Chondrosarcoma of the Larynx

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ABSTRACT: **Background:** Chondrosarcoma of the larynx is a rare tumor. The most common symptom is hoarseness. Treatment is controversial.

Objectives: To describe six patients with laryngeal chondrosarcoma from a single center.

Methods: The medical records of a major tertiary hospital were reviewed for all patients with laryngeal chondrosarcoma diagnosed and treated from 1959 to 2010. Data on background, clinical treatment and outcome were collected.

Results: Six patients, all males with a mean age of 53.3 years, were identified. Partial laryngectomy was performed in three patients, and total laryngectomy, local excision, and partial cricoidectomy in one patient each. Four patients had a permanent tracheostomy after surgery. One patient required postoperative chemotherapy and one radiotherapy. Follow-up time was 12–216 months (mean 102 months). Recurrence developed in two patients 2 and 8 years after initial treatment and was treated by salvage surgery in both patients. One patient died during the follow-up from an unrelated cause. The others are currently alive.

Conclusions: This study supports earlier reports recommending initial treatment with partial or total laryngectomy for laryngeal chondrosarcoma. Long-term follow-up for recurrence is advised. We recommend preserving the larynx, if possible, even if a permanent tracheostomy is necessary.

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KEY WORDS: chondrosarcoma, larynx, recurrence, treatment, resection

Chondrosarcoma of the larynx is an uncommon tumor, accounting for approximately 1% of all laryngeal neoplasms [1]. It is, however, the most common non-epithelial neoplasm of the larynx [2]. It was first described by Travers in 1816 [3], but the term chondrosarcoma at this site was introduced by New in 1935 [4]. Most of the reports on laryngeal chondrosarcoma to date have been limited in size [5-10]; the largest was published by Thompson and Gannon in 2002 [1] and included 111 patients. The literature contains a number of case series, but a review showed that many describe the same patients from the same database(s) [1,11,12].

The etiology of laryngeal chondrosarcoma is unknown, although it is usually assumed to derive from disordered ossification of the laryngeal cartilage [5,13]. No relationship to tobacco use or alcohol consumption has been proved [1].

The mean age at diagnosis is 60 to 64 years, with a male predominance [2,14]. Symptoms vary, depending on the location of the mass, and include hoarseness (most patients) caused by narrowing of the glottic plane and compression of the inferior laryngeal nerves; dyspnea and airway obstruction due to endolaryngeal and subglottic growth; dysphagia due to extralaryngeal growth, originating in the posterior cricoid; and painless neck mass due to tumor involvement of the thyroid cartilage (when present) [1,5,13].

The tumor arises from hyaline – and not elastic – cartilage: cricoid cartilage in 75% of patients, thyroid cartilage in 17%, arytenoid cartilage in 5%, and epiglottis and accessory cartilages in 3% [13]. Chondrosarcoma is a macroscopically smooth and lobular mass. Microscopically it has high nuclear-to-cytoplasmic ratio and hyperchromatic nuclei. Mitotic figures are not common [1].

The histological diagnosis is based on the criteria for cartilaginous malignancies reported by Lichtenstein and Jaffe in 1943 [15]. There are three different grades of chondrosarcoma:

- **Grade 1** – similar to a chondroma, more than two nuclei, no mitoses, some areas of calcification and actual bone tissue (70–80% of cases).
- **Grade 2** – increase in cell number, low nuclear/cytoplasmic ratio, scarce mitoses (10–15% of cases).
- **Grade 3** – multinucleated cells, increased nuclear/cytoplasmic ratio, high number of mitoses (5–10% of cases) [16] [Figure 1].

About 80% of laryngeal chondrosarcomas are low grade compared to chondrosarcoma at the more common sites, namely pelvis, long bones, sternum, and ribs; those located on the larynx are usually well differentiated with a less aggressive pattern [17].

Imaging studies have some diagnostic value, although it is

Figure 1. Well-to-moderately differentiated chondrosarcoma of the larynx, and elastic-rich cartilage nodules with invasive component

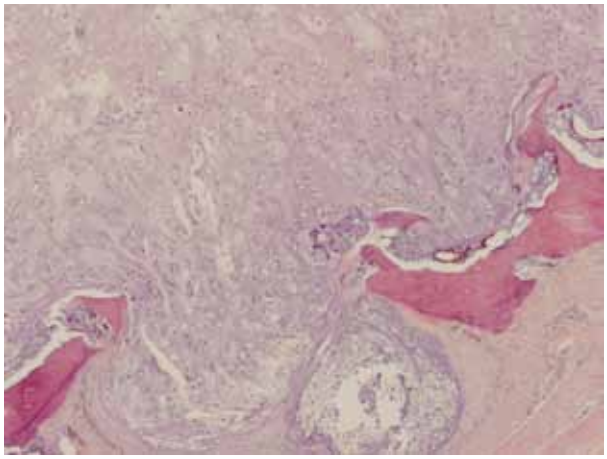
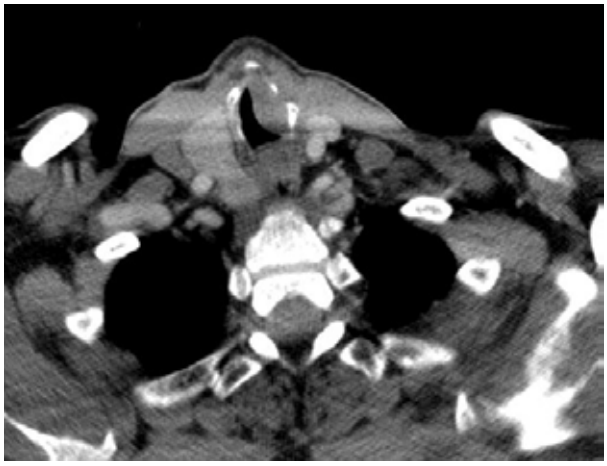


Figure 2. Chondrosarcoma of the larynx. Note the mass in the cricoid cartilage



impossible to distinguish chondromas from chondrosarcomas. The imaging modality of choice is computed tomography [18]. Typical findings are a hypodense, well-circumscribed mass containing mottled calcifications, with displaced structures and destruction of the cartilage [Figure 2]. Magnetic resonance imaging has the added advantage of superior contrast resolution of the tumor and paralaryngeal tissues [19]. On X-ray the tumor appears as a popcorn-like calcification [1].

The initial treatment for chondrosarcoma of the larynx is surgery [16] in order to excise all tumor extensions with negative margins [20]. For low-grade tumors, options include CO_2 laser resection, hemicricoidectomy, or hemilaryngectomy. For high-grade tumors and recurrences, the only choice is total laryngectomy. Attempts are made to spare the larynx for preservation of voice and normal swallow. When cricoidectomy is performed, laryngotracheal anastomosis or reconstruction by free flap is used to close the defect. The effectiveness of radiotherapy is still controversial. Chemotherapy is not considered an option [21].

Recurrence is common, with rates of 35-40%. Nevertheless, the long-term prognosis of laryngeal chondrosarcoma is good (95%, 10 year survival) and metastasis is rare (up to 10%) [1].

The aim of this study was to describe our experience with six patients diagnosed and treated for chondrosarcoma of the larynx at a major tertiary center in the last 51 years.

PATIENTS AND METHODS

Review of the medical records and computerized database of Rabin Medical Center from 1959 to 2010 yielded six patients diagnosed and treated for chondrosarcoma of the larynx. Data were collected on background characteristics, clinical analysis, treatment and outcome.

RESULTS

The results are shown in Table I. All six patients were male. The mean age at diagnosis was 53.3 years (range 24–66 years). Five patients presented with hoarseness and dyspnea; in the sixth patient, laryngeal chondrosarcoma was found incidentally on cervical CT performed for an unrelated reason. Histological study showed a well-differentiated tumor in one patient, well-to-moderately differentiated tumor in four, and a poorly differentiated tumor in one.

Surgery was performed in all patients as follows: partial laryngectomy in three, and total laryngectomy, endolaryngeal laser excision and cricoidectomy in one each. The patients treated with cricoidectomy and partial laryngectomy also had a permanent tracheostomy. Four patients were referred for additional therapy (radiotherapy/chemotherapy) because of recurrence, positive margins, or advanced-stage disease.

As of the end of 2011 the mean duration of follow-up was 102 months (range 12–216 months). The tumor recurred during follow-up in two patients, after 2 years and 8 years, and both underwent a second surgical procedure. No recurrence has been diagnosed in other patients or in those patients after the second surgical procedure. Five patients are currently alive. One patient died during follow-up from a cause unrelated to the chondrosarcoma.

DISCUSSION

This is our single-center experience. In the present series of six patients with chondrosarcoma of the larynx, the low-grade histologic findings in five cases prompted treatment with surgery with preservation of the larynx; only one

Table 1. Clinical and treatment characteristics of six patients with chondrosarcoma of the larynx

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6
Year of diagnosis	1959	1993	1997	2007	2008	2010
Age (yrs) Gender	64 M	56 M	24 M	57 M	66 M	53 M
Presenting symptom	Hoarseness, dyspnea	Incidental finding of neck mass on CT	Hoarseness, dyspnea	Neck mass, dyspnea	Hoarseness, dyspnea	Hoarseness, sensation of foreign body
Histopathology	Well to mod. diff. chondrosarcoma	Well to mod. diff. chondrosarcoma	Poorly diff. chondrosarcoma	Well to mod. diff. chondrosarcoma + papillary ca. of thyroid, 0.4 cm	Well to mod. diff. chondrosarcoma	Well-diff. chondrosarcoma
Treatment	Total laryngectomy + rt. hemi-thyroidectomy	Partial laryngectomy + permanent tracheostomy	Laser endolaryngeal subtotal laryngectomy	Lt. hemi-thyroidectomy + resection of cricoidal mass + permanent tracheostomy	Cricoidectomy + permanent tracheostomy	Partial laryngectomy + tracheostomy + reconstruction with buccal mucosa
Additional therapy	None	None	Chemotherapy	None	Radiotherapy	Radiotherapy
Follow-up (yrs)	12	18	14	4	2	1
Recurrence	None	None	At 8 yrs	At 2 yrs	None	None
Treatment for recurrence	–	–	Open resection of cervical mass	Partial laryngectomy + completion thyroidectomy + lt. lateral neck dissection	–	–
Additional therapy	–	–	Chemo-radiotherapy	Radiotherapy	–	–

patient underwent initial total laryngectomy. Four of the five patients treated with partial surgery had positive margins. Two of them failed treatment, one in the larynx and one in the neck, and required postoperative chemo/radiotherapy. There was no evidence of a second failure during 1 to 4 years of follow-up. The four patients treated with cricoidectomy or partial laryngectomy remain dependent on tracheostomy for breathing; all have excellent speech. The patient after total laryngectomy is alive and well with no evidence of recurrence 10 years after treatment.

Our study confirms other observations in the literature that radical surgery is usually unnecessary for laryngeal chondrosarcoma because of the relatively benign course of the disease [1,16]. The role of postoperative radiotherapy is controversial. In our series, the disease recurred in the patient with positive margins treated with post-surgery radiation, placing the value of radiation in question. None of our patients had distant metastases, confirming the assumption that chemotherapy should not be part of the treatment regimen.

In conclusion, the good prognosis in our six patients with chondrosarcoma of the larynx serves as further justification for the use of minimal surgery provided that the whole tumor is excised. Given the importance of preserving the larynx to patients' quality of life, we suggest doing so even when a permanent tracheostomy is necessary. The only risk is recurrence, which is treated by a second surgery with or without additional oncological therapy. In our series, there were no metastases, and no patient died from the disease. We recommend long-term follow-up, as one recurrence in our series developed 8 years after the initial diagnosis. Larger studies are needed to standardize the therapeutic approach to chondrosarcoma of the larynx.

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