



Synovial Sarcoma of the Larynx: Case report from the perspective of swallowing, voice and quality of life

Sarcoma sinovial da laringe:
Relato de caso sob a perspectiva da
deglutição, voz e qualidade de vida

Sarcoma de laringe sinovial:
reporte de un caso desde la perspectiva
de la deglución, la voz y la calidad de vida

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Abstract

Introduction: Synovial sarcoma is an aggressive tumor commonly found in the lower limbs and rarely described in the larynx. The literature recommends multimodal treatment with radiotherapy, chemotherapy and total laryngectomy. **Purpose:** To describe the functional and quality of life impacts on voice and swallowing of an adolescent who developed a monophasic synovial sarcoma of the larynx. **Method:** A 16-year-old adolescent who developed a monophasic synovial sarcoma of the larynx in childhood, who

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underwent an extended arytenoidectomy due to laryngopharyngeal access associated with radiotherapy at the age of 11. The less aggressive cancer treatment was adequate for cancer control and functional results of the case described here, presenting a voice with satisfactory quality and exclusive oral route. The endoscopic evaluation showed a larynx with infantile proportions, revealing growth interruption at the time of treatment. The patient has a 5-year and 6-month survival rate, higher than the average two-year survival rate described in the literature, referring to a good quality of life. **Conclusion:** The adolescent has oncological control and satisfactory functional results of voice and swallowing, without major interferences in his quality of life. Family support and specialized multidisciplinary care proved to be paramount.

Keywords: Head and neck cancer; Laryngectomy; Dysphagia; Voice; Pediatrics.

Resumo

Introdução: O sarcoma sinovial é um tumor agressivo comumente encontrado nos membros inferiores e raramente descrito na laringe. A literatura recomenda tratamento multimodal com radioterapia, quimioterapia e laringectomia total. **Objetivo:** Descrever os impactos funcionais e de qualidade de vida em voz e deglutição de um adolescente que desenvolveu um sarcoma sinovial monofásico da laringe. **Método:** Adolescente de 16 anos que desenvolveu um sarcoma sinovial monofásico da laringe na infância, submetido a uma aritenoidectomia estendida por acesso laringofaríngeo associado à radioquimioterapia aos 11 anos de idade. O tratamento oncológico menos agressivo foi adequado para o controle oncológico e resultados funcionais do caso aqui descrito, apresentando voz com qualidade satisfatória e via oral exclusiva. A avaliação endoscópica demonstrou uma laringe com proporções infantis, revelando a interrupção do crescimento no momento do tratamento. O paciente apresenta sobrevida de 5 anos e 6 meses, superior à sobrevida média de dois anos descrita na literatura, referindo boa qualidade de vida. **Conclusão:** O adolescente apresenta controle oncológico e resultados funcionais de voz e deglutição satisfatórios, sem grandes interferências em sua qualidade de vida. O suporte familiar e atendimento multiprofissional especializado demonstraram-se primordiais.

Palavras-chave: Neoplasias de cabeça e pescoço; Laringectomia; Disfagia; Voz; Pediatria.

Resumen

Introducción: El sarcoma sinovial es un tumor agresivo que se encuentra comúnmente en las extremidades inferiores y rara vez se describe en la laringe. La literatura recomienda el tratamiento multimodal con radioterapia, quimioterapia y laringectomía total. **Propósito:** Describir los impactos funcionales y de calidad de vida en la voz y la deglución de un adolescente que desarrolló un sarcoma sinovial monofásico de la laringe. **Método:** Un adolescente de 16 años que desarrolló un sarcoma sinovial monofásico de la laringe en la infancia, que se sometió a una aritenoidectomía prolongada debido al acceso laringofaríngeo asociado con radioterapia a la edad de 11 años. El tratamiento contra el cáncer menos agresivo fue adecuado para el control del cáncer y los resultados funcionales del caso descrito aquí, presentando una voz con calidad satisfactoria y una vía oral exclusiva. La evaluación endoscópica mostró una laringe con proporciones infantiles, revelando la interrupción del crecimiento en el momento del tratamiento. El paciente tiene una tasa de supervivencia a 5 y 6 meses, superior a la tasa de supervivencia promedio de dos años descrita en la literatura, que se refiere a una buena calidad de vida. **Conclusión:** El adolescente tiene control oncológico y resultados funcionales satisfactorios de voz y deglución, sin grandes interferencias en su calidad de vida. El apoyo familiar y la atención multidisciplinaria especializada resultaron ser primordiales.

Palabras clave: Neoplasias de cabeza y cuello; Laringectomía; Disfagia; Voz; Pediatría.

Introduction

Synovial sarcoma is a rare and aggressive tumor of connective tissue, usually originating in mesenchymal tissue. It accounts for 5% to 10% of all connective tissue sarcomas and most commonly affects the upper and lower limbs¹⁻³. Primary synovial sarcoma of the head and neck is rare, accounting for less than 5% of all synovial sarcomas. The most common site of synovial head and neck sarcoma is the hypopharynx, rarely described in the larynx^{1,2}.

In a systematic review of 2019, 28 independent case reports and 4 case series were found containing 11 cases of synovial laryngeal sarcoma worldwide¹. The literature recommends multimodal treatment with radiotherapy and total laryngectomy with an average survival of two years². Several aspects of synovial sarcoma are not adequately understood, such as the real implication of the tumor size, the histological subtype and the subtype of the SYT-SSX fusion gene in prognosis and functionality^{1,4}.

In order to optimize the overall treatment results, a multidisciplinary patient-centered approach should be adopted. Despite its importance, there are few reports of specialized care provided by multiprofessional health services, including speech therapy, physiotherapy and psychology¹. Laryngectomy and the consequent laryngeal dysfunction may be associated with reduced quality of life and vocal and swallowing disorders. For young patients affected by synovial laryngeal sarcoma, this impact is likely to be disastrous due to impairment in development^{4,6}. Providing rehabilitation and psychological support for these patients is essential.

The aim of this study was to describe the clinical aspects and functional and quality of life impacts on voice and swallowing of a teenager who developed a monophasic synovial sarcoma of the larynx.

Case report

The study was approved by the Institution's Ethics and Research Committee under number 89042418.7.0000.5274. The patient and family members signed an informed consent form.

An 11-year-old male patient with reports of difficulty on swallowing solids, progressing to

fluids two months before diagnosis in 2013. Due to dysphonia and the presence of a compressive mass, he underwent partial tumor resection and emergency tracheostomy. Videolaryngoscopy revealed a vegetative mobile polypoid lesion in the supraglottic region. The lesion obstructed the vocal folds and extended to the base of the tongue, being visible in the oropharynx. The immunohistochemical evaluation was positive for vimentin, B12, EMA, CD56 and CD99, indicating neoplasia of synovial sarcoma and the cytogenetic evaluation revealed the monophasic subtype. The tumor was classified as E3 and G III.

The patient was submitted to the neoadjuvant European chemotherapy protocol (CT) (2 cycles of ifosfamide and doxorubicin) for cytoreduction in the same year. After these procedures, a surgical approach was performed by means of laryngofissure associated with enlarged arytenoidectomy on the right, and the histopathological examination revealed positive margins. The surgery was followed by radiotherapy (RT) with a dose of 59.4Gy and CT (3 cycles) adjuvant.

Speech therapy was started in the preoperative period in order to manage complaints and symptoms. In the immediate postoperative period, the fundamental objective was to enable decannulation and, subsequently, to enable safe swallowing and vocal rehabilitation. The patient had a rough and breathy voice and severe dysphagia, due to edema, absence of tissues and reduced mobility. Strategies such as masako, mendelsohn and postural maneuvers associated with swallowing function, occlusion of the tracheostomy with coaptation exercises and glottic vibration were used. The exercises were suggested in repetitions of ten with a frequency of three times a day.

Four years after the end of treatment, at the age of 16, the neck and lower third of the patient's face were disproportionate to the rest of the body. The endoscopic evaluation indicated that the larynx had infantile proportions: epiglottis and prominent arytenoids and the membranous portion of the vocal folds reduced for the age group. There was an atypical formation of a fibrous fold in the surgical region, which helped to close the laryngeal vestibule during swallowing and, thus, prevented food from entering the lower airway (Figure 1).

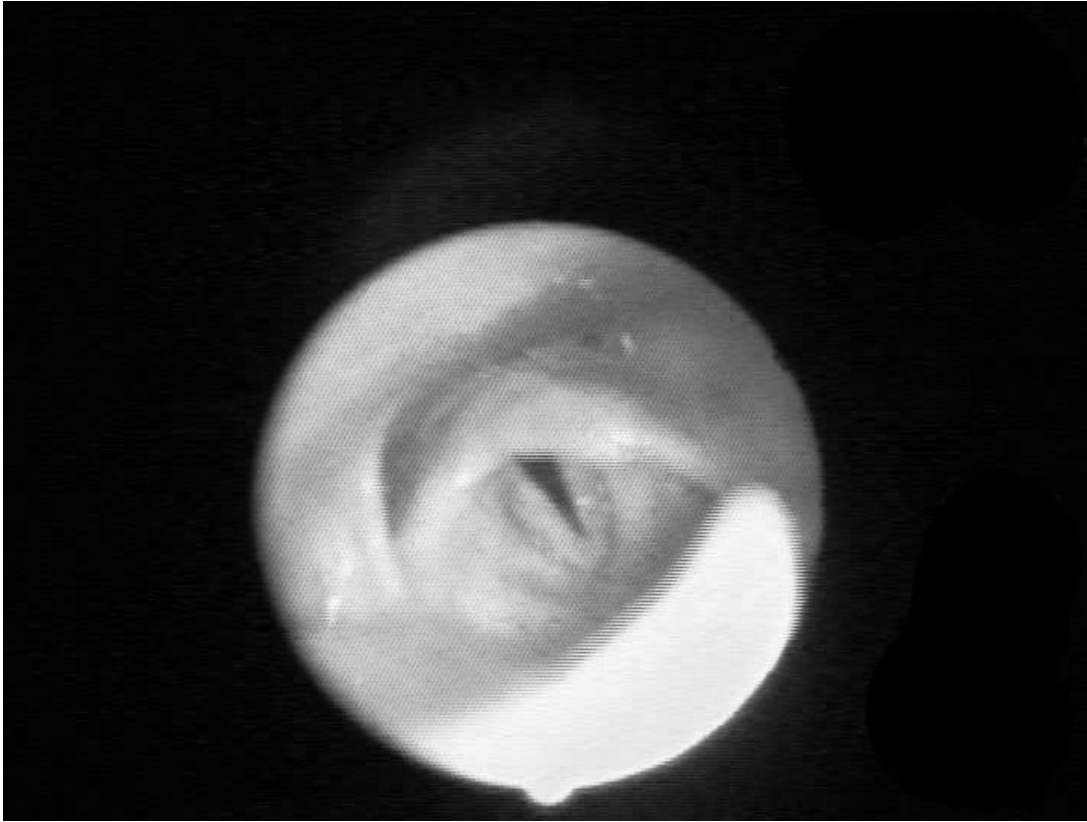


Figure 1. Visualization of pharyngolaryngeal structures during vocal fold abduction. Note the absence of the right aryepiglottic fold, as well as the arytenoid on the same side. The prominent left arytenoid and fibrous fold in the surgical region on the right.

The patient reported difficulty on swallowing solid foods (requiring occasional compensations) and complaints of dry mouth at the time of the objective evaluation of swallowing, using artificial saliva under speech therapy guidance. The functional evaluation of swallowing by means of videofluoroscopy for thin liquid, semi-liquid and pasty, with volumes of 5, 10 and 20 ml, respectively, demonstrated an effective velopharyngeal mechanism, good oral motor control and adequate hyolaryngeal excursion. However, the patient presented incoordination between the oral and pharyngeal events of swallowing in thin liquids, with stasis in the vallecula and left piriform recess (in anteroposterior view), requiring multiple swallowing for whitening. Despite the changes described, during this exam there was no laryngotracheal penetration and aspiration (Figure 2).

The auditory-perceptual assessment was promoted through the Consensus Auditory-Perceptual Evaluation of Voice (CAPE V) and the acoustic analysis of the voice was performed using the VoxMetria® 3.0 (CTS) program with sustained vowel analysis / e /, automated speech (counting from 1 to 20) and spontaneous speech. The patient had good pneumophonic coordination with satisfactory airflow control. The vocal quality was rough and tense with a high pitch, agreeing with the fundamental frequency (f_0) of 244.57 Hz of the sustained vowel / e /.

According to the voice quality of life protocol, the Voice Handicap Index (VHI), the patient did not have significant impacts on quality of life, despite the dysphonia described above⁷. It was found that the patient is aware that he is dysphonic, however, this aspect does not severely impact his quality of life (Total Score is 25).



Figure 2. Videofluoroscopic image after swallowing 5ml of thin liquid. Stasis in piriform recess, vallecula and fibrous fold from the surgical procedure.

Despite the functional impairment and current swallowing complaints, through the Quality of Life in Dysphagia protocol (SWAL-QOL) its score was 182, well above the cutoff point of 100, which is the minimum level for comfortable swallowing with low social impact⁸. The patient considers himself to have a good quality of life and seeks to have a normal adolescence, with sports activities (basketball) and singing.

At the end of the follow-up of this study, the patient was in good general health, with no sign of recurrence of the cancer disease (five years and six months after treatment). He remains under outpatient follow-up for cancer control and speech therapy rehabilitation. Despite all the structural and functional changes in force, the stomatognathic functions of the larynx (breathing, swallowing, voice and speech) remained with good results in activities of daily living.

Discussion

Synovial laryngeal sarcoma is a rare tumor that requires multimodal treatment with combined CT and QT, traditionally associated with total laryngectomy. There is no report in the Brazilian literature that describes such pathology in a child. The publication of national practices and divergent from traditional cancer treatments are essential for improving the performance of this type of pathology in the population, as well as the development of less aggressive techniques with better functional and quality of life results².

Radiotherapy treatment has an immediate and long-term effect, causing reduced development of the irradiated region in relation to the rest of the body. In addition, functionally, the patient complains of dry mouth and stasis, justified by fibrosis in the cervical and laryngeal tissue, damage to the

salivary glands and less mobility of the structures that act on swallowing. Therefore, it is possible to observe a reduced hyolaryngeal performance, alteration in the propulsion of the bolus due to the impairment of the tissue at the base of the tongue and a lower amplitude of pharyngeal contraction^{5,6,10}. All of these aspects can justify a change in the oral motor control for the liquid and deficits in the propulsion of the solid food.

The surgical procedure, associated with the effects caused by radiation, promoted the formation of a fibrous fold in the pharyngeal region. Aryepiglottic folds have the function of directing food from the vallecula to the ipsilateral piriform recess, as a pressure-independent swallowing mechanism to protect the lower airways. In this case report, the patient's arytenoid and right aryepiglottic fold were sectioned, which could represent a risk to the patient during swallowing, generating laryngotracheal penetration and aspiration. The appearance of the fibrous crease compensated for the absence of the dried crease, helping to protect the lower airways¹⁰. At the same time, the fibrous fold reduces the pharyngeal space for the passage of the bolus, justifying the complaint / difficulty in swallowing with some solids that, due to anatomical irregularity, are ecstatic in the pharyngeal region requiring multiple swallows for cleaning.

All teenagers go through vocal changes. These changes are part of a natural physiological process, in which the vocal tract acquires mature structural characteristics. Studies on vocal disorders in pediatric patients undergoing treatment for head and neck cancer were not found in the literature. The patient's high pitched voice can be justified by the hypo-development of the larynx and neck, with infantile proportions and fibrosis from RT. However, the breathiness of the voice and the intense roughness resulting from the absence of an arytenoid partially disguise the vocal deviation and make this voice socially acceptable⁶.

Negative effects on the quality of life and health of childhood cancer survivors are expected^{5,6}. However, little is known about the late effects of this treatment during the natural course of aging⁶. In a pediatric tumor, it is necessary to consider rehabilitation and treatment after diagnosis with caution, in order to guarantee functional results that enable the development of childhood, adult life, aging and all its complexity. Management aims to rehabilitate, enable and prevent late dysfunctions⁴.

In all assessments of quality of life, the patient, in this study, had scores within the standard values, which is probably due to the constant multiprofessional and family care^{1,7,8}.

Conclusion

Laryngopharyngeal arytenoidectomy associated with chemotherapy and radiotherapy was adequate for the oncological control and functional results of swallowing and voice in the case described here, in an 11-year-old male child. The patient has a 5-year and 6-month survival, with moderate dysphonia and mild dysphagia without relevant interference in quality of life. Endoscopic evaluation shows the larynx in infantile proportions. The results came from specialized and family multiprofessional support at all times of treatment.

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