Chondrosarcoma of the Epiglottis: A Case Report and Literature Review

Jonathan Choi, BS; Andrew Victores, MD; Ashley Wenaas, MD; Julina Onkgasuwan, MD

Abstract

EDUCATIONAL OBJECTIVE: At the conclusion of this presentation, the participants should be able to discuss the epidemiology, etiology, histopathology, symptoms, radiologic features, and treatment of laryngeal chondrosarcoma (LCS).

OBJECTIVE: To report the sixth known case of LCS localized in the epiglottis and to discuss the literature regarding the classification scheme, etiology, histopathology, presentation, and treatment of LCS.

STUDY DESIGN: Case report and comprehensive review of the literature.

METHODS: This study reviewed the medical records of a patient with LCS of the epiglottis treated at a tertiary academic medical center. The MEDLINE database search contained the following key words: “larynx AND chondrosarcoma” and “chondrosarcoma AND epiglottis.”

RESULTS: A 71-year-old male smoker presented with sore throat, dysphagia, and odyphagia for 3 months. Computed tomography scan revealed an inhomogeneously enhancing necrotic mass emanating from his right epiglottis and traveling along the aryepiglottic fold. The patient subsequently underwent direct laryngoscopy with biopsy and histopathological examination of the excised specimen confirmed low-grade LCS of the epiglottis. Endoscopic and lobar resection was performed of the mass including the lateral portion of the epiglottis, pharyngoepiglottic fold, aryepiglottic fold, and arytenoid. The patient underwent pre and post-operative swallowing therapy and was able to take all food by mouth at discharge.

CONCLUSION: LCS is an exceedingly rare malignancy and least frequently report in the epiglottis. As of this writing, only five additional cases have been reported in the literature. This case study reinforces the need for histopathological examination in forming the basis for ongoing management and follow-up observations. This is noteworthy since chondrosarcoma (CS) are often misdiagnosed as chondromas. Timely surgery remains the optimal, primary means of treatment notwithstanding the rarity of LCS.

Introduction

Chondrosarcomas (CS) are a group of heterogeneous, malignant bone neoplasms originating to the chondroid (cartilaginous) matrix.1 While not restricted to any one location in the body, most cases of CS are localized in the long bones and pelvis, and approximately 10% are localized in the head and neck (e.g., sinonasal region, jaws, larynx, and skull base).2,3 CS is exceedingly rare in the head and neck account for approximately 0.1% of head and neck neoplasms.4 Laryngeal CS (LCS) accounts for approximately 1% of all laryngeal neoplasms and, though rare, is the most frequent non-epithelial neoplasm of the laryngeal region.5,6 Note that, as of this writing, this 1% estimate is the most frequently cited figure by modern scholars; however, earlier reviews have suggested an estimate ranging from 0.07% to 0.2% of all laryngeal neoplasms.7 Since the literature commonly documents CS via case reports, many of which are repeatedly cited by independent scholars, the more recent findings may be reporting an inflated incidence rate.8 The rarity of CS remains undisputed notwithstanding these potentially conflicting findings.

Laryngeal chondrosarcoma (LCS) primarily develops in the cricoid cartilage (75%-80%), and in particular the posterior lamina. LCS is then most prevalent in the thyroid cartilage (20%) which typically feature malignancies in the inferolateral wall, and is followed by LCS in the arytenoid cartilage (3%). LCS in the vocal cords and the epiglottis (1%) are exceedingly rare.1 As of this writing, only 5 additional cases of CS of the epiglottis have been reported in literature. We describe a case of CS of the epiglottis, which was treated via endoscopic en bloc laryngectomy.

Case Report

A 71-year-old man presented to the otolaryngology clinic with a 3-month history of sore throat. The patient was in his usual state of health until 3 months prior to initial presentation when he began to have a sore throat, more so on the right side. Since that time, the patient felt like his symptoms had progressed. The patient also had odynophagia and dysphagia to solids. The dysphagia improved when he turned his head to the right. The patient denied any voice changes or significant weight loss. He had a 75-pack-year smoking history. Flexible laryngoscopy revealed a mass extending from the right side of the epiglottis into the vallecula. The mass was ulcerative on both the lingual and laryngeal surfaces of the epiglottis. The two vocal folds were mobile and symmetric. Computed tomography (CT) scan demonstrated an inhomogeneously enhancing necrotic mass emanating from the right epiglottis and traveling along the aryepiglottic fold (Fig. 1). We performed direct laryngoscopy with biopsy. Histopathological examination of the excised specimen confirmed low-grade CS of the epiglottis. The patient underwent endoscopic en bloc larynx resection of the mass including the lateral portion of the epiglottis, pharyngoepiglottic fold, aryepiglottic fold, and arytenoid (Fig. 2 and 3). The postoperative course was uneventful; the patient underwent post-operative swallowing therapy and was able to take all food by mouth at discharge. The patient has remained asymptomatic.

Figure 1. CT scan with contrast: well-circumscribed heterogeneously enhancing necrotic mass emanating from the right epiglottis.

Figure 2. Submucosal mass on the vallecular surface of the right side of the epiglottis traveling along the aryepiglottic fold.

Figure 3. Resected specimen of laryngeal chondrosarcoma of epiglottis.

Conclusion

LCS is an exceedingly rare malignancy and least frequently reported in the epiglottis. Approximately 600 cases of CS have been reported in the literature, and of this total approximately 150 to 250 are cases of LC and LCS. The epiglottis is the least common primary site of LCS, and as of this writing only five cases have been reported in the literature. Thus, this paper’s findings are significant in part due to its rarity. In addition, this case study reinforces the need for histopathological examination in forming the basis for ongoing management and follow-up observations. This is noteworthy since CS are often misdiagnosed as chondromas. Timely surgery remains the optimal, primary means of treatment notwithstanding the rarity of LCS.

References

1. Epidemiology

Age of presentation 40-70 years.9,10
Male-to-female ratio 3 to 1.10,11
Grimes more likely to affect Whites than Blacks.2,11

Etiology and Histogenesis

Etiology not clear. Potentially associated with Teflon injection, radiation, or other neoplasms (e.g., spindle cell sarcomatoid carcinoma). Smoking identified in excess of 30% of patients.9

Histogenesis not clear. Five theories of neck and larynx (CS): (1) cartilage remnants; (2) ossified cartilage; (3) pluriportent mesenchymal/stem cells; (4) ischemic change; and (5) precursor syndromes and disorders.

Histopathology

Lichtenstein and Jaffe diagnostic criteria for malignant cartilaginous neoplasms of extralaryngeal bone origin adopted as diagnostic criteria for LCS.12

Symptoms, Radiologic Features, & Treatment

Most common symptoms: hoarseness (74%), dyspnea (56%), and dysphagia (21%). May also present with stridor. Dyspnea tends to increase gradually followed by an acute increase in severity. Vocal-cord paresis or fixation present in 50% of cases.14

CT A reliable modality for evaluating LCS particularly with respect to preoperative planning of partial laryngeal resection. Calcification displayed in 80% of cases. CT may also reveal newly formed ossous elements. However, MRI more useful than CT in delineating the extent of the lesion due to its improved soft-tissue contrast resolution.

Surgery the principal treatment modality. Various treatment modalities may be delineated into the following categories: (1) endoscopic removal; (2) laryngofissure; (3) extralaryngeal approach with respect to thyroid cartilage; (4) partial laryngectomy; and (5) total laryngectomy.1,3,9,10

Contact

Jonathan Choi
Baylor College of Medicine
Email: choi.joseph@gmail.com
Website: http://www.bcm.edu/oto
Phone: 817-542-1322