Laryngeal Pilar Cyst Masquerading as an Internal/External Laryngocele

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ABSTRACT

OBJECTIVES: This study aimed to document and describe a case of a laryngeal pilar cyst and to review the literature.

METHODS: We describe the case of a 65-year-old woman with a laryngeal pilar cyst presenting with occasional ear pain and positional dyspnea, with imaging studies suggesting external/internal laryngocele. We also review the existing clinical literature.

RESULTS: Pilar cysts are adnexal skin lesions most commonly found in the scalp of elderly women. They generally have a benign course, but in rare instances single or multiple foci of proliferating cells can lead to the neoplastic formation of proliferating trichilemmal cysts, which carry malignant potential. Depending on the location of the cyst, pilar cysts may also present functional challenges for the patient.

CONCLUSIONS: Herein, we describe a pilar cyst in and around the larynx appearing initially as a laryngocele. Pilar cysts may present surrounding the larynx and may be mistaken for a vast array of pathologies. It is important to keep the differential broad when evaluating laryngeal masses.

KEYWORDS: Pilar cyst, trichilemmal cyst, larynx, laryngocele

Introduction

Pilar cysts (PCs), also known as trichilemmal cysts, are benign adnexal skin lesions typically occurring in areas with dense hair follicle concentrations, with approximately 90% presenting as scalp lesions.1 Per a recent review, PCs have also been documented in the neck, trunk, groin, mons pubis, vulva, and gluteal regions.1 There is a heavy female predominance to these lesions.1

Grossly, these cysts generally appear as firm, smooth-walled, rounded, cream white intradermal swellings containing semi-solid cheese-like keratinous debris.1,2 The histologic hallmark of PCs is the presence of a sharply circumscribed cyst with an “abrupt transition of a nucleated epithelial cell to an anucleate, keratinized cell without the formation of [an intervening] granular layer”.1 Leppard and Sanderson described regions of focal maturation within the epithelial wall of the PC, as well as around areas of calcification; these regions represent a potential budding zone that may produce multiple daughter cysts when the lesion undergoes a proliferative process.3

Case Presentation

A 65-year-old woman presented to clinic with a 10-year history of an anterior neck mass. She described a history of intermittent shortness of breath. She also described an extensive smoking history. On flexible fiberoptic laryngoscopy, a mass was noted that extended to the right supraglottic larynx and appeared to be just deep to the mucosa of the right aryepiglottic fold and the right pharyngoepiglottic fold. Computed tomography (CT) revealed a right-sided cystic-appearing lesion that appeared consistent with an internal/external laryngocele (Figure 1).

Based on this evidence and the possibility of laryngoceles harboring malignancy, the decision was made to take the patient to the operating room for direct laryngoscopy and excision of the neck mass.

On direct laryngoscopy, there was a submucosal mass of the right aryepiglottic fold. There also appeared to be an external component to the mass. Palpation of the external component on the anterior and lateral neck caused bulging of the mass in the hypopharyngeal and supraglottic regions. The presence of a significant external component extending anteriorly necessitated an external approach to excision.

The mass was identified just inferior to the right submandibular gland through a thyroplasty incision. The gland was retracted superiorly along with the marginal mandibular nerve. Dissection of the mass was carried back towards the thyrohyoid membrane, as the mass appeared to have an external and internal component in the supraglottic larynx. The mass was carefully dissected from the thyrohyoid membrane region. The bulk of the mass was adherent to the right thyroid ala, and this extended superiorly into the laryngeal vestibule and anteriorly around the thyroid cartilage to the contralateral side. The mass was carefully dissected off the thyroid cartilage and away from the laryngeal mucosa without violating the mucosa. On gross
examination, the mass appeared cystic and well encapsulated; it was filled with cheesy, curd-like material (Figure 2).

A direct laryngoscopy was again performed at the end of the procedure, revealing an improved laryngeal airway. The hemotoxylin and eosin (H&E) histopathology slides demonstrated a cyst lined with stratified squamous epithelium, without evidence of a granular layer (Figure 3). These findings were consistent with those seen in a PC. There was no evidence of malignancy.

Discussion

PCs are derived from dermal appendages and generally present as solitary intradermal or subcutaneous lesions. Clinically indiscernible from epidermal cysts, PCs can be distinguished from epidermal cysts histopathologically by their lack of keratohyalin granules. Both lesions exhibit keratinization.

PCs are a common benign finding in hair-bearing regions of 5% to 10% of the population, representing the most common cutaneous cyst in the scalp and the second most common cyst found in the head and neck region. To our knowledge, this report is the first documentation of a PC in and surrounding the larynx.

With consideration of radiographic imaging and flexible laryngoscopy, our initial impression of the neck mass was that of an internal/external laryngocele. Laryngoceles generally arise from the saccule at the anterior aspect of the laryngeal ventricle and track out of the larynx through the thyrohyoid membrane. The lesions may present functional difficulties for the patient, including dysphonia, dyspnea, dysphagia, cough, and a neck mass that becomes more prominent during Valsalva maneuver.

Laryngoceles carry a 10% to 15% risk of harboring malignancy, and given the patient’s symptoms the recommendation was made to excise the lesion for definitive diagnosis. Other submucosal lesions in the differential in this location include saccular cysts, granular cell tumor, and malignancy such as chondrosarcoma.

In addition, PCs carry a 2% risk of neoplastic transformation when foci of replicating cells result in a proliferating trichilemmal cyst (PTC). PTCs can also arise de novo. Although being generally benign lesions, they may grow quickly and aggressively, leading to ulcerations and, in rare instances, malignant transformation resulting in metastasis. Anderson and Hodgkinson described the case of a malignant PTC of the scalp—the important clinical feature of this lesion that distinguished it from a benign cyst was the presence of superficial ulceration. Currently, there exist no definitive criteria to clinically differentiate a benign from a malignant PTC.
making careful histopathologic examination an important part of diagnosis and treatment due to its potential for atypical behavior and malignancy.

There is much debate over the cause of neoplastic transformation leading to epithelial proliferation in PCs. Karaman et al. documented a case of PC in the neck region where trauma from shaving may have resulted in an inflammatory process that resulted in proliferation. When trauma results in a breach of the PC wall, inflammatory cells aggregate inside the cyst to induce healing from along the margins of the breach and in most cases leads to uncomplicated resolution. In some instances, the inflammation results in hyperproliferation to produce a pseudoepitheliomatous change.3 The latter is concerning in that it can resemble a well-differentiated squamous cell carcinoma,3 which must also be included in the differential diagnosis for a PC lesion. Other important considerations in the differential include thyroid and sweat gland tumors.1

The symptoms of a PC in the neck region are similar to those of many other neck masses. Our patient had complaints of a visible anterior neck mass along with hoarseness and intermittent shortness of breath but no pain. At 1-month follow-up after surgical excision of the lesion, the patient reported that she no longer had any positional dyspnea and that her hoarseness was improving.

Conclusions
Herein, we describe a PC in and around the larynx appearing initially as a laryngocele. PCs may present surrounding the larynx and may be mistaken for a vast array of pathologies. It is important to keep the differential broad when evaluating laryngeal masses.

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Author Contributions
CMK was the primary author of the manuscript. MAH and KAN were involved in the operation and obtained imaging, intra-operative photographs, and pathology. All authors reviewed the manuscript.

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