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Case Report

Case report of atypical carcinoid tumors of the larynx

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ABSTRACT

Neuroendocrine tumors (NET) of the larynx are considered one of the rare disease; they represent only 1% of the laryngeal tumors. These tumors are very aggressive with a high rate of metastases. Diagnosis is done by computed tomography and confirmed by histopathological examination. Neuroendocrine tumors can be classified into four types based on histopathology: typical carcinoid tumors, atypical carcinoid tumors, small cell neuroendocrine tumors and paragangliomas. Atypical carcinoid tumor of the larynx is considered rare in occurrence. We report a rare laryngeal neuroendocrine tumor case.

Keywords: Carcinoid, Atypical, Larynx

INTRODUCTION

Laryngeal neuroendocrine tumors are extremely rare and counts <1% of all primary laryngeal tumors.[1,2] The neuroendocrine tumors (NET) can spread throughout several organ systems, but most commonly to the lungs and digestive system. Many classifications have been developed for neuroendocrine neoplasms of the larynx but the most common classification of neuroendocrine tumors subdivided them into four major types, paraganglioma, Small cell carcinoma, Atypical carcinoid, and typical carcinoid (TC).[3,4] Atypical Carcinoid tumors of the larynx are considered one of the rare tumors; they are only around 200 cases in the literature.[5,6] Atypical carcinoid is located most commonly in the supraglottic region; most patients are males and smoker in the sixth-to-seventh decade of life. Most commonly, patient present with hoarseness, dysphagia, odynophagia and dyspnea. Surgical excision is considered as the definitive treatment for laryngeal atypical carcinoid tumor. Partial or total laryngectomy may be indicated taking into account the site and the extent of the primary tumor.[7] In this paper, we present a rare and interesting case of a nonsmoker female patient with atypical carcinoid tumor of the larynx.

CASE REPORT

A 69 year old female, known case of hypertension, hyperthyroidism, presented to the hospital with history of dysphagia, hoarseness and odynophagia for two-month duration. She is nonsmoker or Alcoholic. Her medical history was unremarkable apart from using anti reflex medications for her GERD. Family history is unremarkable. On physical examination, the patient appeared well. Otorhinolaryngological exam was normal apart from fiberoptic laryngoscopy, which revealed a fungating mass arising from the right aryepiglottic fold extended to the ventricles, the mobility of the vocal cord was normal. Neck exam unremarkable with no palpable

lymph nodes or masses. Systemic exam were unremarkable. Her laboratory workup was normal. Computer tomography (CT) of the head and neck was performed and showed large supraglottic mass reaching the vocal cords. It demonstrated large moderately enhancing mass of the epiglottis causing obstruction of the air passage. There is significant effacement of the valleculae bilaterally and distortion of the oropharynx (It may be noted that the distortion of oropharynx is also complemented by the highly tortuous left internal carotid artery. The aryepiglotc folds are unidentifiable. Inferiorly, the mass extend to reach the vocal cords, which are showing disturbed configuration, especially the right appears to be more laterally displaced and the left appears to be relatively medial in location with arytenoid cartilage. Pre epiglottis space appears to be infiltrated (Figure 1 and 2).

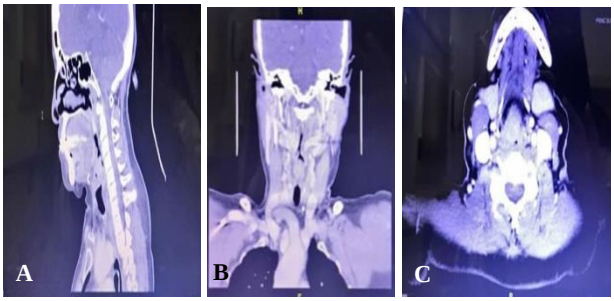

Figure 1: CT scan of the head and neck figures.

A and B: It demonstrated large moderately enhancing mass of the epiglottis causing obstruction of the air passage. There is significant effacement of the valleculae bilaterally and distortion of the oropharynx (It may be noted that the distortion of oropharynx is also complemented by the highly tortuous left internal carotid artery. The aryepiglotc folds are unidentifiable; C: Inferiorly, the mass extend to reach the vocal cords, which are showing disturbed configuration, especially the right appears to be more laterally displaced and the left appears to be relatively medial in location with arytenoid cartilage. Pre epiglottis space appears to be infiltrated.

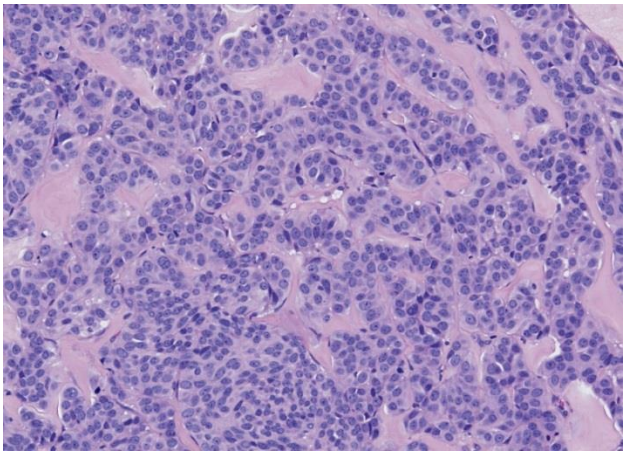

Figure 2: Monomorphic cells arranged in trabecular pattern.

The cells show vesicular nuclei, inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm.

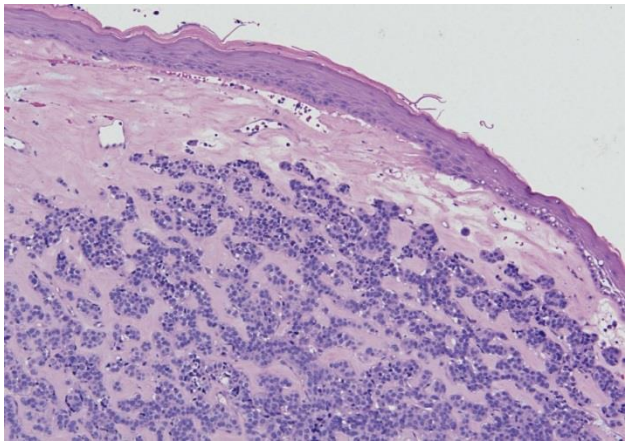

Figure 3: Sub epithelial proliferation of monomorphic cells.

Initial histopathological examination of supraglottic mass revealed well-differentiated type neuroendocrine carcinoma. It showed Monomorphic cells arranged in trabecular pattern, the cells show vesicular nuclei, inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm (Figure 3). Wide local excision of the right aryepiglottic mass was performed with KTP laser, histopathological exam of the surgical specimen was done and it revealed that tumor has infiltrative growth pattern and there is perineural invasion, therefore it is consistent with atypical carcinoid, no necrosis identified. It is also strongly immunoreactive to CK, chromogranin and synaptophysin while not reactive to S100. Therefore, histopathological diagnosis was atypical carcinoid tumor, which is moderately differentiated neuroendocrine carcinoma. Postoperatively, the patient did not receive radiation therapy. At follow up 2 years later, the patient was symptoms free and her voice quality was good.

DISCUSSION

Laryngeal neuroendocrine neoplasms are extremely rare and represent <1% of all primary laryngeal tumors. Carcinoid tumors were first recognized in 1907 by Oberndorfer.[7] who distinguished them from other carcinomas. In 1959, Blanchard and Saunders reported the first description of laryngeal neuroendocrine tumor.[9] Since then, over 500 cases have been reported. They are usually found in the gastrointestinal tract (55%) or the bronchopulmonary tract (10%).[8] Atypical carcinoma of the larynx occurs in more than 90% of the cases in the supraglottic area, prevailing on the aryepiglottic fold, arytenoids and larynx side of epiglottis. It is more common in males (3:1 ratio), especially smokers. Our patient was female, 69 years old, though in the literature the incidence occurs more commonly in smoker males.[9] The symptoms depend on the site of the lesion. Patients commonly present with dysphagia and dysphonia, as in our patient. Glossopharyngeal neuralgia can occur before tumor being clinically clear and the current patient did report pain.[10] Computed tomography of the larynx is

useful for evaluating local and regional tumor spread. Microscopy of atypical carcinoid tumor shows perineural invasion and mitioc figure less than 2/10 HPF which also occur in typical type. Though, besides typical type present better prognosis, they do not present positive signs in the immunohistochemical analysis for CK, chromogranin and synaptophysin and the diagnosis was not questionable due to perineural invasion. The surgical excision is the mainstay of treatment for laryngeal atypical carcinoid tumor. Depending on the site and extent of the primary tumor Partial or total laryngectomy may be indicated. As most tumors are located in supraglottic region, supraglottic laryngectomy is often the procedure of choice for supraglottic location. And that what was done for the current patient with KTP laser. Elective neck dissection is indicated due to high incidence of early cervical lymph node metastasis.[11] Once again, our patient did not have any cervical lymph nodes involvement. Radiotherapy or chemotherapy is not effective in this type of tumor.[11,12]

CONCLUSION

We report a 69 years old female with a rare atypical carcinoid tumor of the larynx**,** which is managed successfully through wide local excision with KTP laser. No recurrence in 2 year follows up.

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