



Chondrosarcoma of the Larynx: Report of One Case and Review of the Literature

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Abstract

Laryngeal chondrosarcoma is a rare tumor whose treatment is essentially surgical. The prognosis generally favorable depends mainly on the histological grade. The objective of this report is to describe, from this case, clinical, radiological and histological features of laryngeal chondrosarcoma, and discuss its therapeutic management.

Keywords: Chondrosarcoma; Larynx; Laryngectomy

Introduction

The laryngeal chondrosarcoma is a rare histological type with less than 1% of laryngeal cancer. The pathogenesis is still unknown [1,2]. Diagnosis can be difficult, especially in histology. Surgery is the best treatment and may vary from an endoscopic surgery to partial surgery or total laryngectomy, depending on the extension and the histological grade of the cancer [3-5]. The prognosis is usually good especially for the low-grade chondrosarcoma [6,7]. This study reports a recent clinical case and analyzes the epidemiological, clinical, histological, radiological, and treatment of this rare location of chondrosarcoma.

Case Presentation

A 46-year-old North African man has this past 6 months dysphonia that was complicated by laryngeal dyspnea that required an emergency tracheotomy. Clinical examination found a patient in good general condition with Karnofsky Index 100%, well maintained tracheostomy cannula surrounded by normal skin absence of lymphadenopathy. A systemic examination was unremarkable. The flexible endoscopy showed a laryngeal sector obstructed by a large diffuse supraglottic process looks bumpy in places (Figure 1). Computed tomography showed a tumor process centered on the two arytenoid (Figure 2) which are unrecognizable with a dual chondroid and tissular component with a calcification. This process extends the vocal folds, ventricles and ventricular strips on both sides, noted the absence of cervical lymphadenopathy.

Direct Laryngoscopy showed a diffuse supraglottic neo formation, covered with a lining inflammatory. The exploration of the two floors glottic and sub glottic is impossible. Histological study of biopsy done at a low grade, well differentiated chondrosarcoma and classed T4N0M0. Figure 3 total laryngectomy without lymphadenectomy was planned. Histological study of the work piece Laryngectomy showed low grade chondrosarcoma invading the thyroid cartilage and the opposite muscles with healthy resection limits of the trachea and epiglottis.

The postoperative course was uncomplicated. At 13 months follow-up, there was no recurrence.

Discussion

The laryngeal chondrosarcoma is rare, with less than 1% of laryngeal cancer. But it is the most common mesenchymal cancers. Predilection site is the cricoid cartilage (75%) followed by the thyroid cartilage (17%), the arytenoid (5%) and epiglottis (2%) [8]. It usually occurs in patients between 50 and 70 years, mainly men.

The exact pathogenesis remains to be elucidated; some hypotheses attribute this condition to 3 local lesions, abnormal ossification, chronic inflammation and metabolic disorders related to old age [9]. Dysphonia is the main clinical sign. While dyspnea associated when the cancer increases in size, dysphagia is rarely present. A neck mass can also be revealing. In direct laryngoscopy, swelling submucosa and mucosa intact can be detected. Moreover, the first sign of the posterior cricoid involvement may be the stiffness of the vocal cords, due to the blockage of the cricothyroid articulation

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Figure 1: Endoscopic examination shown laryngeal sector obstructed by a large diffuse supraglottic covered by intact mucosa.

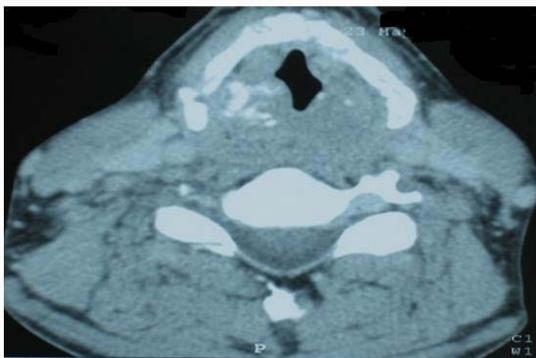


Figure 2: CT scan showed a tumor process centered on the two arytenoid which are unrecognizable with a dual chondroid and tissular component with a calcification.

arytenoidic and not because of the infiltration of the recurrent nerve [10,11]. The computer tomography is the examination of choice to analyze the nature, location and extension of cartilaginous tumors. These tumors, centered on cartilage, are well-defined contours, hypodense with moderate contrast enhancement. Calcifications characteristics are found in 80% of cases appearance like a "Popcorn". The CT scan also determines the degree of obstruction of the laryngeal sector and relations with adjacent structures of the tumor. MRI is less specific but to better clarify the extension and the relationship of the tumor to the surrounding structures [12].

Although the imaging may indicate the origin of the cartilaginous lesion, but does not distinguish the chondroma of low grade chondrosarcoma. Diagnosis of chondrosarcoma is given certainty by histological analysis. The histological appearance of the mesenchymal cancer was classified by Evans et al. [6] (1977) in different malignancy grades: a malignant form of low grade (first level) is characterized by chondrocytes in small, dense nuclei, binucleated cells, mitotic activity and low cellularity. The intermediate form and the high-grade malignant type are characterized by increased cellularity, many binucleated and multinucleated cells and numerous mitotic [6]. Surgery is the best treatment for these tumors, it must allow the eradication of cancer with sufficient safety margins and in the case of low-grade, must be functional and conservative [3,13]. Chondrosarcoma is considered less sensitive to radiotherapy. It may nevertheless be considered when surgery is impossible or if the excision is incomplete [7]. Most authors, however, agree that there is

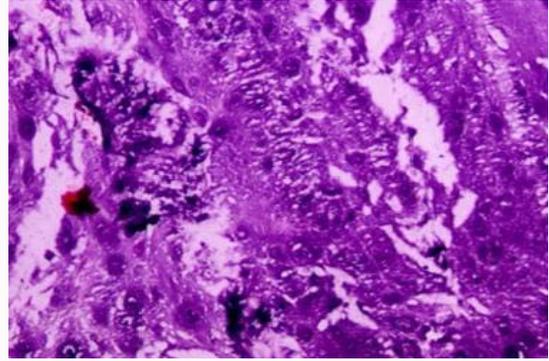


Figure 3: Histopathology showing lobular and patchily hypercellular proliferation of atypical chondrocytes. Magnification $\times 200$.

little evidence for postoperative adjuvant radiotherapy after complete resection [15]. Chemotherapy has no curative role [16]. The major prognostic factors are histologic grade, location, extent and quality of the initial resection. Local recurrence of chondrosarcoma is frequent. They have low metastatic potential. Lung and cervical lymph nodes are the most commonly affected sites [17].

Conclusion

Laryngeal chondrosarcoma is a rare tumor. Imaging-based scanner's role is to clarify the seat and tumor extension, it does not distinguish between chondroma and low-grade chondrosarcoma. This distinction remains as histological dilemma. The treatment is surgical. The prognosis is generally good, and basically depends on the histological grade.

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