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REVIEW ARTICLE

Laryngeal chondrosarcoma – Ten years of experience☆

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KEYWORDS
Chondrosarcoma;
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Abstract
Introduction: Laryngeal involvement by cartilaginous tumors is rare. However, although accounting for only 1% of laryngeal tumor pathology, they are the most frequently occurring non-epithelial neoplasms. The most probable location is the endolaryngeal surface of the cricoid cartilage. Their symptoms are variable, depending on the size and location, and may include hoarseness, stridor, and dyspnea. Treatment is based on surgical excision. Some centers take into account the degree of differentiation and whether it is a case of relapse when deciding to perform a radical resection.
Aim: To evaluate this disease in a sample of the Portuguese population.
Methods: A review of the medical records from 2002 to 2012 by assessment of clinical processes was performed. Data on demographics, clinical treatments, and outcomes were collected.
Results: Six patients were included in the study. Five of them underwent total laryngectomy, and in one case, partial excision of the thyroid cartilage was performed. None of the patients had either metastases or tumor-related death.
Conclusion: Laryngeal chondrosarcomas remain a rare disease of unknown etiology, with slow and insidious symptoms. The treatment is surgical, with favorable prognosis, and metastases rarely occur. The main concern regards their propensity to relapse.

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PALAVRAS-CHAVE
Condrossarcoma;
Doenças da laringe;
Eoplasias laríngeas

Condrossarcoma da laringe – 10 anos de casuística

Resumo
Introdução: O acometimento laríngeo por tumores cartilaginosos é raro. No entanto, apesar de representarem 1% da patologia tumoral laríngea, são as neoplasias não epiteliais mais frequentes. Localizam-se mais frequentemente na face endolaríngea da cartilagem cricóide. Tem sintomatologia variável consoante o tamanho e a localização, podendo incluir disfonia, estridor e dispneia. O tratamento é essencialmente cirúrgico. Alguns centros levam em conta o grau de diferenciação e de se tratar ou não de recidiva, quando da decisão de ressecção mais ou menos radical.
Objetivo: Avaliar esta patologia numa amostra da população portuguesa.
Método: Revisão da casuística no intervalo de tempo 2002-2012, através de consulta dos processos clínicos. Foram coligidos os dados demográficos e clínicos relevantes, os tratamentos efetuados e os resultados.
Resultados: Foram incluídos seis pacientes. Cinco foram submetidos à laringectomia total e um foi submetido à excisão da asa esquerda da cartilagem tiroide. Nenhum apresentou metástases ou morte relacionada com o tumor.
Conclusão: Os condrossarcomas laríngeos permanecem como patologia rara, de etiologia desconhecida, com crescimento lento e clínica insidiosa. O tratamento é cirúrgico, com prognóstico favorável, com a metastização a ocorrer raramente. A maior preocupação decorre da sua propensão à recidiva.

Introduction

Rare and insidious pathological entities require extra attention for their appropriate diagnosis and treatment. Laryngeal involvement by cartilaginous tumors is rare.[1–4] However, despite representing only 1% of laryngeal tumor pathology, they are the most frequently occurring non-epithelial neoplasms.[2] Chondromas are the most common benign tumors, most often affecting the cricoid cartilage (75%).[3] Only approximately 0.1% of cartilaginous tumors are chondrosarcomas, which are most commonly low grade.[1,2,5]

Chondrosarcoma is a slow-growing malignant tumor, most commonly located in the pelvis, femur, ribcage, humerus, scapula, fibula, sacrum, and sternum. More rarely, it is found in the head and neck, in approximately 10–20% of the total number of cases.[2,5] In the larynxchondromas they are located in most cases in the endolaryngeal surface of the posterior portion of the cricoid cartilage.[1,2,5,6] It less often affects the arytenoid cartilage or the thyroid cartilage in the inferior border of its laryngeal surface.[5,7]

Its etiology remains unknown, but there are some hypotheses regarding the contribution of medullary-cervical trauma and vertebral instability,[3] repeated laryngeal trauma,[5] radiotherapy, Teflon® injection,[6] and irregular ossification of laryngeal cartilage.[7]

As for the epidemiological characteristics, it is most often observed in adult patients in the sixth and seventh decades of life,[1] although it can be found at any age. It affects more males than females, at a ratio of 3:1.[3] Cartilaginous tumors with higher degrees of malignancy are found at older ages.[6]

Symptoms are variable and depend on tumor size and location. Tumors may be asymptomatic while small, and their growth can trigger dysphonia, stridor, dyspnea, and dysphagia. The clinical picture very rarely includes pain.[1,3,5] There have been cases described with concurrent vocal cord paralysis, which is thought to originate from cricoarytenoid joint fixation and not the recurrent laryngeal nerve lesion.[1]

Endoscopic assessment must be considered, taking into account the tumor's common subglottic location. The clinician must search for submucosal bulging, usually with regular mucosa and in a more posterior location, which is small-sized (usually smaller than 3–4 cm).[5] Vocal cord immobilization can be found.

During imaging assessment and in the presence of larger masses, an anterior displacement of the larynx in the lateral cervical radiograph can be observed. However, its clinical suspicion requires the performance of computed tomography (CT) or magnetic resonance imaging (MRI). Some authors suggest that CT is the imaging examination of choice,[1–3,8] disclosing a hypodense, well-defined image with calcifications inside, cartilage destruction, and structure distortion.[9]

Others report the superiority of MRI due to its greater accuracy in distinguishing between tumor and other paralaryngeal tissues.[5,8] The signal strength is low on T1 and high on T2, with a characteristic mosaic pattern.[8] The F-18 fluorodeoxyglucose-positron emission tomography (FDG-PET) is helpful in tumor grading, metastasis detection, and local recurrence assessment. The uptake value of 1.3 was set as the limit between benign and malignant lesions, with the higher uptake showing greater differentiation.[8,10]

In spite of clinical suspicion and the above-mentioned complementary diagnostic tests, the diagnosis can only be definitely established by histological study. Often, this can only be correctly achieved through the study of the surgical