CLINICAL CASE

LARYNGEAL SYNOVIAL SARCOMA – A CASE REPORT

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Abstract

Laryngeal synovial sarcoma is an extremely rare mesenchymal tumor. During the past four decades, literature findings revealed under 40 cases with this localization. We are bringing forward the case of a 70-year-old male patient who was referred to the ENT Clinic as a respiratory emergency – a tumor obstructing the laryngeal glottis, for which an emergency tracheotomy was initially performed, followed by a CT scan and a biopsy under general anesthesia through suspension videolaryngoscopy. The histopathological report was suggestive of a biphasic synovial sarcoma. A multidisciplinary team set the surgical indication for total laryngectomy followed by chemo-radiotherapy. Long-term follow-up is mandatory for recurrence prevention. Patient has checked in for follow-up every 3 months over the past 2 years, with no macroscopic signs of tumoral relapse.

Keywords: larynx, synovial sarcoma, total laryngectomy, rare tumors

Introduction

Synovial sarcoma represents a type of soft tissue sarcoma usually encountered in para-articular sites [1], with a predilection for the extremities. It accounts for up to 10% of all soft tissue sarcomas, out of which approximately 10% occur in the head and neck region [1-4].

In 1975 Gatti and Miller described the first case of laryngeal synovial sarcoma, with the next 4 decades seeing about 40 published cases of synovial sarcomas with a laryngeal starting point [5,6].

While synovial sarcomas may affect all age groups, it is most commonly encountered within the second and third decades of life, with a second peak of incidence in the fifth and sixth decades, with men being more commonly

affected (a 3 to 1 ratio when compared to women) [2,4].

Synovial sarcomas may be monophasic, biphasic or undifferentiated, where the biphasic phenotype represents a tumor made of malignant spindle cells interposed with epithelial elements [1, 2].

Patients may report hoarseness in the early stages, given the nonspecific symptoms of the slowly growing and painless tumor mass, with dyspnea, upper respiratory distress and at times even dysphagia in the more advanced stages of the disease [1,2].

Treatment generally consists of surgical resection of the tumor, with negative soft-tissue margins, followed by multimodal therapy consisting of both radiotherapy and chemotherapy. Local recurrence may develop in

up to 25% of patients, with another 25% having distant metastases, most commonly in the lungs [1,2,4]. Does not usually involve cervical lymph node metastasis. Better prognosis associated with smaller, localized tumors and young age.

Case presentation

A 70-year-old male patient, heavy drinker and smoker, living in the countryside, was admitted in the ENT Clinic for severe inspiratory dyspnea, cyanosis and wheezing. Upon clinical examination of the pharynx and larynx, a violaceous, corneous, bilobed tumor is identified, arising from the laryngeal ventricle, the petiole (root) of the epiglottis and the ventricular folds commissure.

An initial diagnosis of acute respiratory insufficiency owing to a laryngeal tumor is placed.

We perform an emergency tracheostomy under local anesthesia, securing the patients airway.

A CT scan of the neck, performed both precontrast (native-phase) and postcontrast, revealed an obstruction of the laryngeal lumen by way of a heterogeneous, iodophilic mass of tissue, which extends within the hypopharynx. No cervical lymphadenopathy identified.

The patients' medical history did not reveal any significant pathologies, with blood tests, chest X-ray and cardiological examination – all within normal limits.

We decided to perform a tumor biopsy, under general anesthesia with tracheal intubation, through suspension videolaryngoscopy. The histopathological report indicates a diffuse malignant epithelial and mesenchymal proliferation, with cells revealing a pronounced nuclear pleomorphism. Resembles epithelial-mesenchymal tumor - biphasic synovial sarcoma.

Following a multidisciplinary oncology board and after obtaining the informed patient consent, we proceeded in performing a total laryngectomy with bilateral cervical lymphadenectomy, under general anesthesia with tracheal intubation.

The retrograde laryngeal resection began with a Glück-Sorensen incision, followed by cervical flap dissection, neck dissection and cervical lymph node dissection (Figures 1 and 2). The larynx is eventually lifted, from the bottom up,

after separating it from the trachea (Figure 3). The new tracheostomy is created. Thus, after the larynx has been separated from the hypopharynx, we continued by closing the anterior pharyngeal and esophageal wall through sutures.

Figure 1 and 2 – Left (1) and right (2) cervical lymph node dissection

Figure 3 – Removal of the larynx

The surgical technique involved modified radical dissection of the ipsilateral cervical lymph nodes and functional dissection of the contralateral nodes (Figure 1, 2). Modified radical node dissection type III included lymph node excision of all lymph node levels I-VI, whilst preserving the non-lymphatic structures (internal jugular vein, accessory spinal nerve and sternocleidomastoid muscle). No cervical lymph node metastasis has been identified.

Upon opening the removed larynx, the macroscopic endolaryngeal mass was exposed (Figure 4) showing a violaceous, bilobed tumor with a corneous surface; its point of insertion located within the laryngeal ventricle, the petiole (root) of the epiglottis and the ventricular folds commissure. Histopathological examination of the tumor and lymph nodes, together with local examination, indicated T3N0M0.

Figure 4 – The removed larynx with endolaryngeal mass

Evolution of patient in postoperative period was favorable, under prescribed antibiotics, pain relievers, antisecretory agents, anticoagulant and intravenous balancing solutions.

The patient was recommended to the oncology department and underwent radio-chemotherapy, with 30 radiotherapy sessions and 6 of chemotherapy, both with favorable results.

Patient has checked in for follow-up, every 3 months over the past 2 years, with no macroscopic signs of tumoral relapse.

Discussions

A search of literature is suggestive for the very low number of cases, accounting for less than 1% of head and neck malignancies [8]. Most published articles on the matter are suggestive for the bimodal distribution of age groups, our case coming in towards the end of the second incidence peak.

A literature review of laryngeal synovial sarcomas summarizes 39 cases, reported up until 2019, with the management remaining a subject of debate. Surgical excision adapted to the tumor extension represents the treatment of choice [7].

Multimodal treatment is governed by surgery, often combined with chemotherapy and radiotherapy. Depending on the stage of the tumor at presentation, one can opt for either CO₂ laser transoral surgical technique, or an external surgical technique such as total laryngectomy in cases involving massive tumors, recurrence or positive tumor margins after an endoscopic procedure. In our case the patient presented to the emergency room, with acute respiratory failure that required emergency tracheotomy and following the CT scan aspect and the biopsy result, the surgery indication was for total laryngectomy.

The involvement of a multidisciplinary team in the treatment of laryngeal malignancies is essential in regard to both the oncological approach and the rehabilitation of phonatory function. The treatment plan is discussed and established by a team comprising of an ENT surgeon, an oncologist, a pathologist, a radiation therapist and a nutritionist, with the options forwarded to the patient and family, so they may partake in treatment decision [9].

Total laryngectomy is a mutilating intervention, which affects the patient's quality of life but also the socio-professional and family reintegration. Consequently, the postoperative care of the patient consists of a multidisciplinary team that in addition to the ENT surgeon includes the vocal therapist and the psychologist. Given the incidence of laryngeal synovial sarcoma among young patients, the impact of the disease is even more important [10].

The vocal rehabilitation methods available to the laryngectomized patient are comprised of the following: learning to use the esophageal voice (as in our case), the laryngophone (external laryngeal amplifier) or opting for a phonatory implant/vocal prosthesis (communication through a surgically constructed tracheo-esophageal puncture) [11-13].

Conclusion

Biphasic synovial sarcoma located in the larynx is extremely rare, with literature data providing information on some cases, without studies in this direction.

Laryngeal synovial sarcoma is a high malignancy soft-tissue tumor, with a poor prognosis, relating to the degree of infiltration, resectability and the presence of metastasis (usually involving the lungs).

The gold standard consists of surgical treatment, combined in selected cases with chemotherapy and/or radiotherapy. Long-term follow-up is mandatory for recurrence prevention.

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