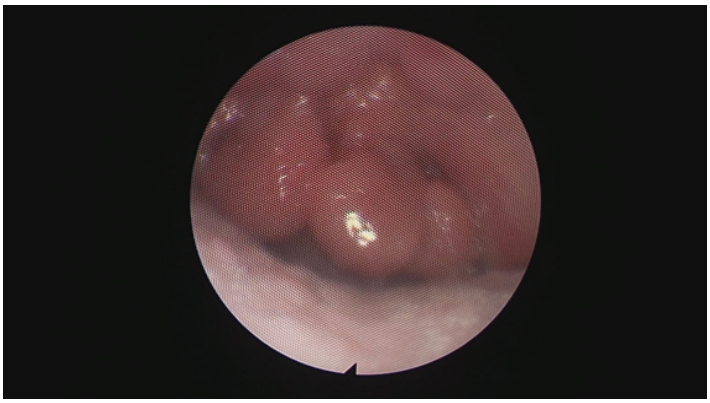

Figure 1: *Exophytic lesion of the uvula and soft palate.*

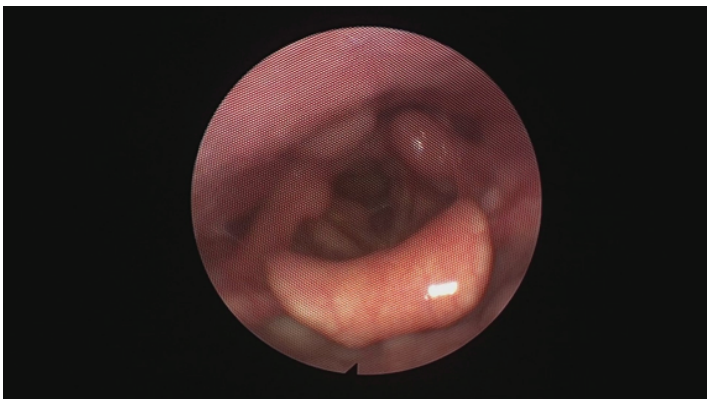

Figure 2: *Flexible nasolaryngoscopy showing submucosal nodules in the supraglottis and left arytenoid, and non-obstructing lesion on the true vocal folds with the cords abducted (A) and adducted (B).*

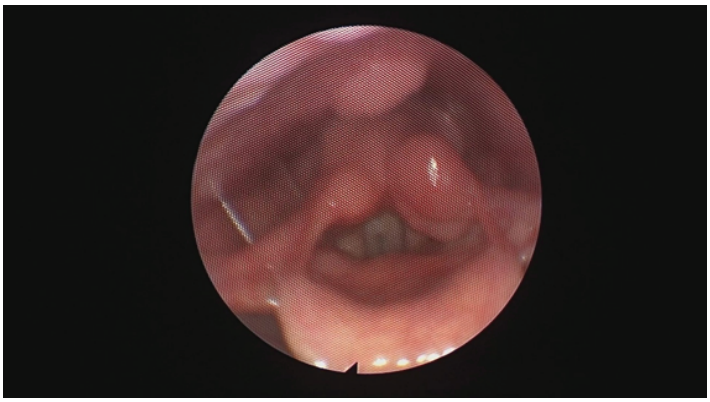

Figure 3: *Sections show a skin excisional biopsy with extensive sheet-like dermal involvement by medium to large blasts with round to ovoid or indented nuclei with open chromatin. No epidermotropism is identified.*

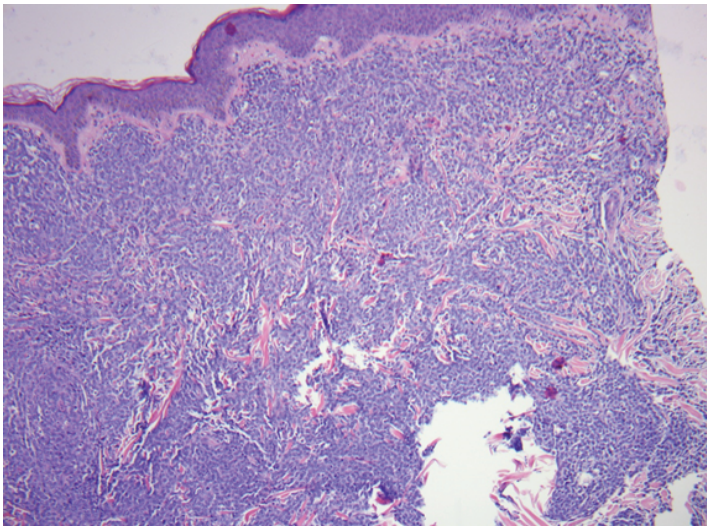

Figure 4: *Section shows a skin excisional biopsy with mild perivascular dermatitis and no evidence of the leukemic infiltrate.*

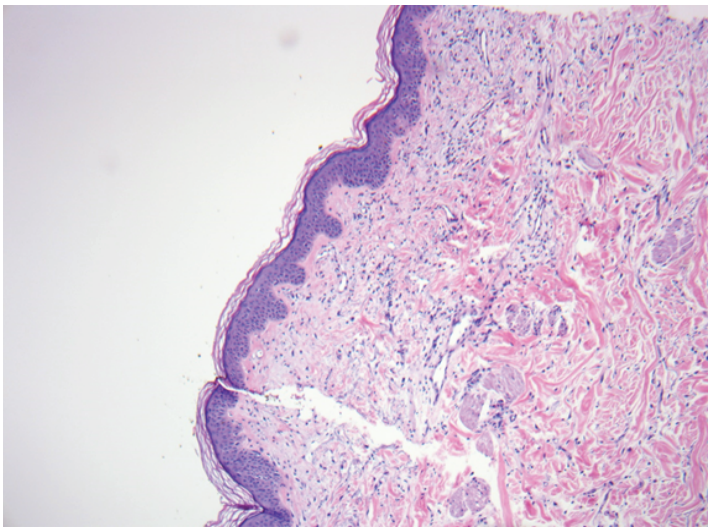

Figure 5: *Flexible nasolaryngoscopy showing resolution of previously seen subglottic, pharyngeal, and laryngeal lesions.*

CD4, weak expression of CD56, and weak to absent expression of CD123, which helps to exclude blastic plasmacytoid dendritic cell neoplasm. CD 33 was uniformly moderately expressed. The results confirmed the diagnosis of AML of monocytic lineage involving the skin. A bone marrow biopsy showed no marrow involvement or other extramedullary involvement, and normal cytogenetics. Per the hematology / oncology physicians, she was started on induction chemotherapy with cytarabine and mitoxantrone and had remarkable improvement. A repeat punch biopsy of the patient's chest at a site of a previous lesion was performed by dermatology. The biopsy results showed mild superficial perivascular dermatitis with no evidence of leukemic infiltrate (Figure 4).

One month after completing chemotherapy, patient presented for her follow up otolaryngology clinic visit. At that time she reported resolution of dysphonia, dysphagia, and cutaneous lesions. Repeat flexible nasolaryngoscopy demonstrated no oropharyngeal, pharyngeal, or laryngeal masses or lesions (Figure 5). The visualized portion of the subglottis was clear and all submucosal masses and nodules seen prior had resolved. Patient is pending stem cell transplant. All necessary informed consents were obtained from the patient.

Discussion

Leukemia cutis is a rare presenting sign or symptom of AML [7] occurring in 2.0-3.7% of patients with AML [3]. The identification of leukemia cutis is vital for both early diagnosis of systemic disease and for prognostic assessment. Previous studies have proposed that leukemia cutis has no preferred sites and the trunk, extremities, and head are all equally involved [1]. Leukemia cutis lesions can take different appearances and are highly variable in presentation. They are often firm and rubbery, and may look like flesh colored to violaceous plaques, nodules, or papules [8,9]. In rare cases, leukemia cutis lesions may precede the development of systemic disease, known as aleukemic leukemia cutis. This often occurs in patients who will eventually develop AML.

The pathophysiology and mechanism leading to the migration of leukemic cells to the skin is poorly understood. The proposed etiology is that various chemokine integrins and other adhesion cell molecules promote homing of T and B leukemic cells [9].

Oral manifestations of leukemia may occur, but they usually present as laryngeal pain, lymphadenopathy, gingival bleeding, oral blisters or ulceration, and gingival hyperplasia [8,10]. Ulcers and blisters occur less commonly on the skin.

There are no reported cases in the literature of leukemia cutis lesions appearing in the pharynx or larynx. In the case above, the lesions in the pharynx and on the skin resolved with chemotherapy, along with the patient's symptoms of dysphonia and dysphagia. Although a biopsy of the pharyngeal or laryngeal lesions was not done, the fact that the lesions resolved with chemotherapy at the same time as the skin lesions confirms that the lesions were the same pathologically leukemia cutis.

Pharyngeal and laryngeal manifestation of leukemia cutis is rare as shown by the lack of reported cases in the literature. Patients presenting with skin lesions consistent with leukemia cutis should be referred to an otolaryngologist for evaluation of pharyngeal lesions, especially if symptoms such as dyspnea, dysphagia, or dysphonia are endorsed. If these lesions do not respond to chemotherapy, or worsen with chemotherapy induction, then emergent intubation or operative intervention may be indicated.

Conclusions

Leukemia cutis is a rare lesion that can present in the pharynx or larynx. This is the first case report of leukemia cutis with laryngeal lesions causing dysphonia and pharyngeal lesions causing dysphagia. Laryngeal leukemia cutis should be considered in the differential diagnosis of poorly differentiated skin lesions with symptoms of dysphonia, dysphagia, or dyspnea. .

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*Correspondence: Jaijeet S. Toor, Mosc Department of Surgery, Rush University Medical Center, Chicago, Illinois, USA, Tel: 510-305-3308; E-mail: Jaijeet_S_Toor@rush.edu

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