Laryngeal Chondrosarcoma with dedifferentiation to Spindle Cell Sarcoma of the Larynx

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ABSTRACT

Chondrosarcomas are rare malignant tumors of mesenchymal origin. First described in 1816, laryngeal chondrosarcomas are rare tumors of the head and neck.1 Laryngeal chondrosarcomas account for <0.2% of head and neck malignancies and <1% of laryngeal neoplasms.2 Due to the infrequent occurrence, treatment strategies are not well defined and borrow principles of management from sarcomas arising in many commonly affected areas. While conservative surgery is the primary treatment, laryngeal preservation does not supersede the application of sound oncologic principles.1

CASE REPORT

A 55 year old male with a history of alcohol and tobacco use, gout, diabetes mellitus and hypertension presented with hoarseness. Upon further evaluation, he was diagnosed with a low grade chondrosarcoma of the right cricoid cartilage. The patient was subsequently taken for a tracheotomy and laryngofissure to excise the lesion with a posterior cricoid split and rib graft interposition to reconstruct the larynx. Of note, bone-cutting instruments were required to perform the laryngofissure and tracheotomy. Postoperatively, the patient did well from this procedure (Figure 1). The patient was later decannulated, tolerated an oral diet and continued to do well for several years. Follow-up bronchoscopy at two years demonstrated a patent airway with a small right crico-arytenoid mass with mild luminal extension (Figure 2). Given the normally indolent growth of a low-grade chondrosarcoma, a decision to wait and observe clinically was followed. The tracheo-bronchial airways were without extension of disease. A year later he re-presented with dyspnea and stridor and was found to have an obstructing right posterior cricoid lesion on imaging and laryngoscopy (Figure 3). Biopsy confirmed the lesion to be a spindle cell sarcoma (Figure 4). After an extensive discussion of treatment options, the patient was taken for a total laryngectomy. The postoperative course was unremarkable and the patient did very well from the surgical standpoint he received post operative radiation and has new pulmonary lesions suspicious for metastases.

DISCUSSION

Introduction

Chondrosarcomas are the most common sarcoma found in the larynx.3 Chondrosarcomas can be classified as primary or secondary. Chondrosarcomas that arise de novo in cartilage and other tissues are primary, while chondrosarcomas that develop in pre-existing benign cartilaginous tumors are secondary. Chondrosarcomas of the larynx typically arise in the hyaline cartilage of the cricoid cartilage. No definitive etiology for laryngeal chondrosarcomas is known, although there is some acceptance of the speculation that disordered ossification of the laryngeal cartilages may be a sentinel event.4

Clinical Presentation

Men are more commonly affected than women with a ratio of men to women of up to 4:1.2,5,6 Laryngeal chondrosarcomas typically occur in the 6th to 9th decade of life with a peak in the 7th decade.2,5 Presenting complaints typically include hoarseness, dyspnea, dysphagia, odynophagia and a palpable neck mass.5,6 The cricoid cartilage is the most commonly affected laryngeal site followed by the thyroid cartilage, arytenoid cartilage and epiglottis.2,4 Computed tomography (CT) is the imaging modality of choice in head and neck chondrosarcomas, as it allows evaluation of abnormal areas of ossification and nodal status in the neck. Although magnetic resonance imaging (MRI) can define the extent of the tumor, it is a poor choice for delineating the calcified matrix seen in chondrosarcomas. The endoscopic appearance of the overlying surface mucosa of chondrosarcomas is generally smooth, intact and uninvolved by the tumor.2

Pathology & Staging

Histopathologic grading of chondrosarcomas was first described in 1943 by Lichtenstein and Jaffe.2 The current grading system (grade 1, 2, 3) stratifies disease on a continuum from low grade to high grade based upon microscopic features such as cellularity, nuclear size and pleomorphism, necrosis and mitotic activity. The histopathological classification has implications for tumor behavior and prognosis, but is subject to inter-observer variability.9 High grade tumors and metastases have been associated with a worse prognosis.10 The American Joint Commission on Cancer (AJCC) developed a staging system for chondrosarcomas, but due to its complexity and limited consideration for local invasion into vital head and neck structures it is rarely applied to head and neck chondrosarcomas.7

Figure 1. Bronchoscopy showing a well epithelialized rib graft (arrow).

Figure 2. Bronchoscopy showing a right crico-arytenoid mass (arrow). Note the smooth, uninvolved appearance of the overlying surface mucosa.

Figure 3. CT Neck axial image demonstrating a 2.5 x 2.7 x 2.0 cm obstructing right posterior cricoid lesion.

Figure 4. Light micrograph 20X demonstrating spindle cell sarcoma (A) adjacent to low grade chondrosarcoma (B), suggesting a dedifferentiated chondrosarcoma.

Figure 5. Light micrograph 20X demonstrating high grade spindle cell sarcoma.

Figure 6. Total laryngectomy specimen with tumor (arrow) within the cricoid cartilage.

Metastases & Recurrence

Local metastatic disease in the neck is rare and distant metastases are even less frequently encountered.14 The lung is the most common site of distant metastases in several reports.2,14 Recurrence rates in the literature have ranged from 18% to 40%, although given the nature of the disease and its treatment – it is difficult to discern whether a primary tumor was excised or if it recurred.2 There is some suggestion that chondrosarcomas may biologically progress with up to 13% of recurrent chondrosarcomas exhibiting a higher grade of malignancy than the original neoplasm.7 Given the propensity for late recurrences, head and neck chondrosarcomas should be followed continuously with endoscopic and radiographic examinations.7,15 Death directly attributable to chondrosarcoma is rare and generally the result of direct tumor growth into adjacent vital structures of the neck.2

Dedifferentiated Chondrosarcoma

Dedifferentiation has been noted to occur in several reports, as well as in our case.2,12,14 Dedifferentiated chondrosarcomas account for 10% - 15% of all chondrosarcomas. Dedifferentiated chondrosarcoma is a high-grade non-cartilaginous sarcoma typically arising from a pre-existing cartilage-forming tumor with a sharp interface between the two lesions (Figure 4).2 Limited reports of the genetic analysis of dedifferentiated chondrosarcomas and their adjacent lesions found that the two lesions possess genetic aberrations that suggest they may share a common precursor lesion that leads to both of their development.2 Dedifferentiated chondrosarcoma bears a significantly worse prognosis than their adjacent counterparts.12,13

CONCLUSIONS

This report describes a case of a laryngeal chondrosarcoma lesion recurring with dedifferentiation as a spindle cell sarcoma several years after initial diagnosis. Laryngeal chondrosarcoma is a rare disease entity with a relatively indolent course. Therefore, long term follow-up and monitoring is appropriate. However, as exhibited in our patient, laryngeal chondrosarcoma can undergo dedifferentiation to a more aggressive sarcoma. Treatment options include observation, radiation, conservative surgery and complete excisional surgery. Given the possibilities for disease progression and the various treatment options, a frank discussion between the physician and patient is of the utmost importance.

REFERENCES