

## Case Report

# A case of laryngeal paraganglioma and literature review

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**Abstract:** Paragangliomas are common in carotid body, jugular tympanic cavity and cervical vagus nerve, but uncommon in larynx. We report clinical and pathological features of a 40-year-old woman with laryngeal paraganglioma. Tracheotomy combined with vein anesthesia was performed to isolate tumors. The patient was followed up for 3 months after surgery treatment and recovered well. We also provide a better understanding of this disease through a review of relevant literature. Laryngeal paraganglioma is rare, but should require long-term follow-up for possible recurrence.

**Keywords:** Paraganglioma, larynx, diagnosis, prognosis

### Introduction

As a part of parasympathetic nervous system, parasympathetic ganglion is composed of neuroendocrine cells [1]. Pheochromocytoma arises from adrenal medulla, while paraganglioma is a kind of extra-adrenal tumors [2]. There are two kinds of paragangliomas: sympathetic and parasympathetic paragangliomas. Paragangliomas are benign tumors, accounting for 0.6% of head and neck tumors. They are common in carotid body, jugular tympanic cavity and cervical vagus nerve, but uncommon in larynx [2, 3]. Our article aims to present a case of laryngeal paraganglioma (LP) and review relevant literatures.

### Case report

A 40-year-old woman presented to our hospital with hoarseness and throat discomfort for 4 years. She reported excessive phlegm, sleep snoring, and occasional suppress wake. On physical examination, a dark red, smooth neoplasm at the inside of left arytenoid blocked the glottis. The left vocal cord was dark red. The right vocal cord, ventricular band and arytenoid were unremarkable. Bilateral vocal cord was good in the movement amplitude, but poor in closure. On electronic laryngoscope exami-

nation, a smooth neoplasm was found in the left arytenoid. An angiomatoid neoplasm was found in the laryngeal by enhanced CT (**Figure 1**). The patient received tracheotomy combined with vein anesthesia in Nov 2014. Tumor was isolated from the left side of pharyngeal, the size of which was 3×2.5×2 cm. A immunohistochemistry revealed positive for expression of CD56, chromogranin A (CgA), SYN, P63, S-100, Vimentin, CD34, CD31, but negative for expression of Calponin, CK5/6, Actin, SMA, HMB45, Melan-A, IV collagen (**Figure 2**). The patient was diagnosed with LP and recovered well after 3 months follow-up.

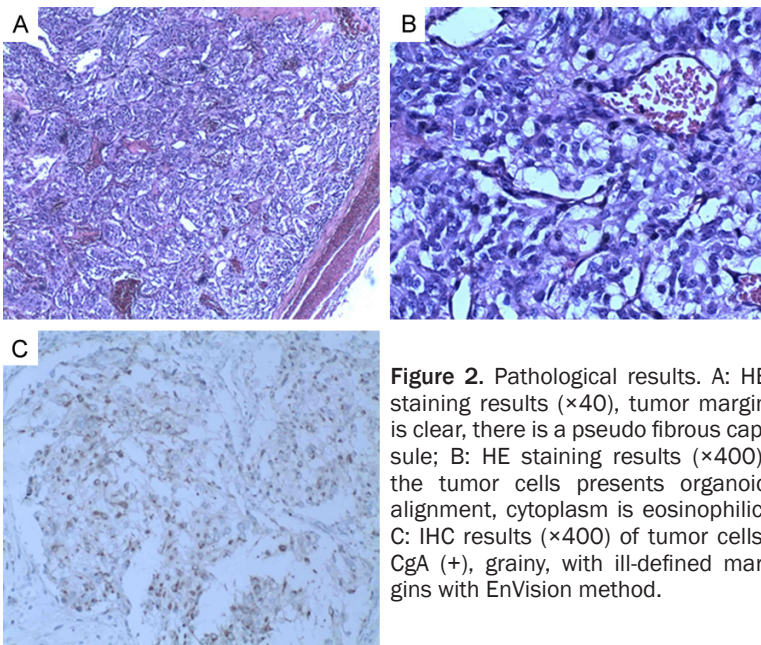
### Discussion

There are two pairs of laryngeal paraganglia. Superior laryngeal paraganglia locates in the edge of thyroid cartilage. While inferior laryngeal paraganglia lies in any part from inferior cornu of the thyroid cartilage to cricoid cartilage. The superior laryngeal paraganglia tissue accounts for 90% of LPs [4]. LP is divided into two types in clinic. Type I primarily presents with hoarseness [4], while type II may lead to wheezing sound and hemoptysis [5].

LP is the only one of laryngeal neuroendocrine tumors occurred in women and the ratio of



**Figure 1.** The results of contrast-enhanced CT on larynx: a significantly enhanced block shadow is seen on left side of throat with size of 2.4×2.9×3 cm, no obvious abnormalities found in the rest parts.



**Figure 2.** Pathological results. A: HE staining results (×40), tumor margin is clear, there is a pseudo fibrous capsule; B: HE staining results (×400), the tumor cells presents organoid alignment, cytoplasm is eosinophilic; C: IHC results (×400) of tumor cells: CgA (+), grainy, with ill-defined margins with EnVision method.

[2]. Moreover, LP rarely shows tissue necrosis and vascular invasion, and the canceration rate is 2% [6]. In our case, the patient reported hoarseness and uncomfortable throat. Mucous membrane is penetrated in tumor biopsies, resulting in the risk of tumor bleeding. Therefore, the patient was received intraoperative frozen. Enhanced CT, MRI and angiography are preoperative necessary to assess the location, the relationship with the surrounding tissues and organs, and malignant change of tumors [7]. Immunohistochemistry is an important method for distinguishing LP from laryngeal cancer, angiosarcoma, and neuroendocrine cell carcinoma [8]. LP is typical cell sphere structure, also known as Zell-ballen structure, which shows tumor cell nested bubble pulse constructed by primary and supporting cells and surrounded with vascular fibrous stroma. In this case, the patient was diagnosed with supraglottic LP by enhanced CT and immunohistochemistry. It was observed that oval, reddish brown, and flexible tumor, coated with incomplete capsule. The diameter of tumor was about 2.5 cm.

Operation is major treatment for LP, including removal under endoscopic or via external carotid pharyngeal. The latter is appropriate for supraglottic LP, in which thyroid artery is ligated to reduce bleeding [9]. While endoscopic surgery may lead to uncompleted tumor resection and risk of hemorrhage [10]. In

women to men is 3 to 1 [6]. The location and size of tumors represent clinical characteristic of LP, and hoarseness is a major symptom of LP

our case, the patient was treated via external carotid pharyngeal for large extension, deep tissue invasion and rich blood supply of tumors.

Radiotherapy is used as a major or auxiliary treatment under special situation, such as malignant tumor with intracranial metastases, multi-site paraganglioma and patients unsuitable for surgery [11]. Complete resection of tumor is the key to prevent recurrence and improve survival. Radiotherapy may be a better choice to delay tumors growth those are large volume or exist in special areas. A post-operation follow-up is necessary for LP. The recurrence rate is 17% and local recurrence may be a sign of malignancy [11]. The patient recovered well after 3 months follow-up in our case.

## Disclosure of conflict of interest

None.

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