Case Report / Olgu Sunumu

Laser excision of a typical carcinoid tumor of the larynx: a case report

Tipik larenks karsinoid tümörünün lazer eksizyonu: Olgu sunumu

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Neuroendocrine carcinomas of the larynx include a range of rare tumors which have variable biologic behavior, affecting treatment and prognosis. Among these, typical carcinoid tumors are the least common type. Prognosis of typical carcinoid tumor is better than atypical carcinoid tumor and small cell carcinoma of the larynx. Conservation surgery is the preferred treatment modality. Transoral CO_2 laser surgery can be a good alternative for appropriate cases because of the functional results and less morbidity. In this article, a 71-year-old female presented with complaints of feeling a mass during swallowing. Fiberoptic examination of the larynx revealed a mass located on the right aryepiglottic fold and biopsy revealed the tumor as a typical carcinoid tumor. We describe CO_2 laser excision of a typical carcinoid tumor of the larynx in this case report.

Key Words: Carcinoid tumor/diagnosis/surgery; laryngeal neoplasms; laser therapy; paraganglioma/diagnosis/ surgery.

Larenksin nöroendokrin karsinomları nadir görülmekle birlikte değişken biyolojik davranışları nedeniyle tedavi ve prognozları farklıdır. Bunlar arasında tipik karsinoid tümörler en sık görülen tipidir. Larenksin tipik karsinoid tümörünün gidişatı atipik karsinoid tümöre ve küçük hücreli karsinoma göre daha iyidir. Konservatif cerrahi tercih edilen tedavi yöntemidir. Transoral CO_2 lazer cerrahisi, fonksiyonel sonuçları ve morbiditenin düşük olması nedeniyle uygun olgularda iyi bir seçenek olabilir. Bu yazıda, yutkunurken boğazında kitle hissi yakınması ile başvuran 71 yaşında kadın hasta sunuldu. Fiberoptik muayenesinde sağ ariepiglottik (pilika) yerleşimli kitle ile karşılaşıldı ve alınan biyopsi sonucu tipik karsinoid tümör olarak bildirildi. Bu olgu sunumunda larenks yerleşimli tipik karsinoid tümörün CO_2 lazer ile çıkarılması bildirildi.

Anahtar Sözcükler: Karsinoid tümör/tanı/cerrahi; larenks tümörleri; lazer tedavisi; paragangliyom/tanı/cerrahi.

Carcinoid tumors arise from the enterochromaffin (Kulchitsky) cells, part of the APUD (amine precursor uptake and decarboxylation) system.[1] They are thus found in sites where these cells are widely distributed: the lungs, bronchi and gastrointestinal tract. The larynx is an infrequent site of origin.[1-6] They make up just a little less than 1% of all laryngeal neoplasms.[1,5,6] The World Health Organization (WHO) described four different neuroendocrine tumor types: typical carcinoid, atypical carcinoid, small cell carcinoma, and paraganglioma. Among these, atypical carcinoid tumors of the larynx

Received / *Geliş tarihi:* June 11, 2010 Accepted / *Kabul tarihi:* August 8, 2010

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(moderately differentiated neuroendocrine tumors) are the most common, typical carcinoid tumors are least common. These tumors represent a heterogeneous group of disorders with different prognosis and rationales of management. The prognosis of typical carcinoid tumors is favorable and the biological behavior is more indolent than atypical carcinoid tumors and small cell neuroendocrine tumors.[1,2,5,6] Furthermore they do not metastasize and are less sensitive to irradiation.[6] Therefore the preferred treatment is limited excision with preservation of laryngeal function as much as possible. Preferably ultrasound can be used for evaluation of neck lymph nodes. Elective treatment of the N0 neck is not necessary, as the likelihood of cervical lymph node metastasis is not comon.[1,5,6]

CASE REPORT

A 71-year-old female presented with complaints of feeling a mass during swallowing for two years. She was a chronic smoker since 35 years and she drank 1-2 glasses of alcohol every day. There were no complaints of dysphagia, odynophagia or respiratory distress.

Fiberoptic examination of larynx revealed a mass located on the right aryepiglottic fold (Figure 1). The vocal cord movement was normal. On examination under general anesthesia the tumor was a firm, 1.5 cm well-defined mass on the right aryepiglottic fold. Arytenoid cartilage and ventricle were free of tumor. The biopsy taken identified the tumor as typical carcinoid tumor, with low grade features. Octreotid scan was performed to rule out spread of tumor and no additional sites were identified (Figure 2).

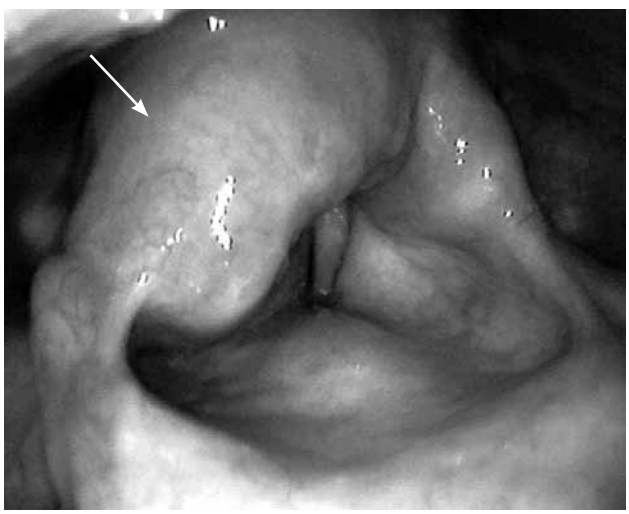

Figure 1. *Submucosal mass in the right aryepiglottic fold.*

Given the location of the tumor the least invasive method allowing adequate tumor excision was decided to be an endoscopic laser excision. The tumor mass was divided along the midline with laser to observe the depth of invasion and plan the resection margins. The tumor was identified not to be extending to the ventricle and the arytenoid cartilage. It was thus possible to preserve vocal cords and arytenoid cartilage. The tumor was removed in three pieces including margins of at least 2 mm of normal-looking tissue. The aryepiglottic fold and false vocal cord were included in this excision. Biopsies were taken from the resection margins to assure adequate removal. Histopathological evaluation confirmed the diagnosis of a completely removed low grade typical carcinoid tumor.

No tracheotomy was performed. The patient was discharged home after one day of observation. However, because of swallowing problems with aspiration detected by videoflouroscopy, one week after the surgery a nasogastric feeding tube was placed as an outpatient procedure. Ten days later another videofluoroscopy revealed no aspiration. The feeding tube was removed and oral intake resumed. One month after the operation a second look operation was performed under general anesthesia. The biopsies were tumor-free.

DISCUSSION

The 1991 WHO (World Health Organization) classification of laryngeal tumors divided neuroendocrine neoplasms into carcinoid, atypical carcinoid, small cell carcinoma, and paraganglioma. Neuroendocrine tumors of the larynx are very rare.[1,3,6] No more than 700 cases have been reported in the literature. The atypical carcinoid tumor is most common, while typical carcinoid is least common.[6]

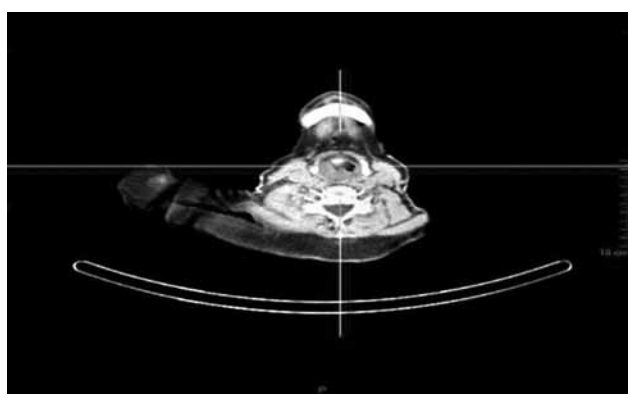

Figure 2. *Octreotide-computed tomography scan showing uptake by the tumor located on the right aryepiglottic fold.*

The atypical carcinoid tumors are poorly differentiated with a more aggressive course than typical carcinoids. Histologically, the atypical carcinoid tumor is characterized by increased mitotic activity and more necrosis. Classification is very important to distinguish between the different behaviors of these tumors. Metastatic lymph nodes in the neck are very common. Soga et al.[7] described finding metastases in 66.7% of 199 cases of atypical carcinoid tumor of the larynx. Apart from regional metastases, atypical carcinoid can metastasize to the skin and subcutaneous tissues. Lung, bone, liver, heart, peritoneum and brain are other places of metastasis.[1] Death occurs due to distant metastases. The survival rate is between 40 and 50% at five years and 30% at 10 years. Soga[8] reported that the five-year survival for atypical carcinoid tumor was 47%. Treatment is usually supraglottic or total laryngectomy depending on the site and stage of the tumor. In the literature, only few cases have been treated with CO_2 laser surgery. Ferlito et al.[1,3,6] advise elective bilateral selective neck dissection (levels II-IV, preserving sublevel IIB) in N_0 cases and radical or modified radical neck dissection for N^+ necks. Responses to chemotherapy and radiotherapy are less well documented and their use are controversial in cases without metastasis.

Small cell neuroendocrine tumors of the larynx show similar features to lung small cell carcinoma.[1,6] Prognosis is poor and metastasis occurs early. Various paraneoplastic syndromes are common. Treatment of small cell neuroendocrine carcinoma is similar to pulmonary small cell carcinoma, using systemic chemotherapy and radiotherapy.[1,6] Surgery is seldom indicated and two and five-year survival rates are 16 and 5%, respectively.[1,6]

The biological behaviour of paraganglioma of the larynx is benign and the prognosis is excellent. This tumor shows a female preponderance, with a male-female ratio of 1:3. Only one metastatic case has been reported in the literature who developed a metastasis to the lumbar spine 16 years after diagnosis of the tumor in the larynx.[1,2,6] Generally, this tumor is characterized by a submucosal mass and treated with partial laryngectomy. One case has been reported with an advanced laryngeal paraganglioma treated successfully with transoral CO_2 laser microsurgery.[1]

Typical carcinoid tumors occur predominantly during their 6^{th} and 7^{th} decades of life in men who have been heavy smokers. Presenting symptoms are dysphagia, odynophagia, feeling of a lump in the throat, hoarseness, otalgia and hemoptysis.[1,2,6] Our patient was 71-years-old female, also developed complaints of feeling of a lump in her throat for approximately two years. The commonest site is the supraglottic larynx, with a predilection for the aryepiglottic fold.[9] They may be polypoid, pedunculated, or nodular and are often submucosally located.[1,6]

Soga et al.[7] observed that 33% of 42 patients with typical carcinoid of the larynx also developed metastases. Batsakis et al.[10] reported distant metastases in four of 13 (31%) reported cases of typical carcinoid.[1,2,4] We used octreotid scan for staging and found no metastasis.

Typical carcinoid tumor of larynx can be treated with conservative surgical excision. Total laryngectomy should be reserved for extensive tumors only. Radiotherapy and chemotherapy are reported to be not effective.[1,3,5,6] Our patient's tumor size was 1.5 cm, so we made an attempt for transoral CO_2 laser microsurgery because of the expected better functional result.[11,12] Starting the excision by splitting the tumor into half, enables a better visualization of the depth of invasion and has enabled us to preserve the vocal cord and arythenoid cartilage.

Elective neck dissection is not indicated if lymph node metastases are not detected by ultrasound. The five-year survival rate for typical carcinoid of the larynx is 50%. The low survival rate might partly be due to the fact that some cases of atypical carcinoid tumor are sometimes erroneously classified as typical carcinoid.[1,6]

Laser surgery has a lower incidence of complications (permanent tracheotomy, gastrostomy) and better functional results compared with open surgical techniques and therefore can be used in early supraglottic cancer.[11] Karatzanis et al.[11] found endoscopic laser surgery complication rates (10.2%) to be lower than horizontal laryngectomy. Hinni et al.[12] have reported complication rates of transoral CO_2 laser microsurgery for 117 advanced laryngeal carcinoma patients and in their study only two patients had a permanent tracheotomy and five patients had permanent feeding tubes. Peretti et al.[13] have reported on 595 glottic laryngeal carcinomas treated by laser, in their study no patient needed a permanent tracheotomy and in only one patient was a feeding tube needed for five days. A feeding tube was

necessary in our patient to prevent aspiration and no tracheotomy was needed.

In conclusion, laryngeal neuroendocrine tumors are extremely rare. Treatment of these tumors are very different due to biologic behaviors. CO_2 laser surgery can be a good alternative for selected cases because of good functional results and less morbidity. Especially in typical carcinoids transoral CO_2 laser microsurgery is the preferred method to avoid long term morbidity associated with external surgery.

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