



Images In Medicine

A Slow Growing Non-Calcified Airway Mass

Edmund H. Sears, MD, Matthew D. Jankowich, MD, and Terrance T. Healey, MD

LARYNGEAL CHONDROSARCOMA IS A RARE RELATIVELY SLOW-GROWING tumor arising from the laryngeal cartilages. The slowly progressive nature of symptoms means that many patients will undergo multiple diagnostic studies, and recognition of the imaging characteristics of the tumor may assist significantly in diagnosis. We present a case of a laryngeal chondrosarcoma lacking the calcifications which are often a diagnostic clue.

CASE REPORT

A 67-year old male presented with complaints of progressive hoarseness and stridor. The patient first noticed hoarseness of the voice five months prior to presentation at which time he was evaluated with CT of the neck (Figure 1), which was unremarkable. He also underwent direct laryngoscopy which revealed normal cords without lesions and normal movement. The hoarseness continued and a repeat CT was obtained 3 months later, which showed mildly increased prominence of the soft tissues in the posterior subglottic trachea. The patient then underwent rigid laryngoscopy under general sedation which showed mild supraglottic edema, but normal appearing cords with normal movement. Supraglottic and posterior laryngeal biopsies were done which showed hyperplastic squamous epithelium with keratosis.

Three days after his procedure the patient complained of shortness of breath, dyspnea, stridor, dysphagia, and drooling and presented to the ED for evaluation. Flexible laryngoscopy showed mobile cords with edema, and inability to evaluate the subglottic area.



Figure 1. Baseline CT of the neck at the time of onset of symptoms.

Chest radiography was unremarkable. CT of the neck with IV contrast showed abnormal soft tissue in the posterior aspect of the subglottic trachea surrounding the cricoid cartilage, with mild airway narrowing (Figure 2). There was no evidence of abnormal lymphadenopathy, and the visualized thyroid appeared normal.

Given his ongoing respiratory distress in the setting of an obstructing mass lesion, the patient underwent urgent tracheostomy in order to secure his airway. Biopsies were taken at the time of the procedure as well as by flexible bronchoscopy several days later. These revealed areas of acute inflammation and surface ulceration, as well as fragments of atypical cartilage, with enlarged chondrocytes, irregularly distributed on a basophilic matrix. The atypical chondrocytes demonstrated hypercellularity, nuclear hyperchromasia, and occasional binucleated forms (Figure 3); consistent with a diagnosis of laryngeal chondrosarcoma.

The patient subsequently underwent total laryngectomy, and all margins were clear of residual tumor. Review of the surgical specimen showed a well delineated posterior chondrosarcoma measuring 3.8 x 1.0 x 1.2 cm arising from the cricoid cartilage (Figure 4), and the examining pathologist noted that the “airway was narrow and rigid, barely allowing for the passage of the smallest finger.” After recovery from surgery, the patient is doing well with no evidence of residual disease.

DISCUSSION

Chondrosarcoma is a common tumor of bone, which very uncommonly presents in the larynx. While chondrosarcomas are the 3rd most common tumor of bone¹, and the most com-

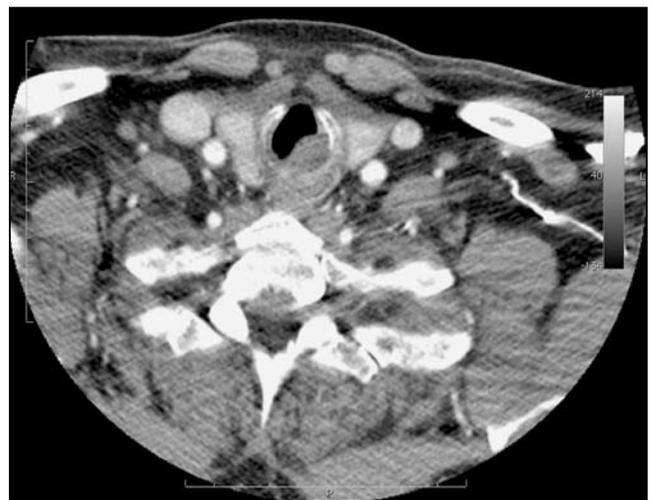


Figure 2. A soft tissue mass is seen surrounding the cricoid cartilage, narrowing the posterolateral trachea. The mass is smoothly delineated and lacks calcification.

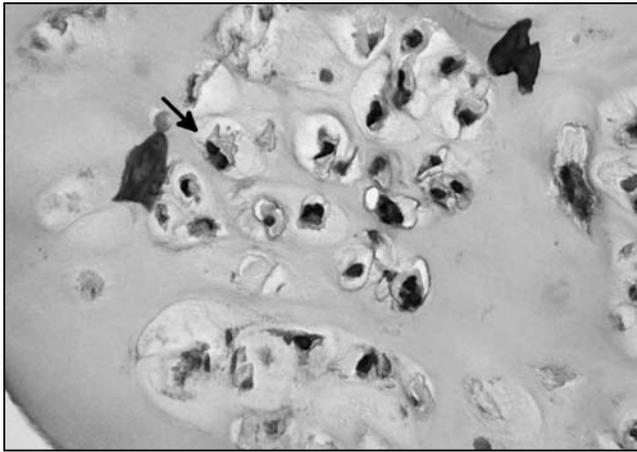


Figure 3. Hematoxylin and eosin stain of biopsy specimen showing atypical pleomorphic chondrocytes, hyperchromatic nuclei, and occasional pleomorphic forms (arrow). 400x magnification.

mon sarcoma diagnosed in patients over 50 years of age, they represent only about 0.5% of all laryngeal tumors,² although estimates of incidence are complicated by confusion with laryngeal chondromas, and certain other rare clinical entities. The pathogenesis of these tumors is not well understood although there has been speculation about the possibility that they arise from disordered ossification of laryngeal cartilages², or ischemic degeneration of pre-existing benign chondromas.²

The mean age of presentation of laryngeal chondrosarcoma is the mid-sixth decade of life; and although most series have found a male predominance, there does not appear to be any significant age difference at presentation between the genders². Most patients have symptoms attributable to vocal cord dysfunction or direct compression of the larynx such as hoarseness, dyspnea, and dysphagia. Similar to this case, almost all patients have a prolonged duration of symptoms prior to diagnosis, with a mean duration of 28 months in the largest published series².

Radiographic studies generally show mass lesions of mixed density with hypodense, isodense, and hyperdense areas compared to surrounding muscle. The mass is usually well-defined, with displacement, replacement and destruction of surrounding cartilaginous structures. Invasion of vascular structures is rare³. Reports describe a range of fine punctuate to stippled coarse (“popcorn”) calcifications, seen in 75-80% of reported cases⁴, although this feature was lacking in our patient.

Pathologically, most laryngeal chondrosarcomas arise from the cricoid cartilage, as in this case, although the other laryngeal cartilages can be also be involved. At time of resection the tumor is usually described as lobular, blue-gray, and “glistening”. Microscopically, chondrosarcomas are defined by loss of normal cartilaginous structure and distribution of chondrocytes in basophilic to metachromatic matrix⁵. Grading of chondrosarcomas is divided by degree of invasion, cell irregularity, and the presence of multinucleate cells, and nuclear hyperchromasia⁶.

Laryngeal chondrosarcomas are generally considered to be relatively slow growing and nonaggressive tumors, although their location can make management difficult. Conservative larynx-sparing surgery is usually attempted when possible, and conservative surgery does not generally negatively impact survival². Chemotherapy and radiation are not generally effective

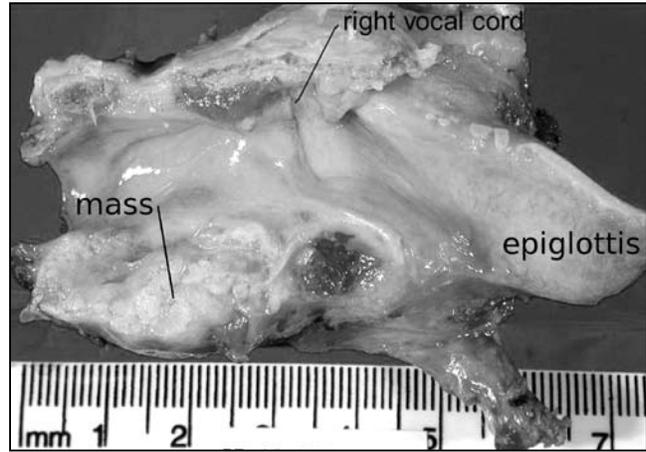


Figure 4. Resected larynx showing vocal cord and posterior mass, with glistening, grayish cut surface.

modalities, although case reports of primary or adjuvant radiation therapy exist⁷. The rate of metastatic disease varies by reported series between 2-10%, and death from laryngeal chondrosarcoma is rare with survival rates of 90% or greater².

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Edmund Sears, MD, is a Fellow in the Division of Pulmonary, Critical Care, and Sleep Medicine at the Warren Alpert Medical School of Brown University.

Matthew D. Jankowich, MD, is an Assistant Professor of Medicine in the Division of Pulmonary and Critical Care Medicine at the Warren Alpert Medical School of Brown University and staff physician at the Providence VA Medical Center.

Terrance T. Healey, MD, is an Assistant Professor of Diagnostic Radiology in the Department of Diagnostic Imaging at the Warren Alpert Medical School of Brown University, and Director of Thoracic Imaging at Rhode Island Hospital.

Disclosure of Financial Interests

CORRESPONDENCE

Edmund H. Sears, MD
Rhode Island Hospital
593 Eddy St.
Providence, RI, 02903
e-mail: esears@lifespan.org