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Chondrosarcoma of the Larynx: CT Features

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PURPOSE: To define the typical CT features of chondrosarcoma of the larynx. PATIENTS AND METHODS: Results of CT studies, performed on 10 patients with pathologically proved chondrosarcoma of the larynx, were retrospectively reviewed and correlated with clinical presentation. RESULTS: In all patients, the mass was detected on CT. The most frequent site of origin of the tumor was the cricoid cartilage (nine cases) followed by the thyroid cartilage (one case). Coarse or stippled calcification within the tumor was the most helpful radiologic finding and was seen in every case. In eight patients, the tumor had both an endolaryngeal and an extralaryngeal growth pattern, whereas in two patients the tumor was entirely endolaryngeal. Hoarseness, dyspnea, and dysphagia were the most common symptoms. In all patients presenting with dyspnea, the tumor exhibited endolaryngeal components. In patients presenting with hoarseness, three tumors had endolaryngeal and extralaryngeal components and two tumors were entirely endolaryngeal. CONCLUSION: Cross-sectional imaging afforded excellent evaluation of the airway as well as the extralaryngeal component of the tumor.

Index terms: Larynx, computed tomography; Larynx, neoplasms; Chondrosarcoma

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Chondrosarcomas of the larynx are rare, slowly growing, malignant neoplasms of cartilage tissue origin that can often be successfully controlled with local excision (1–6). Although these tumors have been amply documented in the pathology literature (4–14), radiologic correlation has been scant and primarily limited to plain films, barium esophagrams, and tomography (4, 7, 9, 15). The few published studies involving newer radiologic methods are limited to case reports (12, 14, 16, 17). We retrospectively reviewed the computed tomography (CT) examinations in 10 patients with chondrosarcoma of the larynx in an effort to define the typical CT features of this lesion.

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Materials and Methods

From 1970 to 1991, 98 cases of pathologically proved chondrosarcoma of the larynx have been registered into the archives of the Armed Forces Institute of Pathology (AFIP); CT scans were available for review in six; four additional cases with CT examinations were obtained from the teaching archives of the Mallinckrodt Institute of Radiology. The diagnoses of all 10 cases were confirmed by biopsy.

Clinical information available included age at presentation, sex, history, duration of symptoms, operative findings, and cartilage of origin. Imaging studies were evaluated for evidence of mass, CT density of the lesion, presence and pattern of calcification, definition of lesion margin, and pattern of growth. The density of the mass was rated as hypodense, isodense, or hyperdense compared with adjacent muscle. Fine, punctate calcifications were termed stippled and large collections of irregular calcifications were termed coarse. The overall appearance of a lesion was recorded as being predominantly solid or cystic. Margins of the mass that were clearly separable from adjacent tissue for at least 50% of the tumor circumference were considered well defined; otherwise, the margins were considered ill defined. Pattern of growth was judged as endolaryngeal if the tumor was confined by the outer margin of the cartilage of origin and grew primarily inwardly. An extralaryngeal pattern of tumor spread was defined as growth that had extended beyond the outer circumference of the cartilage into the paralaryngeal tissues.

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The relationship of the lesion to adjacent structures such as the carotid arteries and pharynx was evaluated for displacement or invasion. Evidence of airway compromise and presence and location of adenopathy also was recorded.

The CT imaging equipment and protocols used varied according to the time and place in which the study was performed.

Results

The results are summarized in Table 1. The patients, nine men and one woman, ranged in age from 60 to 79 years (mean, 70.1 years). The most frequent symptoms were dyspnea (six patients), hoarseness (five patients), and dysphagia (two patients). Two patients experienced stridor and one patient complained of a neck mass. Symptoms persisted for days to 10 years (mean, 27 months).

The most frequent site of origin of the tumor was the cricoid cartilage (nine patients) (Figs. 1–3) followed by the thyroid cartilage (one patient) (Fig. 4). The terminology of the surgical pathology diagnoses varied and included: chondrosarcoma (one patient); chondrosarcoma, low grade (four patients); chondrosarcoma, grade I (one patient); chondrosarcoma, low grade II (one patient); chondrosarcoma, well differentiated (two patients); and malignant chondrosarcoma (one patient) (Table 1). Nine patients eventually underwent laryngectomy.

CT showed a soft-tissue mass in all cases. Four lesions had mixed CT densities with hypodense, isodense, and hyperdense components. Three lesions were isodense and hyperdense. Two lesions were only hyperdense. One lesion was hypodense and hyperdense. This latter lesion was predominantly cystic (Fig. 4); the remaining nine lesions were solid. Calcification was demonstrated in every case. The pattern of calcification was coarse (Fig. 3) in three patients, stippled (Fig. 1) in two patients, and a combination of both stippled and coarse in five patients. Tumor margins were well defined in six patients and ill defined in four patients. Eight patients had CT evidence of both endolaryngeal and extralaryngeal growth of the tumor. In two patients, the tumor was confined to the cartilage of origin (Fig. 2).

Tumor either displaced (four patients) or did not involve (six patients) the carotid arteries; no tumor invaded a carotid artery. The pharynx was displaced in one patient and unaffected in another patient. Because of poor tissue contrast, the pharynx in each of the remaining eight patients could not be evaluated. The vocal cords were displaced in all patients. The airway was displaced anteriorly in nine patients and was circumferentially narrowed in one patient. Of the six patients who underwent complete CT surveys of the neck, none demonstrated adenopathy.

Discussion

Nonepithelial neoplasms arising from the supporting tissues of the larynx are rare and account for only 2% of primary laryngeal neoplasms (1). Within this group, cartilaginous lesions have been well documented in the pathology literature (3–14, 18, 19). The two largest series include reviews of 22 cases by Goethals et al (9) and 39 cases by Hyams et al (10). The latter experience was culled from the AFIP from 1929 to 1969. Neither of these large series or the other isolated reports emphasized the radiologic presentation of these tumors.

Approximately 70% of cartilage tumors in the larynx are chondrosarcomas (4, 8–12). Criteria for pathologic diagnosis of chondrosarcoma include the presence of many cells with large, irregular and/or multiple nuclei, and giant cartilage cells with large single or multiple nuclei and nuclei containing clumped chromatin (20). Chondrosarcomas usually occur in the fourth to sixth decades (2) and have a reported male to female predominance ranging from 5:1 (1) to as high as 10:1 (15). Our findings were consistent with these earlier series and showed an age range from 60 to 79 years (mean, 70.1 years); men predominated 9:1.

Chondrosarcomas of the larynx typically originate in hyaline cartilage. The cricoid cartilage is involved in approximately 70% of cases (11). The posterior lamina is the most frequent site within this cartilage (10). The thyroid cartilage is the next most common site of origin. In the original AFIP series, 50% of the chondrosarcomas arising in the thyroid cartilage originated on the external surface of the thyroid lamina; no tumors arose from the epiglottis, corniculate, cuneiform, or triticea cartilages (10). Huizenga et al (4) reported an additional three cases of chondrosarcoma arising from the arytenoid cartilage. Scattered reports of chondrosarcoma arising in juxta laryngeal structures such as the hyoid bone and tracheobronchial tree have also been reported (21, 22). In our series, nine tumors arose from the cricoid cartilage and one from the thyroid cartilage; these findings are in agreement with previous reports.

TABLE 1: CT findings in chondrosarcoma of the larynx

Case No.	Clinical Data							
	Age (years)	Sex	History	Duration of Symptoms	Location	Surgical Pathology	Radiology Mass	
1	68	Μ	Dyspnea Dysphagia	10 years 10 years	Cricoid	Chondrosarcoma, grade I	+	
2	68	M	Unavailable	Unavailable	Cricoid	Chondrosarcoma, low grade	+	
3	60	M	Dyspnea	2 years	Cricoid	Chondrosarcoma, low grade		
4	70	Μ	Hoarseness Dysphagia	4 years Unavailable	Cricoid	Chondrosarcoma, low grade	+	
5	77	M	Neck mass	Weeks	Thyroid	Chondrosarcoma, low grade (II)	+	
6	63	Μ	Dyspnea Hoarseness	8 months 8 months	Cricoid	Chondrosarcoma, well differentiated	+	
7	66	M	Hoarseness	1 year	Cricoid	Malignant chondrosarcoma	+	
8	79	Μ	Dyspnea Hoarseness	5 months 5 days	Cricoid	Chondrosarcoma	+	
9	75	F	Hoarseness Dyspnea Stridor	2 years 5 months 2 years	Cricoid	Chondrosarcoma, well differentiated	+	
10	75	Μ	Stridor Dyspnea	3 months 3 months	Cricoid	Chondrosarcoma, low grade	+	

	Radiology								
Case No.		CT Density		Calcification	Predominate Appearance	Margins	Pattern of Growth		
	Hypodense	Isodense	Hyperdense						
1	+	+	+	+, Stippled and coarse	Solid	III defined	Endo, exo		
2		+	+	+, Stippled	Solid	III defined	Endo, exo		
3			+	+, Coarse	Solid	Well defined	Endo, exo		
4	+	+	+	+, Stippled	Solid	III defined	Endo, exo		
5	+		+	+, Stippled and coarse	Cystic	Well defined	Endo, exo		
6			+	+, Coarse	Solid	Well defined	Endo, exo		
7	+	+	+	+, Coarse	Solid	Well defined	Endo		
8	+	+	+	+, Stippled and coarse	Solid	III defined	Endo, exo		
9		+	+	+, Stippled and coarse	Solid	Well defined	Endo		
10		+	+	+, Stippled and coarse	Solid	Well defined	Endo, exo		

C	Radiology							
Case No.	Carotid Arteries	Pharynx	Vocal Cords	Airway Displacement/ Obstruction	Adenopathy			
1	Unaffected	*	Displaced	+, Anterior	-			
2	Unaffected	*	Displaced	+, Anterior	_			
3	Unaffected	*	Displaced	+, Anterior	-			
4	Displaced	*	Displaced	+, Anterior	_			
5	Displaced	*	Displaced	+, Anterior	_			
6	Unaffected	*	Displaced	+, Circumferential obstruction	*			
7	Unaffected	Unaffected	*	+, Anterior	-			
8	Unaffected	*	Displaced	+, Anterior	*			
9	Displaced	Displaced	*	+, Anterior	*			
10	Displaced	*	Displaced	+, Anterior	*			

Note.—Abbreviations: M, male; F, female; endo, endolaryngeal; exo, extralaryngeal; +, present; -, not present; *, unable to evaluate.

The symptoms of cartilage tumors depend upon the location of the mass. Endolaryngeal and subglottic growth causes dyspnea as the airway is progressively obstructed; whereas extralaryngeal growth, originating in the posterior cricoid, usually produces dysphagia (3, 8). Limited laryngeal mobility causes hoarseness (8). This is more likely due to restriction of the vocal cords by the mass, rather than from paralysis of the recurrent laryngeal nerve (15). One reported indolent chondrosarcoma of the cricoarytenoid region caused a previously misdiagnosed vocal cord paralysis



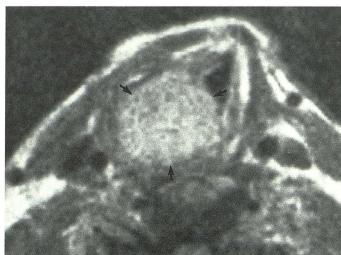


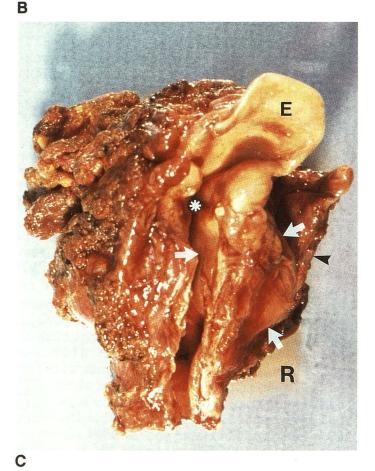
Fig. 1. Case 4.

A

A, Axial contrast CT scan demonstrates stippled calcification (arrowheads) within a large mass (small arrows) arising from the cricoid cartilage and extending into the extralaryngeal tissues. The thyroid cartilage (large arrows) is displaced anteriorly and to the left.

B, Axial proton density-weighted MR scan (1500/35) (TR/ TE) shows the margins of the tumor (arrows) but fails to demonstrate the calcifications.

C, Gross surgical specimen observed posteriorly with the patient's right side (R) oriented to the reader's right side. The large subglottic tumor (arrows) is submucosal and is covered with intact mucosa. Epiglottis (E), laryngeal ventricle (asterisk), right thyroid cartilage lamina (arrowhead).



(14). Tumors involving the thyroid cartilage are more likely to produce a painless neck mass (10).

In our small series, tumor location and symptoms correlated poorly. This may have been related to the subjective nature of the complaints and the abbreviated histories available in some patients. In the six patients with dyspnea, CT demonstrated a significant endolaryngeal component of tumor growth with resultant narrowing and displacement of the airway. In the three patients without dyspnea, however, the airway was similarly displaced.

In three of the patients with hoarseness, the true vocal cords had CT evidence of displacement by tumor. The cords were displaced in four other patients who did not present with hoarseness.

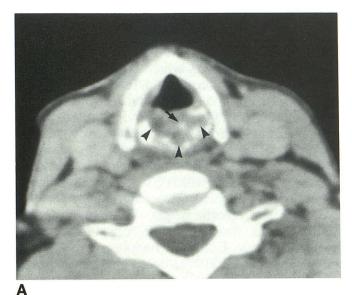




Fig. 2. Case 7: axial CT scans demonstrating primarily endolaryngeal pattern of growth in a chondrosarcoma arising from the cricoid cartilage.

- A, The mass (arrowheads) contains coarse calcifications at this level (arrow).
 - B, The subglottic airway (arrows) is narrowed.

(Illustration from Glazer HS, Balfe DM, Sagel SS. Neck. In: Lee JKT, Sagel SS, Stanley RJ, eds. *Computed body tomography with MRI correlation*. New York: Raven Press, 1989:130. Reprinted with permission.)

Eight patients had extralaryngeal growth of tumor but only two had dysphagia. In the patients with dysphagia, tumor had extended posteriorly or posterolaterally. In one of these patients, a barium esophagram demonstrated displacement of the pharynx. The single patient whose tumor originated in the thyroid cartilage complained of a neck mass.

Because of the indolent behavior of this tumor, symptoms may persist for months (3, 4). In our series, symptoms persisted for days to 10 years (mean, 27 months).

One of the most helpful radiologic signs suggesting chondrosarcoma is the coarse or stippled calcification within the mass (23) which is felt by several authors to be pathognomonic when present (11, 15). Early series that contained plain films analysis reported the occurrence of calcification from 40% to as high as 80% (1, 2, 9, 11, 12, 15). In our series, calcification was identified in every patient. The pattern of calcification was stippled, coarse, or a combination of both, and varied from minimal to extensive.

Because of improved tissue contrast resolution, one would expect CT to be more sensitive than plain radiography in detecting this calcification. Although magnetic resonance (MR) imaging may superbly demonstrate tumor extent, it cannot identify the calcific matrix as accurately. Such was the case in one of our patients who was examined with both MR and CT (Fig. 1).

Eliminating consideration of the characteristic calcification, the CT appearance of the tumor is otherwise nonspecific. In our series, lesions tended to be solid and mixed in density. One lesion was notably cystic, probably due to necrosis; however, no correlation with histologic grade was apparent. CT evaluation of extralaryngeal extent of tumor is superior to plain films but can be difficult when isodense portions of the tumor blend imperceptibly with the surrounding tissues.

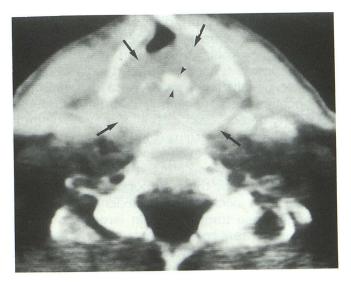


Fig. 3. Case 10. axial CT showing large chondrosarcoma (arrows) arising from the cricoid cartilage. Coarse calcification (arrowheads) is seen within the tumor. Stippled calcification was evident on adjacent slices (not shown).

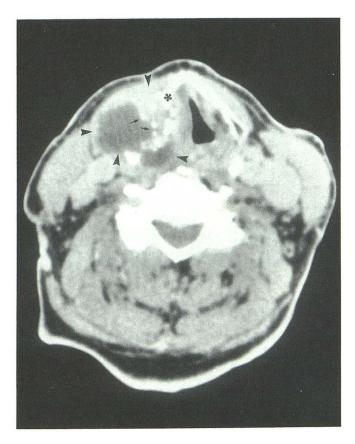


Fig. 4. Case 5: axial CT showing a large chondrosarcoma (arrowheads) arising from the right thyroid cartilage lamina (asterisk). Coarse calcifications (arrows) permeate the tumor at this level.

Detection of extralaryngeal spread of tumor was aided by identification of displaced structures such as the carotid arteries. Evaluation of the pharynx was frequently difficult because of the lack of tissue contrast with the adjacent tumor; oral contrast would have been helpful. Well-defined tumors tended to have hypodense, hyperdense, or calcified margins, or to have margins encroaching the laryngeal air column. Although MR poorly detects calcification, it may be helpful in defining tumor margins (Fig. 1).

CT was helpful in evaluating the carotid arteries. No invasion was detected reflecting the indolent behavior of these tumors. CT effectively evaluates the neck for lymphadenopathy (24). Although these tumors remain largely confined to the larynx, rare metastases to soft tissues and lungs have been reported (4, 10). Of our six patients whose studies included surveys of the neck, no adenopathy was detected. Routine complete neck scans should be included as part of the evaluation for local extension and nodal disease.

Because many authors advocate primary excision for control of laryngeal chondrosarcoma (3-6), preoperative CT is essential for defining extralaryngeal disease, which could escape clinical detection. A complete differential diagnosis for chondrosarcoma of the larynx is quite lengthy but the practical differential is more limited. Squamous cell carcinoma should always be considered for any bulky mass in the larynx. Most squamous cell carcinomas arise above the level of the cricoid cartilage. In cases in which CT detects a subglottic mass, more cephalad scans will usually show a glottic or supraglottic origin. Moreover, calcified tumor matrix should prompt consideration of a cartilage tumor. Epithelial neoplasms rarely calcify, with one exception being the pleomorphic carcinoma, which can contain osteoid, cartilage, and bone matrix (1). This latter tumor is quite rare, however,

The differential diagnosis of chondrosarcoma should also include benign chondroma. Although hypercellularity may be evident, the cells remain histologically similar to normal cartilage on photomicroscopy (2). Chondromas can arise from either hyaline or elastic cartilage as well as other soft tissues of the neck (10). Chondrosarcoma, however, originates in the hyaline cartilage. Chondrosarcoma usually presents after the fifth or sixth decade of life, whereas chondroma usually presents decades earlier (10).

The radiologic distinction between chondrosar-coma and chondroma is extremely difficult if not impossible (3, 8, 11, 18). One of the values in radiologic diagnosis is more appropriately to separate chondroid lesions, which contain characteristic calcification, from noncalcified tumors. Lavertu concluded that the pretreatment distinction between low-grade chondrosarcoma and chondroma was of minimal value since the outcome of tumors initially identified as chondroma did not differ from those labeled chondrosarcoma prior to treatment (5). Furthermore, patients with recurrence of previously treated chondroma often have evidence of chondrosarcoma in the original specimens upon review (5).

Less common than chondroma and squamous cell carcinoma are a long list of rare conditions, including fibrosarcoma, rhabdomyosarcoma, osteosarcoma, rhabdomyoma, granular cell myoblastoma, nerve sheath tumor, cylindroma, hemangioma, lipoma, and nonneoplastic cyst (1, 11). Although these lesions can be grouped by clinical findings, the radiologic manifestations can be extremely nonspecific. Cross-sectional imag-

ing can establish the cartilage of origin in most cases. Calcification is again a helpful CT finding because it is usually not present to significant degree in these other lesions. Although fibrosarcoma can arise from the cricoid region, it is usually found in the anterior commissure and vocal cords (1, 13). Rhabdomyosarcoma usually develops by the fourth decade (13). Osteosarcoma could potentially present with an osteoid matrix, but this tumor is so rare that only a few case reports document its existence (25, 26). Other sarcomas, we well as neurogenic tumors and cylindromas, tend to occur in the supraglottic rather than infraglottic region, a distinguishing feature (1).

Hemangiomas can be either the adult or infantile type. Although the infantile type can present with a bulky mass, the compressible character of the mass and age at presentation virtually eliminate chondrosarcoma as a reasonable possibility. The adult-type hemangioma is usually small, nodular, reddish in color, and located in the glottic or supraglottic region (1). Lipomas can be distinguished because of the fat attenuation values on CT.

In summary, the diagnosis of chondrosarcoma of the larynx is established by histologic review of biopsy material obtained from evaluation of a slowly growing mass. CT can suggest the diagnosis prior to biopsy and can direct the biopsy site, particularly since small specimens can be difficult to interpret and may show a sampling error. Radiologic evaluation is essential to define the extent of the lesion since surgery can be curative in these slowly growing tumors, to assess the regional spread which, although rare, can escape detection on physical examination, and to monitor for local recurrence in those patients who have had conservative therapy or who have carried the histologic diagnosis of chondroma.

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