CASE SERIES

CHONDROSARCOMA OF THE LARYNX – A SINGLE INSTITUTE EXPERIENCE

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Abstract

Chondrosarcoma is a rare tumor of the head and neck, accounting for approximately 0.1% of all head and neck malignancies and 1% to 2% of laryngeal neoplasm. A retrospective study of laryngeal chondrosarcoma cases in the past 20 years was done. Six patients diagnosed with laryngeal chondrosarcoma were included in the study. The clinical presentations, diagnostic work-up, management and outcome were presented. All patients were male, between 45-60 years old. All patients presented with voice changes, dyspnea later on stridor that required tracheostomy to secure airway. The site of origin was the cricoid cartilage in 3 cases, the thyroid cartilage in two cases and the arytenoid cartilage in one case. All patients underwent total laryngectomy depending on clinical presentation & size of lesion. Laryngeal chondrosarcoma is a slowly growing tumor that is rarely metastasizing to neck lymph nodes. Airway obstruction is the most presenting symptoms. Once the patient needs tracheostomy, a sort of laryngectomy Mostly, total laryngectomy is recommended. No chemo-radiotherapy is recommended for organ preservation protocol.

Keywords*: larynx, laryngeal neoplasms, chondrosarcoma*

Introduction

Chondrosarcoma is a rare tumor of the head and neck, accounting for approximately 0.1% of all head and neck malignancies and 1% to 2% of laryngeal neoplasm [1]. Although chondrosarcoma of the larynx (LCS) is a rare upper airway malignancy, with fewer than 600 cases described in literature, it represents the most common non-epithelial laryngeal tumor [2]. The etiology is unclear; however multiple theories were postulated such as initial disordered ossification of laryngeal cartilage [3], initial ischemic changes in a chondroma [4], radiation therapy [5] or after Teflon injection [6].

LCS often occurs in 6th and 7th decades with males more affected than females (3.6:1) [7]. Sites of predilection within the larynx are in the following order: cricoid is the most common site, then thyroid, arytenoid [8]. The most common presenting symptoms in a decreasing frequency, are hoarseness, dyspnea, dysphagia, dysphonia, cough, neck mass, stridor. The onset of symptoms is insidious given the indolent nature of this tumor [1,3].

Diagnostic work up includes laryngoscopy and imaging. CT-Scan is considered the imaging of choice, it shows a hypo dense mass with calcifications inside and cartilage destruction [7, 9]. Barium swallow may show a filling defect in the post cricoid region or the pyriform sinus [9]. Other authors prefer MRI due to its ability to distinguish between tumor and other para laryngeal tissue. The signal strength is low on T1

and high on T2, with a characteristic mosaic pattern [10].

Histopathological diagnosis can be done accurately through study of the surgical specimen. Diagnostic criteria include the presence hyaline cartilage with giant cartilage cells, hyperchromatic nuclei and pleomorphism [11]. LCS is classified into three grades; Grade I: accounts for 70-80% of cases and it is similar to a chondroma, >2 nuclei, no mitosis and some areas of calcification and actual bone tissue; Grade II is characterized by an increased cell number, a low nuclear/ cytoplasmic (N/C) ratio and few mitoses; and Grade III: characterized by multinucleated cells, an increased N/C ratio and a high number of mitoses [12].

Overall recurrence rate is between 35% and 40%. Recurrence is due to incomplete excision or high-grade tumor [7]. Metastasis occurs mostly in high grade tumors with estimated rate of 12%-15%. The most common sites of metastasis are lung followed by cervical neck nodes [1]. This Low rate of metastasis may be due to the indolent nature, the slow growth rate, and early diagnosis being made due to hoarseness and respiratory compromise [9].

Conservative resection is advocated due to the indolent nature of these tumors [1]; however radical surgery is indicated in the following situations: high-grade lesions, huge lesions with inability to preserve the larynx; an elderly or sick patient who might better profit from a single operation [9], recurrent tumors that cannot be salvaged by conservative resection [3].

LCS has good prognosis with ten- year survival rate of 95%. Prognostic factors include size and site of the tumor and the histologic grade [9]. Mortality from LCS is very rare and results from invasion of vital structures of the neck [7]. LCS has a potential for late recurrence and longer follow up and surveillance is mandatory for early detection of recurrence [3].

Materials and Method

After approval by the ethics committee of the hospital, review of LCS cases in the past 20 years was done. The review included the following parameters:

1. Demographic data
2. Site of origin and side
3. Presenting symptoms
4. Diagnostic work up
5. Neck status (N negative/ N positive)
6. Treatment modality
7. Histologic grade
8. Treatment outcome

Results

Six patients diagnosed with LCS were included in the study. To date it is the first and largest series published in the Arab world. All patients were male, between 45-60 years old. All patients presented with voice changes, dyspnea later on stridor that required tracheostomy to secure airway. Neck mass was also a presentation in tumors originated from thyroid cartilage, while dysphagia occurs in large lesions. The site of origin was the cricoid cartilage in 3 cases, the thyroid cartilage in two cases and the arytenoid cartilage in one case. Diagnosis made by biopsy in five cases, CT guided FNAC in one patient. CT scan was the best imaging modality aided in diagnosis of all cases. All patients underwent total laryngectomy depending on clinical presentation and size of lesion. Follow-up course revealed no recurrence in 5 cases while one case lost from follow up. Two cases are presented here in details and all cases series in summarized in Table 1.

Case 1

A 55-year-old male presented with a slowly growing left neck mass for 3 years duration presented with hoarseness of voice & dysphagia for 6 months. Flexible laryngoscopy examination of larynx revealed left supraglottic bulge with normal overlying mucosa. The patient did not follow up from 3 months and suddenly came to Emergency department with stridor that necessitated tracheostomy. At the same sitting, Direct Laryngoscopy was done, two biopsies were taken. The result came as inconclusive so CT-guided FNAC done and was suspicious for a cartilaginous tumor. CT scan of the neck showed a huge homogenous mass epicentered at the left thyroid lamina with elements of calcification around it with no detected lymph node enlargement. We did total laryngectomy for patient and histopathological results come as

high-grade chondrosarcoma of thyroid cartilage. Now patient with smooth post-operative follow up with no recurrence (Figure 1).

Case 2

A 60-year-old male presented with voice change and dyspnea for 1 month associated with stridor for whom emergent tracheostomy done. Flexible laryngoscopy revealed limited mobility of left vocal cord with subglottic mass obscuring tracheal lumen. CT scan of neck showed heterogonous mass epicentered at postero-lateral left aspect of cricoid cartilage with calcification, no detected lymph node enlargement. Direct Laryngoscopy revealed subglottic mass with normal overlying mucosa. Multiple biopsies were obtained and result came as chondrosarcoma of larynx (Figure 2). Total laryngectomy was done and final histopathology came back as Grade I chondrosarcoma. Now patient is still under surveillance with no recurrence (Figure 3).

Date	Age	Sex	Origin	Side	Symptoms	Diagnosis	Surgery & neck	Histopathology	Follow-up
2002	50	Male	Thyroid	Left huge	Neck mass Hoarseness Dyspnea Dysphagia Stridor	Clinical CT-scan Biopsy	Total laryngectomy Neck (N0)	Grade II	Normal
2003	55	Male	Arytenoid	Right huge	Hoarseness Dyspnea Dysphagia Stridor	Clinical CT-scan Biopsy	Total laryngectomy Neck (N0)	Grade III	Lost
2003	45	Male	Cricoid	Left large	Hoarseness Dyspnea Dysphagia Stridor	Clinical CT-scan Biopsy	Total laryngectomy Neck (N0)	Grade I	Normal
2014	55	Male	Thyroid	Left huge	Neck mass Hoarseness Dyspnea Dysphagia Stridor	Biopsy not conclusive CT-guided FNAC	Total laryngectomy Neck (N0)	Grade II	Normal
2015	59	Male	Cricoid	Right small	Hoarseness Dyspnea Dysphagia Stridor	Clinical CT-scan Biopsy	Total laryngectomy Neck (N0)	Grade II	Normal
2019	60	Male	Cricoid	Left moderate	Hoarseness Dyspnea Dysphagia Stridor	Clinical Biopsy	Total laryngectomy Neck (N0)	Grade I	Normal

Table 1 – Summary of cases of laryngeal chondrosarcoma in the past 20 years; HP = histopathology

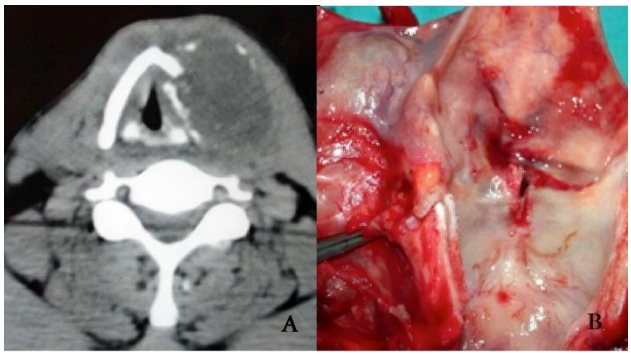

Figure 1 – Laryngeal chondrosarcoma. A – axial CT showing homogenous mass epicentered on LT thyroid cartilage with peripheral calcifications; B – gross specimen showing mass bulging from LT thyroid cartilage

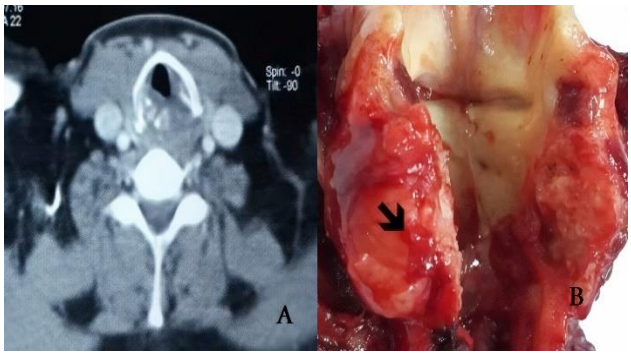

Figure 2 – Laryngeal Cricoid Chondrosarcoma. A – axial CT showing mass arising from cricoid cartilage with calcifications B – laryngectomy specimen showing mass arising from cricoid cartilage (black arrow)

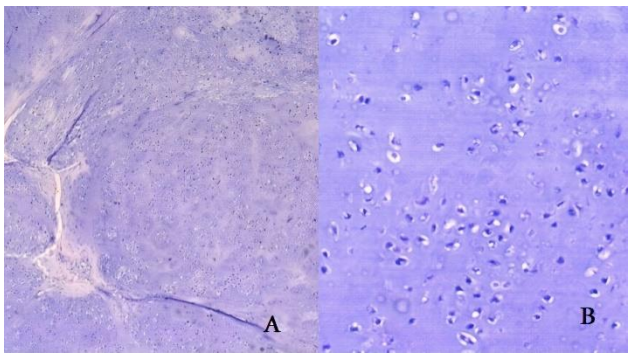

Figure 3 – Final histopathology of cricoid chondrosarcoma. A – low power (×10) lobulated hypercellular cartilaginous neoplastic growth. B – high power (×400) evident hypercellularity, frequent binucleation as well as mild to moderate nuclear atypia and rare mitotic figures

Discussions

The larynx is a rare subsite for all chondrosarcoma arising in the head and neck [13]. Although LCS represents only 1-2% of laryngeal neoplasm [1], it is the most common non-epithelial laryngeal tumor [2].

LCS often occurs in 6th and 7th decades and has a predilection for men (3.6:1) [7]. In our case series all the patients were males between 40-60 years. The cricoid is the most common site of origin within the larynx followed by the thyroid cartilage [8]. In our patients, the site of origin was the cricoid cartilage in 3 cases, the thyroid cartilage in two cases and the arytenoid cartilage in one case. The most common presenting symptoms in a decreasing frequency, are hoarseness, dyspnea, dysphagia, dysphonia, cough, neck mass, stridor. The onset of symptoms is insidious given the indolent nature of this tumor [1,3]. Hoarseness of voice and dyspnea was reported to be the first symptoms in our patients. However, all our patients either did not seek medical advice or lost for follow up until became in advanced stage. CT neck is considered the imaging of choice and presence of a hypo dense mass with calcifications inside and cartilage destruction is diagnostic [7, 9]. CT scan was obtained and was the best imaging modality aided in diagnosis of the six cases. LCS is classified into three grades; and Grade I account for 70-80% of cases [12].

There are many treatment options described in the literature spanning the spectrum from limited resections to radical surgery as total laryngectomy. Indications of total laryngectomy as suggested by Jones [14] are: extensive tumor with inability to preserve airway patency, recurrence, anaplastic tumors. As the cricoid cartilage is the most common site of origin specifically its posterior lamina, any tumor involvement > 50% of this cartilage should be treated with total laryngectomy [15]. In our case series, there was extensive involvement of the cricoid or thyroid cartilage, and this explains the need for total laryngectomy in the six patients. LCS has good prognosis with ten- year survival rate of 95% [9]. LCS has recurrence rate of 35%-40%. Recurrence is due to incomplete excision or high-grade tumor [7]. The metastasis rate is very low (12-15 %) and occurs mostly in high grade tumors [1]. Follow up course in our patients revealed no recurrence or metastasis in five cases while one case lost from follow up.

Conclusion

Laryngeal chondrosarcoma is a slowly growing tumor that is rarely metastasizing to neck lymph nodes. Airway obstruction is the most presenting symptoms. Endoscopic debulking is the preferable treatment option with close follow up. Once the patient needs tracheostomy, a sort of laryngectomy (Mostly total laryngectomy) is recommend. No chemo-radiotherapy is recommended for organ preservation protocol.

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