

Atypical carcinoid of larynx and management using transoral robotic surgery: Supraglottic laryngectomy

ABSTRACT

Atypical carcinoid tumor of the larynx is a particularly rare occurrence. Here, we report a patient with a laryngeal neuroendocrine tumor (NET) who was treated with transoral robotic surgery-supraglottic laryngectomy (TORS-SL). A 73-year-old male patient, a known smoker, presented with throat pain and hoarseness for 1 month. Endoscopic laryngeal examination and magnetic resonance imaging revealed a left supraglottic mass. The results of histopathologic examination and immunohistochemical analysis were consistent with atypical carcinoid tumor of the larynx. A TORS-SL was performed. There was no recurrence during a follow-up period of 18 months. NETs of the larynx are a very heterogeneous group of tumors in terms of pathological features and biological behavior. The histological diagnosis is the most important step in the management of these tumors. Tumor histology is an important determinant of prognosis. TORS-SL is a promising, minimally invasive, endoscopic surgical alternative for the treatment of supraglottic tumors with good functional and clinical outcomes as seen in this case.

Key words: Atypical carcinoid; neuroendocrine tumors of larynx; transoral robotic surgery-supraglottic laryngectomy.

Introduction

Laryngeal cancer accounts for approximately 2-5% of head and neck malignancies.^[1,2] Squamous cell carcinomas account for more than 90% of laryngeal cancers, and it is the most common histological finding. Neuroendocrine tumors (NETs) are the most common nonsquamous types of neoplasm arising in the larynx and represent <1% of all primary laryngeal tumors.^[3]

The first laryngeal NET was reported in 1969 by Goldman *et al.*^[4] These tumors have a predilection for supraglottis. The median age at diagnosis ranges between 50 and 70 years, and the most important risk factor is smoking.

Organ preservation regimen such as radiotherapy and concurrent chemoradiotherapy has been advocated for treatment of laryngopharyngeal head and neck cancer to avoid the morbidity associated with open head and neck surgical approaches. However, they can result in organ

dysfunction, tracheostomy, feeding tube dependence, and poor quality of life.

New modalities like transoral, minimally invasive organ preservation surgeries have been more frequently used and have demonstrated comparable disease control and promising functional and quality of life outcomes.^[5] After the first description of transoral robotic surgery (TORS) by Weinstein *et al.*,^[5] this application gained widespread use as another minimally invasive way of treating a variety of laryngopharyngeal, especially oropharyngeal carcinomas. Here, we report on a patient with a laryngeal NET, who was treated with TORS-supraglottic laryngectomy (TORS-SL).

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Case Report

A 73-year-old male patient, a known smoker, presented with throat pain and hoarseness for 1 month. Endoscopic examination revealed a large supraglottic mass involving left aryepiglottic fold [Figure 1]. The mobility of vocal cords was normal. Examination of neck found no palpable lymphadenopathy. Laryngoscopic biopsy of the mass was suggestive of atypical carcinoid. Magnetic resonance imaging showed the presence of supraglottic mass (1.7 cm × 1.7 cm in size), involving left aryepiglottic fold, left pyriform sinus, and few sub centimeter short axis lymph nodes in bilateral submandibular and deep cervical region. The patient underwent TORS-SL and selective neck dissection (level II-V). Robotic setup time was 8 min with total surgical duration of 50 min and minimal blood loss (<30 ml). The patient did not require tracheostomy or percutaneous endoscopic gastrostomy tube insertion. Oral feeding was started on postoperative day 10 with no peri or postoperative complications. The patient had good swallowing and speech outcome at week 6 postoperatively. Based on the histopathological examination of the surgical specimen, a diagnosis of atypical carcinoid (moderately differentiated NET) was made. All margins were reported negative. Immunohistochemistry revealed expression of calcitonin, chromogranin, and CD56, and negative for thyroid transcription factor, cytokeratin (CK) [Figure 2a-c]. Metastatic carcinoma was present in 2 of 25 lymph nodes. The patient did not require adjuvant treatment and is disease-free after 18 months of follow-up.

Discussion

NETs of the larynx are a rare entity. These are classified into the following subtypes - typical carcinoid (well-differentiated NET,



Figure 1: Endoscopic view depicting a large supraglottic mass involving left aryepiglottic fold

Grade I), atypical carcinoid tumor (moderately differentiated carcinoma, Grade II; large cell neuroendocrine carcinoma), small cell neuroendocrine carcinoma (poorly differentiated neuroendocrine carcinoma, Grade III), and paraganglioma.^[3]

These tumors present as subepithelial lesions without any ulceration of the mucosa. Histologically, cells are arranged in cords, nests, trabeculae, or glandular patterns. Neoplastic cells are larger, and nuclei are often vesicular and contain prominent nucleoli. The tumors are positive marker (in particular calcitonin and somatostatin), and low molecular weight CK as well as other epithelial markers like carcinoembryonic antigen and epithelial membrane antigen.^[3]

Atypical carcinoid tumor in the larynx is very uncommon with a male:female ratio of 3:1. The 5-year survival rate for atypical carcinoid of the larynx is 48.7%.^[6] The prognosis of atypical carcinoid tumor is poorer than that of typical carcinoid tumor.^[3] The treatment of choice for atypical carcinoid tumor of the larynx is surgical excision.^[7] As most tumors are supraglottic in location, supraglottic laryngectomy is often performed.^[8] The usual sites of metastatic involvement are lymph nodes, bones, skin, liver, and lung. Therefore, elective neck dissection is indicated.^[3]

Recent studies have cited potential advantages of TORS over traditional treatment options, including avoidance of the external incision, improved access and visualization, preservation of uninvolved structures, and shorter hospital stay.^[9,10] The main advantage of this approach compared to the traditional approaches is the preservation of organ structure and function. Fewer patients require tracheostomy tubes with the quicker rehabilitation of swallowing and vocal function.^[10-12] Similar to the findings of TORS in other subsites, TORS supraglottic laryngectomy also resulted in minimal

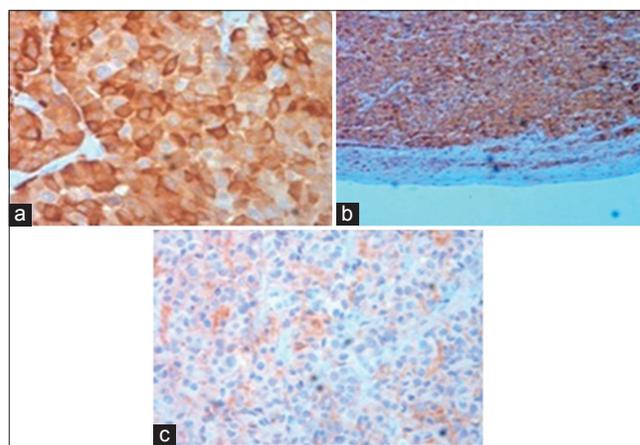


Figure 2: Immunohistochemistry revealing positivity for calcitonin (a), chromogranin (b), and CD56 (c)

estimated blood loss, acceptable robotic setup, and relatively short operative times and length of stay.

Conclusion

NETs of the larynx are a very heterogeneous group of tumors in terms of pathological features and biological behavior. The histological diagnosis is an important determinant of prognosis and also in the management of these tumors. Unfavorable prognostic factors include tumor diameter >1 cm, palliative surgery, evidence of distant metastases, particularly in the skin or subcutaneous tissue.^[5,6]

TORS-SL is a promising, minimally invasive, and endoscopic surgical alternative for the treatment of supraglottic tumors with good functional and clinical outcome. Further studies are indicated with an additional clinical follow-up to investigate long-term functional and oncological outcomes.

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Conflicts of interest

There are no conflicts of interest.

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