Laryngeal Chondrosarcoma a Case Report

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Abstract
Chondrosarcomas of the larynx are rare cancer and are more frequently located at cricoid cartilage level. Frequency, related to other cartilage cancers, is lower than 1%, whereas if related to cancers located in the larynx, it is between 0.07% and 0.2%. They are characterised by a low tendency to metastatic diffusion (low grade). Prognosis is generally good. The treatment of choice is surgery, which may be endoscopic or “open partial surgery”, if extension of the cancer is limited, or total laryngectomy. In this report, a case of low malignancy cricoid chondrosarcoma is described which was treated in the Otolaryngology-Head and Neck Surgery Department of University of Thessaly. Chondrosarcoma of the larynx shows a slow and painless growth, the first symptom is often an ingenuous dysphonia. Laryngoscopy reveals tumefaction of the larynx, covered by intact mucosa. Computerized tomography imaging with contrast and magnetic resonance imaging defines not only coarse calcifications, pathognomonic of chondromatous neoformations but also the relationship of the neoformation with the surrounding tissues. However, histology remains the gold standard for diagnostic purposes.

Keywords: Larynx; Malignant tumours; Chondrosarcoma; Cricoid tumour

Introduction
Chondrosarcoma is a malignant cartilaginous tumor of the larynx exceptional. Frequency, compared to other cancers of the cartilage, is less than 1%, while that associated with cancer of the larynx is located between 0.07% and 0.2% [1-5]. The pathogenesis is unknown. It can occur at any age but is most often described as an adult and there is a male predominance (sex ratio 5:1). Chondrosarcoma is a malignant tumor of very slow growth of the proliferation of hyaline cartilage. It is rarely found at the head and neck, and is normally described in the pelvis, femur, ribs, scapula, scapula, sacrum and sternum. Only 10-12% of chondrosarcoma are located at the head and neck. In the larynx, the most common location is posterolateral region of the cricoid cartilage. It is exceptionally described in the lower portion of the laryngeal surface of the thyroid cartilage or the arytenoid cartilages. The regional and distant metastases when tumors of low malignancy are uncommon [2,6]. Surgery is the treatment of choice and can be either endoscopic or partial “open surgery” or a total laryngectomy, depending on the extension and histological grade of the tumor [4,7-9]. Chondrosarcoma of the larynx generally has a good prognosis (low grade) [10,11]. In this report, a case of cricoid chondrosarcoma grade II is described which was treated at the Otorhynolaryngology Department of the University of Thessaly Greece.

Case Report
A 55 year old male immigrant came to our attention with dysphonia which had been slowly increased. He had obvious inhalation syrinxus with oxygen saturation 95% and did not mention any dysphagia. Fiberoptic laryngoscopy revealed a large neoformation, about 3 cm in diameter, in the right subglottic region, partially obstructing the respiratory space. The neoformation, situated at the level of the cricoid cartilage, was covered by mucosa with a normal appearance. The right vocal cord was fixated. No adenopathy was found upon palpation at laterocervical level.
After a very difficult intubation a biopsy was taken from the neoplasm and a tracheotomy was performed. Considering the grade and size of the tumor a total laryngectomy was performed

The histological report revealed a grade II chondrosarcoma (Figure 3).

After 9 years follow up the patient showed no sings of local recurrence or distal metastasis.

Discussion

Chondrosarcoma represents 10-20% of all malignant primitive bone cancers [2,4,11].

Chondrosarcoma of the larynx is a rare pathological condition, and is certainly the most frequent mesenchymal cancer of this organ. The cricoid is the cartilage most commonly involved (75%) [5] primarily of the posterior lamina; other locations involved (in order of frequency) are: the thyroid cartilage, the arytenoids and the epiglottis [1,2].

The exact pathogenesis still remains to be elucidated; some aetiopathogenetic hypotheses attribute this pathological condition to local injuries [4], ossification anomalies, chronic inflammation and metabolic disorders related to old age [12].

Chondrosarcoma of the larynx generally occurs in the age group comprised between the sixth and seventh decade of life [13]. Dysphonia represents the principal clinical sign, while dyspnoea is associated when the cancer increases in size; dysphagia is rarely present.

In indirect laryngoscopy, a sub-mucosa tumefaction with intact mucosa may be detected; furthermore, the first sign of posterior cricoid involvement may be stiffness of vocal cord, due to blocking of the cricoarytenoid articulation and not because of infiltration of the recurrent nerve [6,13].
CT, is the first-choice radiological test; this method of “imaging”, with contrast medium, allowed us to identify the location, dimensions, limits as well as the adjacent relations of the neoplasm and also to observe its calcification, which is pathognomonic for this mesenchymal cancer [14]. MRI is a complementary test, as it does not yield additional information.

The diagnosis of certainty of chondrosarcoma is given by the histological analysis. Chondrosarcoma can only be diagnosed, with certainty, by means of histological analysis. Biopsy performed in direct laryngoscopy can sometimes be difficult on account of the hard tissue typical of the lesion; furthermore, precise distinction between chondroma and low malignancy chondrosarcoma, on small biopsy specimens is not always easy, and the final diagnosis is often preformed on the surgical specimen. The pathological analysis technique must be rigorous and accurate, respecting, during decalcification of the sample, the cellular and nuclear morphology, which are fundamental elements for the diagnosis [10,15].

The histological aspect of this mesenchymal cancer was classified by Evans et al. (1977) [10] into different grades of malignancy and clinical behaviour: a form with low grade malignancy (1st grade) is characterised by high cellular density with negligible hyaline cartilage matrix, some cells are bi-nucleated or show nuclear anomalies and mitosis; with a not aggressive progress and a low tendency to create metastases.

The intermediate form (2nd grade) and the high malignancy grade form (3rd grade), which show greater cellular and nuclear anomalies and a high miotic index, both have a worse prognosis [10,15].

Chondrosarcoma with a low malignancy grade is the most frequent cancer at larynx level [11,14,16,17].

The treatment of laryngeal chondrosarcoma is surgical, with concern for conservation of function, while respecting the rules of cancer surgery. The main problems with this surgery are the risk of recurrence and postoperative stricture. The treatment of choice for symptomatic tumors is surgical in the absence of contraindications, in case of diagnostic uncertainty. The resection depends on the stage of the tumor extension and is subject to many controversies, the well limited tumors justify a tumor resection in healthy zone followed by a closure by local flap without calibration. In earlier position, the risk of stenosis is higher and has to consider a a tummy army by the introduction of a silicone stent cricotraheal. For larger lesions (greater than 2 cm), the restoration of continuity is called to the establishment of a Montgomery tube or at the resection, anastomosis thyrotrachéale. Achieving unity cricoaryténoidienne, imposes a subtotal laryngectomy Pearson type if the injury does not exceed the median line or total laryngectomy if the invasion is bilateral [3,4,16-18].

The present surgical trend, of most Authors, is to perform total laryngectomy whenever the neoplasm extends beyond the half cricoid cartilage or when the chondrosarcoma shows a high grade of malignancy as we did in our case [7,19].

The role of radiotherapy is uncertain and controversial; indeed, many Authors maintain that the chondrosarcoma displays negligible sensitivity to radiations [2,16]. Others have recently presented positive results with exclusive radiation treatment (60-70Gy), also in low malignancy cases [20,21]. McNaney et al. obtained good local control of the pathology, at three years, with a combination of neutrons and photons [22].

According to most Authors, radiotherapy should be performed exclusively on undifferentiated chondrosarcomas, as post-operative adjuvant treatment [1-3,16].

Chemotherapy does not seem to have a place in the curative treatment; it is reserved for palliative care in case of aggressive tumors with local invasion. Local recurrences of chondrosarcoma are common. Their reported incidence in literature varies from 35% to 40%. Metastases are not particularly common, and occur in 2-10% of cases. Lung and cervical lymph nodes are the sites most often affected. Due to the low metastatic potential and slowing tumor growth, survival at 10 years exceeds 95% and even the onset of recurrence did not significantly affect survival. The main prognostic factors are histologic grade, location, extent and quality of the initial resection [23-25]. Prognosis of this condition depends upon the radicality of the resection, on the extension and histological grade of the chondrosarcoma. The low grade of malignancy rarely relapses and distant metastases are not frequent, even if, in 1982, Neel and Unni published a retrospective study in which pulmonary metastases occurred after 20 years of follow-up [11].

**Conclusion**

Chondrosarcoma of the larynx with low grade of malignancy is characterised by slow growth.

In our opinion, conservative surgery may be performed, but total laryngectomy becomes necessary if the cancer is large, if the margins have infiltrated surrounding tissues or when relapse occurs, and whenever the grade of malignancy is high.

Adjuvant radiotherapy may be useful for undifferentiated chondrosarcomas.

Regular and long-term follow-up is mandatory, in order to detect relapses and metastases.

**References**