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Radiological-Pathological Correlation

Dedifferentiated chondrosarcoma of the larynx: Radiological, gross, microscopic and clinical features

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ABSTRACT

Laryngeal chondrosarcoma is an uncommon malignancy with a predilection for the cricoid cartilage of adult male patients. Although rare, identification of aggressive chondrosarcoma variants, such as dedifferentiated chondrosarcoma (DDCS) may influence preoperative patient counseling, definitive surgical management, potential implementation of post-operative adjuvant therapy and prognosis. Herein we describe clinical and imaging features of laryngeal DDCS, the unique perspective of fresh and formalin fixed macroscopic examination, a spectrum of histopathologic findings, and detail the full course of the patient's disease.

1. Introduction

Sarcomas of the larynx account for < 1% of primary laryngeal neoplasms. Chondrosarcoma represents the most common laryngeal sarcoma and is often centered in the cricoid cartilage, exhibits a male predilection and in most cases, behaves in a low grade manner [1,2]. Risk of airway compromise and submucosal location of the neoplasm contribute to the technical challenges of providing a representative biopsy sample of laryngeal chondrosarcoma (LCS) [3]. Microscopically the classification of any rare neoplasm can be problematic, a challenge that may be compounded by sample fragmentation and/or low grade or bland histologic features of cartilaginous neoplasms. In the largest review of LCS, investigators found that incisional biopsy results were only explicitly documented in approximately 28% of cases, and not uncommonly the initial and final classification were discordant [2]. Herein, we describe a case in which the malignant spindle cell proliferation and atypical cartilage components captured on incisional biopsy sampling facilitated accurate diagnosis of laryngeal dedifferentiated chondrosarcoma (DDCS), followed by frank patient counseling and surgical treatment of this rare and aggressive tumor. In addition to the description of clinical and imaging features, this report highlights the uncommonly documented perspective of a fresh macroscopic examination, the subsequent formalin fixed gross examination, histopathologic findings, treatment and the full course of the patient's disease.

1.1. Case

A 76 year-old female presented to a community otolaryngologist with a six month history of increasing dysphonia. There was no history of radiation therapy to the head and neck. Initial examination demonstrated no concerning laryngeal pathology and appropriate vocal fold movement. The patient was started on an anti-reflux diet and noted no improvement of her symptoms. Increased shortness of breath and stridor prompted subsequent flexible laryngoscopic examination 10 months later, demonstrating a polypoid posterior subglottic mass. A computed tomography (CT) scan of the neck with intravenous contrast demonstrated a bulky subglottic laryngeal mass which appeared intrinsic to the cricoid cartilage. Internal chondroid ring and arc calcifications within the expanded cricoid cartilage narrowed the subglottic airway. More inferiorly, an exophytic soft tissue component projected posteriorly, more in keeping with dedifferentiated tumor (Fig. 1).

The patient was referred to our institution for further evaluation and although originally scheduled for an outpatient direct laryngoscopy with biopsy, presented emergently within the week with worsening dyspnea and stridor. Awake tracheotomy, direct laryngoscopy, and biopsy demonstrated a spindle cell malignancy in addition to fragments of atypical cartilage. The combined histologic findings, when taken together with the radiographic impression of chondrosarcoma favored a diagnosis of DDCS.

A positron emission tomography–CT (PET-CT) scan performed to assess for regional and distant metastasis demonstrated a fludeoxyglucose (FDG)-avid lesion centered within the cricoid, but no evidence

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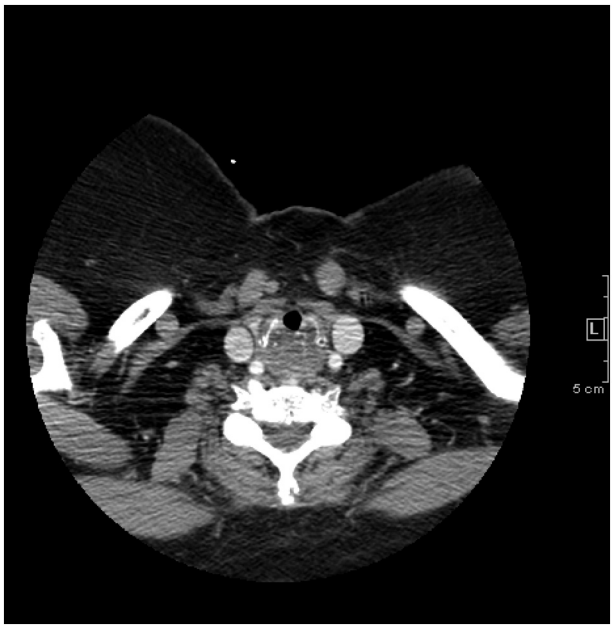

Fig. 1. Computed tomography with contrast, axial view demonstrating the subglottic laryngeal mass within the cricoid cartilage and an exophytic soft tissue component projecting posteriorly, consistent with dedifferentiated tumor.

of regional or distant metastasis. The patient underwent a total laryngectomy without elective neck dissection and primary tracheoesophageal prosthesis (TEP) placement, concordant with the recommendations of the multidisciplinary tumor board. The mass was centered within the cricoid cartilage during the surgery; however, there was a pedunculated portion that extended inferiorly to the level of the tracheotomy. This pedunculated portion was free from any attachment to mucosa or framework of the trachea, allowing for at least 6 mm of clinically uninvolved margin at the tracheal incision.

1.2. Macroscopic findings

Examination of the external surface of the fresh total laryngectomy (TL) specimen revealed abnormal expansion of the posterior laryngeal contours, and a smooth-surfaced red-brown mass obstructing the distal tracheal opening (Fig. 2). As noted during the surgical procedure, this portion of the mass was not attached to the distal tracheal framework, and light finger pressure could partially 'reduce' the mass into the trachea lumen. Opening the TL specimen posteriorly in the midline revealed an expansile tan-grey mass with a gelatinous, mucoid-type cut surface centered in the posterior cricoid cartilage with gross extension into adjacent structures. The irregular tan-grey cut surface of the cricoid lesion abruptly transitioned to a smooth, soft red-brown elongated segment protruding into the airway with a polypoid distal extension, previously identified as the mass occluding the distal tracheal airway (Fig. 3a and b). On subsequent gross examination of the formalin-fixed specimen, additional sections through the cricoid mass revealed classic grey-white lobules of chondroid matrix with near-circumferential cricoid cartilage involvement (Fig. 4a–b). Additional sections through the contiguous formalin-fixed polypoid component now revealed a tan-white homogenous mass infiltrating into the soft tissue and cartilaginous framework of the larynx, to involve the intrinsic laryngeal musculature (Fig. 4c–d).

1.3. Histopathologic evaluation

Sections submitted from the cricoid cartilage mass revealed hypercellular, myxochondroid matrix with increased cytologic and nuclear atypia, in keeping with grade II chondrosarcoma (Fig. 5a–c). Areas

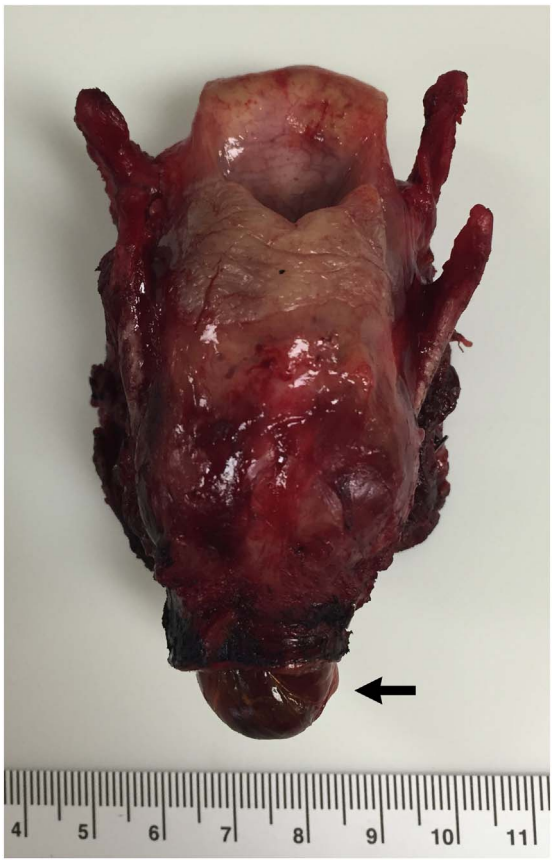

Fig. 2. Examination of the posterior external surface of the fresh total laryngectomy (TL) specimen revealed abnormal expansion of the posterior laryngeal contours, and a smooth-surfaced red-brown mass obstructing the distal tracheal opening (arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

of mineralization distinct from pre-existing ossified cricoid cartilage were focally present within the cartilaginous tumor. Sections of the adjacent polypoid mass showed a relatively abrupt transition from chondrosarcoma to a cellular, non-cartilaginous high grade sarcoma (Fig. 5d). This malignant spindle to epithelioid cell neoplasm, the 'dedifferentiated' component, showed vesicular, pleomorphic nuclei, prominent nucleoli, high mitotic rate and a focal proliferation of giant cells (Fig. 5d–e). Cytokeratin AE1/3 was focally positive in the spindled to epithelioid sarcoma element (Fig. 5f) and negative for high molecular weight cytokeratin and p63. Overlying mucosal dysplasia was absent. The initial diagnosis of DDCS was confirmed, and all surgical margins of the resection were negative for neoplasm. The non-chondroid sarcoma represented approximately 45% of the total laryngeal mass by combined gross and microscopic examination.

1.4. Follow-up

The patient began radiation treatment 6 weeks post-operatively and continued for seven weeks. She was treated to 70 Gray for the area of pre-operative gross disease and to 60.2 Gray for the entire surgical bed without delays or breaks in treatment. The patient's three-month post-treatment PET-CT scan demonstrated widespread soft tissue and bony metastasis (Fig. 6).

The patient was started on palliative chemotherapy after her PET-CT scan and ultimately succumbed to disease eight months after her total laryngectomy was performed.