Case Report

Oncocytic Papillary Cystadenoma, an Unusual Variant Presenting as a Laryngeal Ventricular Cyst

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ABSTRACT: Cystadenoma arising from the larynx is a rare benign minor salivary gland tumor that can show mucinous or papillary morphology. The epithelial lining of the salivary gland tumor can present with oncocytic features, which is attributed to an increased number of mitochondria. We present a rare case of oncocytic papillary cystadenoma (OPC) of the larynx which has a combination of these features. The WHO defines OPC tumors as entities which closely resemble Warthin tumor, but lack its classic lymphoid component. The immunohistochemical profile and molecular genetic features are largely unknown. We present an 84-year-old female, former smoker, who presented with progressive dysphonia, dysphagia, and shortness of breath. Laryngoscopy revealed a large, smooth mass originating from the ventricle of the right vocal fold. Subsequent biopsy demonstrated cyst wall fragments lined by a bilayer of large columnar to cuboidal oncocytic cells that had granular eosinophilic cytoplasm, round to oval nuclei with finely dispersed chromatin, and small but distinct nucleoli. The surrounding stroma was slightly fibrotic with scant lymphoid elements. No nuclear pleomorphism, increased mitosis, or necrosis was identified. In the larynx, benign salivary gland tumors are rare and less frequent than malignant neoplasms. Awareness of rare benign entities like OPC help ensure proper management and aid in avoiding unnecessary therapy.

KEYWORDS: *Cystadenoma, salivary gland, laryngoscopy.*

Introduction

Oncocytic papillary cystadenomas (OPCs) arising in the larynx are rare benign lesions.

These lesions resemble laryngeal cysts but are lined by an epithelial bilayer comprised of columnar or cuboidal oncocytic cells with granular eosinophilic cytoplasm.

The presence of numerous hypertrophied mitochondria account for the eosinophilic cytoplasm of these cells.

OPCs are thought to arise from metaplastic seromucinous gland epithelium associated with degenerative processes, cellular aging, and inflammation from irritants such as smoking [1].

The majority of cases typically involve the minor salivary glands, although cases of major salivary gland involvement have been reported the parotid gland being the most affected [2].

Less than 200 cases of laryngeal OPCs have been reported [3,4]; they comprise less than 1% of all benign tumors of the larynx [5].

Cases of laryngeal involvement are typically confined to the supraglottic region, commonly in the aryepiglottic fold, false vocal folds, and laryngeal ventricles due to the abundance of seromucinous glands in these regions.

The free edges of the true vocal folds are not involved as glandular epithelium is not present in this area [1].

Subglottic laryngeal lesions are extremely rare but have been reported [6].

OPCs of the larynx typically present as single solitary lesions, although diffuse, multifocal occurrences have been reported [4].

The signs and symptoms of OPCs experienced by patients depend on the size of the lesion and its proximity to the glottis; larger lesions and those closer to the glottis are more likely to cause symptoms which may be severe, such as upper airway obstruction [4].

Symptoms can range from hoarseness, dysphonia, dyspnea due to airway obstruction, stridor, or pain.

Hoarseness is the most common symptoms and may last for weeks to years before diagnosis [1].

Diagnosis is based on patient's history, endoscopic exam, and histologic features, which is vital in differentiating OPCs from malignant lesions [7].

Preferred treatment is complete resection using transoral microlaryngeal surgery; however, open laryngeal surgery may be considered depending on the size of the OPC [7].

Herein, we report a classic case of OPC involving the supraglottic region of the larynx with recurrence after marsupialization.

Case Presentation

An 86-year-old female with a five-pack year history of smoking cigarettes, presented with progressively worsening dysphonia.

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She also reported dyspnea on exertion, dysphagia with solid food, and coughing with liquids such as water.

The patient denied hemoptysis, odynophagia, otalgia, recent lower respiratory infection, and weight loss.

Past medical history was unremarkable. Transnasal flexible laryngoscopy with stroboscopy revealed a large, smooth mass originating from the right ventricle that would intermittently ball valve into the subglottis, which was widely patent.

The mass partially obstructed the visual field at the right anterior true vocal fold.

Additionally, the mass disrupted the mucosal wave propagation of the right true vocal fold, thus creating asymmetry posteriorly.

Otherwise, the true vocal folds were fully mobile bilaterally, and there was no evidence of muscle tension dysphonia or glottic gap.

The vallecula and the post-cricoid region were free of lesions.

Under direct suspension laryngoscopy, a CO_2 laser was used to excise the cyst causing cyst rupture with the spillage of thick yellow-white exudative fluid.

Histologic examination of the excised mass revealed clusters of oncocytic epithelial cells and scattered individual cells.

The cells had distinct borders, finely granular cytoplasm, and uniform, round nuclei with small, centrally placed nucleoli (Figure 1).

The exudate was cultured and few *Streptococcus mitis*, rare *Neisseria* species, and *Rothia mucilaginosa* species were isolated and identified.

At one-month follow-up, the patient reported improvement of her voice, however laryngoscopy revealed obvious regrowth of the right ventricular lesion.

The finding was discussed with the patient and observation was recommended given the benign nature of the lesion.

The patient was instructed to return to the clinic in six months or sooner if symptoms developed, but the patient was lost to follow-up.

An informed consent was obtained from the patient regarding the publication of the data.

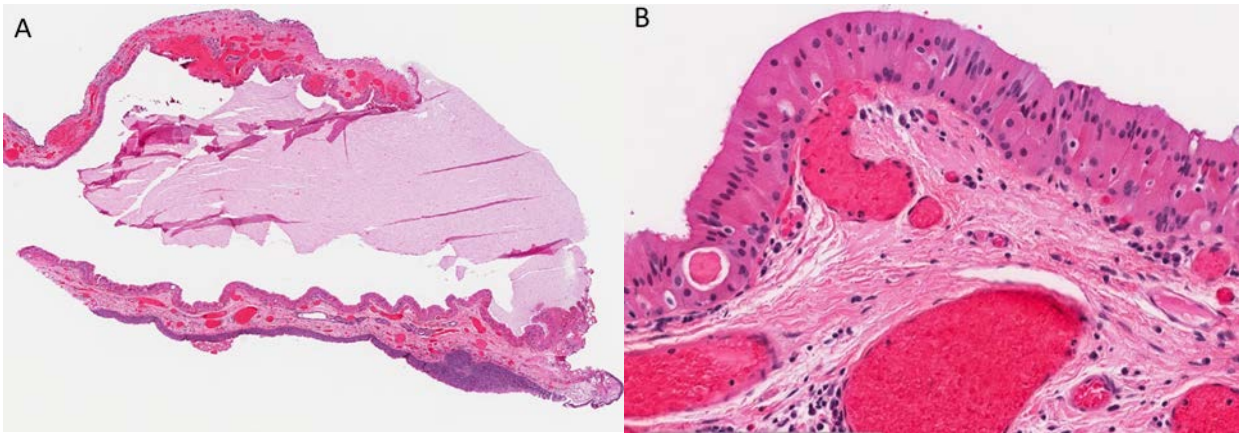

Figure 1. A, H&E 10X, B, H&E 20X: Cyst wall fragments lined by a bilayer of large columnar to cuboidal oncocytic cells with granular eosinophilic cytoplasm, round to oval nuclei with finely dispersed chromatin, and small, distinct nucleoli. The surrounding stroma is slightly fibrotic with scant lymphoid elements.

Discussion

Cystadenomas can be divided into papillary and mucinous subtypes.

The papillary subtype has unilocular or multilocular cysts with intraluminal papillary projections lined by a cuboidal or columnar epithelial bilayer.

Due to histological similarities, OPCs are often confused with Warthin tumors (papillary cystadenoma lymphomatosum).

Both tumors are comprised of papillary structures and oncocytic cells, however Warthin tumors differ from OPCs in that the stroma is rich in lymphocytes whereas the stroma of OPCs lack this feature [2].

Another tumor that is histologically similar is intraductal papilloma. Intraductal papilloma also have papillary projections, but they are more complex than OPCs [2].

Additionally, oncocytoma have oncocytic cells but lack the papillary projections characteristic of OPCs [2].

Laryngoceles may also mimic OPCs as they both present as ventricular masses on imaging and endoscopy, and both have similar clinical presentations [7].

However, a laryngocele has air within its cyst [8].

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Laryngeal OPCs usually occur in elderly patients in their 70s and 80s, however cases in younger patients have been reported [1,4].

Women are more commonly affected than men.

Several reports suggest a link between a history of smoking and laryngeal OPCs [1,4,6,9,10].

Patients with laryngeal cysts typically present with hoarseness, dysphonia, dysphagia, globus sensation, and rarely stridor, pain, or airway compromise [6,11].

The pathogenesis of oncocytic papillary cystadenomas remains a topic of debate.

Oncocytic differentiation has been identified in the epithelial lining of the tongue, salivary and lacrimal glands, pharynx, esophagus, gallbladder, as well as within the parenchyma of the thyroid, parathyroid, thymus, liver, pancreas, adrenal gland, kidney, and breast.

Some authors believe that these lesions are a result of a true neoplastic process.

Others argue that they are a result of epithelial hyperplasia rather than neoplasia due to the papillary infoldings [1,4].

Additionally, it has been hypothesized that oncocytic metaplasia is a result of physiological changes related to functional "wear and tear" [1].

This hypothesis is based on the fact that thyroid tumors composed of oncocytes are rarely functional [12].

OPCs commonly present as solitary lesions with involvement of a single site, nonetheless, multiple diffuse, multifocal cysts may occur [1,4].

Malignant transformation of OPCs in the larynx have not been reported, although Stenner et al. reported a case of squamous cell carcinoma arising in multifocal pharyngolaryngeal OPC.

The report hypothesized that such occurrence was a result of tumor collision rather than malignant transformation due to the absence of clear transitions from oncocytic to carcinoma cells [9].

Conclusion

We present a rare case of oncocytic papillary cystadenoma located in the larynx.

As this is an uncommon site for this entity, histology is essential in differentiating OPC from other tumors with similar features and allows for quick and appropriate treatment initiation.

Complete resection with CO_2 laser or cold instruments is the treatment of choice and is associated with a good prognosis.

However, recurrence is common, especially in cases with multifocal involvement.

Hence, follow-up with endoscopic exams are recommended.

Conflict of interests

None to declare.

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