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Unusual Laryngotracheal Manifestation of Rhinoscleroma in a Pediatric Migrant Patient

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Rhinoscleroma is a rare chronic granulomatous disease caused by the bacterium *Klebsiella rhinoscleromatis*. Although it predominantly involves the nasal cavities, it may also affect the larynx and upper trachea, resulting in various degrees of stenosis.[1,2] While several reports of rhinoscleroma have been detailed in endemic regions, including Central America, tropical Africa, Eastern Europe, and Southeast Asia, few cases have been reported in the United States.[1-3]

Herein, we report a case of rhinoscleroma with widespread laryngotracheal involvement in an unaccompanied pediatric migrant patient. This report was exempt from UCLA internal review board review.

Case Report

A 16-year-old Guatemalan unaccompanied migrant female, who carried a diagnosis of asthma, presented to our emergency department with biphasic stridor, increased work of breathing, and raspy dysphonia. She reported that her now 12-year-old brother had undergone emergency tracheotomy at 10 years of age for presumed asthma; he was later decannulated and symptom-free. After a chest x-ray showed a 5-cm segment of severe tracheal narrowing, she was urgently taken to the operating room for airway evaluation and management.

On microdirect laryngoscopy and bronchoscopy, there was severe redundancy of the anterior glottic complex with circumferential scarring of the anterior true vocal cords to the level of the subglottis (**Figure 1**). A 5-cm length of severely friable stenotic tracheal segments with no visible landmarks and multiple crypts of unclear etiology was noted throughout the proximal trachea. The stenotic laryngotracheal complex was soft and friable. The distal third of the trachea, carina, and mainstem bronchi had normal diameter and mucosa.

We proceeded with laryngotracheal balloon dilation (5 atmospheres of pressure held for 15 seconds, twice) to address the tracheal narrowing, which improved the airway from a diameter of 5 to 10 mm. Bleeding was controlled easily with topical epinephrine on cottonoid pledgets. Cultures were taken. Two granulomatous friable 1-cm lesions arising from the laryngeal surface of the epiglottis were biopsied. Bilateral nasal endoscopy revealed 3-mm nonobstructing friable lesions along the nasal septum, which were also biopsied.

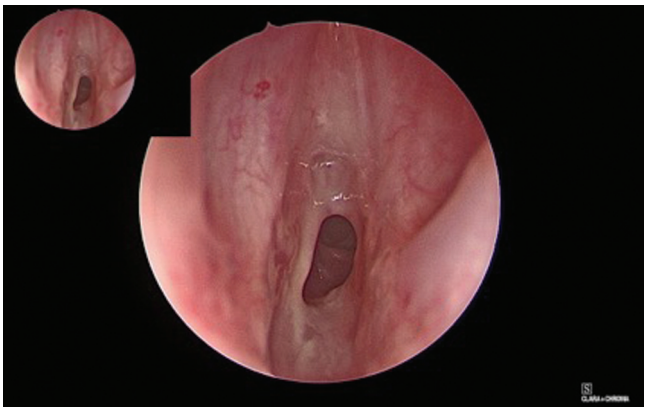

Figure 1. Intraoperative image of the glottis, indicating severe redundancy of the anterior glottic complex with circumferential scarring of the anterior true vocal cords to the level of the subglottis.

Postoperatively, there was immediate improvement with resolution of the biphasic stridor. Pediatric rheumatology was consulted to rule out autoimmune causes of secondary tracheal stenosis, granulomatous polyangiitis, and relapsing polychondritis. The patient had elevated c-ANCA at 1:640 with negative PR3. Biopsies from the septum and epiglottis, with sputum cultures that grew *K rhinoscleromatis*, confirmed

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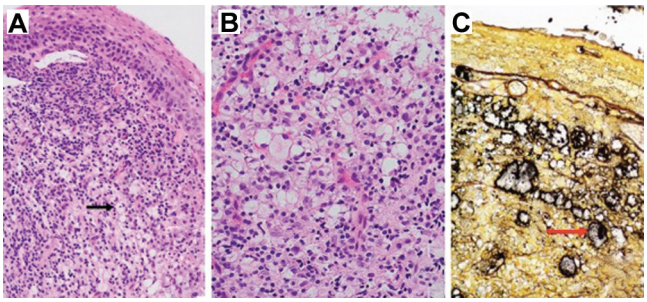

Figure 2. (A) Epiglottic biopsy histologic specimens reveal Mikulicz cells (black arrow) within a background of lymphocytes and Russell bodies. (B) Faintly stained organisms seen on higher magnification. (C) Warthin-Starry stain highlights intracellular bacilli (red arrow) within macrophages.

the diagnosis of rhinoscleroma (**Figure 2**). In consultation with the infectious disease team, we started sulfamethoxazole-trimethoprim, 1 tablet twice daily, for 3 months. The patient was discharged with plans for repeat laryngoscopy and bronchoscopy to determine disease evolution and need for further treatment.

Discussion

Rhinoscleroma is a difficult diagnosis in the United States, given the rarity of its presentation. With the increase in immigration from Central America and other endemic regions, otolaryngologists should be familiar with recognizing and managing this disease, including its less common laryngotracheal manifestations. Interestingly, in our patient, the intranasal aspect consisted of a subtle 3-mm nonobstructive septal lesion as compared with the more extensive laryngotracheal component. Postoperative computed tomography scanning of her sinuses and chest demonstrated no sinopulmonary involvement. Our patient had no symptoms of rhinorrhea or nasal obstruction, which are often documented in patients with rhinoscleroma, indicating that manifestations of this disease are variable.[1]

We showcased the safety and efficacy of limited balloon dilation for rhinoscleromatous laryngotracheal stenosis. Balloon dilation has been noted to be effective in other reports.[2,4] Nonetheless, tracheostomies were at times necessary to secure the airway in published accounts involving late-stage disease presentations.[1-4]

It is important to rule out rheumatologic conditions that present similarly. Patients with granulomatous polyangiitis will have an elevated c-ANCA with positive PR3, and patients with relapsing polychondritis may present with ear and/or nasal cartilage involvement, arthropathy, and ocular inflammation.[5]

Diagnosis of rhinoscleroma is based on histopathology notable for Mikulicz cells (foamy histiocytes with intracellular bacilli, ''moth-eaten'' cytoplasm), Russell bodies (large plasma cells with birefringent inclusions), and pseudoepitheliomatous hyperplasia.[4] Cultures support the diagnosis by confirming presence of *K rhinoscleromatis*. Although several antibiotic choices for treatment are detailed in published reports, sulfamethoxazole-trimethoprim was our choice given the microbiologic susceptibility, patient tolerance, and reports of its success.[3,4]

This unusual laryngotracheal manifestation of a rare disease process requires a familiarity and high level of suspicion when encountering symptomatic immigrants from endemic regions.

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Author Contributions

Laith Mukdad, study concept and design, manuscript drafting and revision; **Samira Nazzar Romero**, manuscript drafting and revision; **Deborah McCurdy**, manuscript drafting and revision; **Nina L. Shapiro**, senior surgeon, study concept and design, manuscript drafting and revision

Disclosures

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