MEDICAL FORUM

LARYNGEAL PARAGANGLIOMA: A CASE REPORT

Le Van Khang*, Vu Dang Luu*, Nguyen Thi Ngoc*, Nguyen Thi Thu Hang**

SUMMARY

Paragangliomas correspond to tumors of neuroendocrine origin of chromaffin and non-chromaffin in the paraganglia of the sympathetic or parasympathetic nervous system. Laryngeal and thyroid paragangliomas that arise from the laryngeal paraganglia are extremely rare. Diagnosing two kinds of paragangliomas preoperatively is challenging due to non-specific cytology, pathology, and imaging features, and overlaps with many other neoplasms. Complications in surgery of laryngeal and thyroid paragangliomas may be associated with significant intraoperative bleeding and excision with adherence to nearby structures, including the recurrent laryngeal nerve.

Keywords: *nonfunctional paraganglioma, supraglottic paraganglioma, laryngeal paraganglioma, thyroid paraganglioma, head and neck paraganglioma.*

I. INTRODUCTION

Laryngeal and thyroid paragangliomas that arise from the laryngeal paraganglia are rare compared to the more common locations of the carotid body, vagal, jugular, and tympanic paragangliomas. The laryngeal paraganglia give rise to both laryngeal and thyroid paragangliomas, comprising both superior and inferior paraganglia [1]. The superior laryngeal paraganglia are located near the superior edge of the thyroid cartilage, near the larynx entrance and the epiglottis. They are fairly constant in location, related to the internal branch of the superior laryngeal nerve. The vast majority of laryngeal paragangliomas are supraglottic in location, arising from the superior laryngeal paraganglia. The inferior laryngeal paraganglia are more variable in location, close to the recurrent laryngeal nerves at the level of the cricoid cartilage and first tracheal ring, and may occur within the thyroid gland [2–4]. Inferior laryngeal paraganglia may give rise to paragangliomas in the thyroid gland, subglottic larynx, or upper cervical trachea [5–7]. These lesions may be associated with significant intraoperative bleeding and complicated excision with adherence to nearby structures, including the recurrent laryngeal nerve. Diagnosing two kinds of paragangliomas preoperatively is challenging due to non-specific cytology, pathology, and imaging features, and overlaps with many other neoplasms.

Case report

A 42-year-old male patient discovered a mass in the right neck area. Clinical diagnosis is a tumor in the right carotid triangle. Ultrasound images demonstrated a hypoechoic mass lying next to the common carotid artery's inner border. The mass was well-defined, hypervascular on color Doppler and its size was up to 15x31 mm.

** Department of Radiology, Hanoi Medical University, Hanoi, Vietnam*

*** Radiology Center, Bach Mai Hospital, Ha Noi, Vietnam*

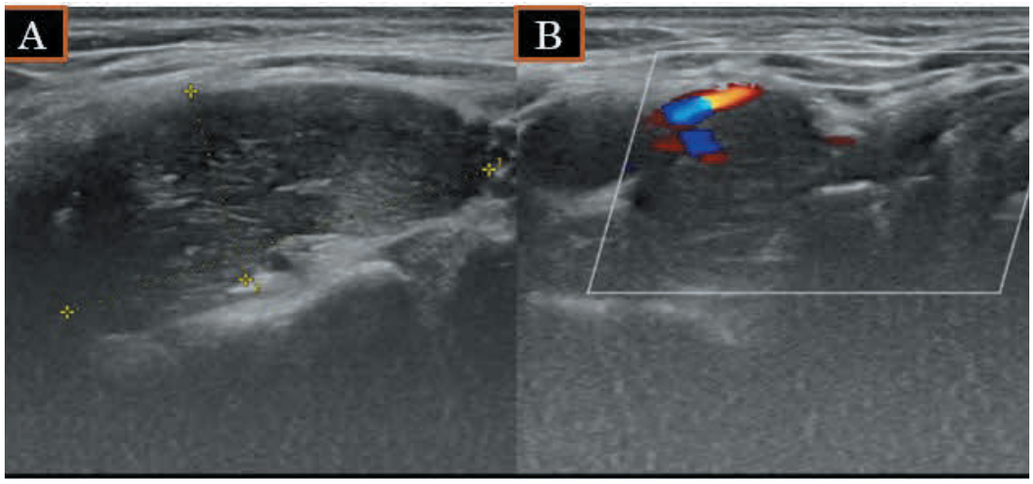

Figure 1. The grayscale (A) and color Doppler (B) ultrasound images demonstrated a hypoechoic mass that lay next to the common carotid artery's inner border and hypervascularity

On CT scan, the mass located medially to the right carotid sheath, below the bifurcation of the common carotid artery, had well-defined borders and enhanced strongly after injection. This mass was fed by both superior and inferior thyroid arteries.

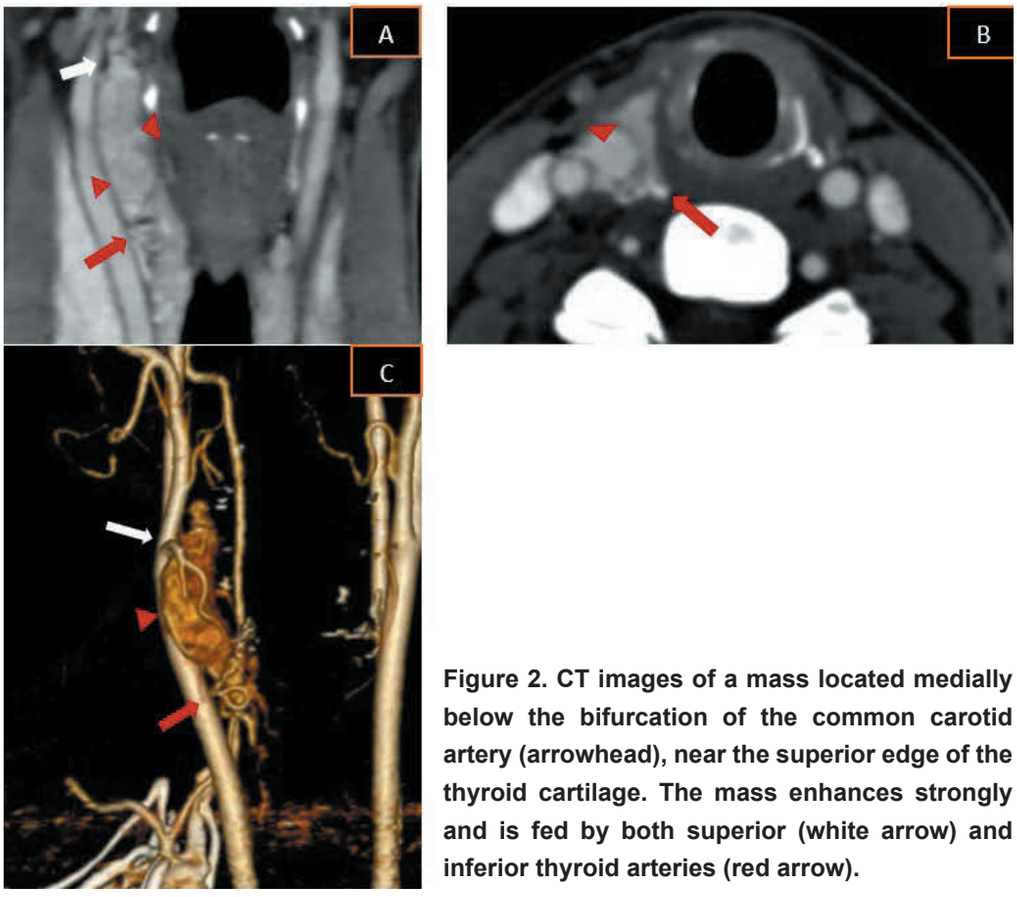

Figure 2. CT images of a mass located medially below the bifurcation of the common carotid artery (arrowhead), near the superior edge of the thyroid cartilage. The mass enhances strongly and is fed by both superior (white arrow) and inferior thyroid arteries (red arrow).

FT4 (15 pmol/L), and TSH (0.95 µU/mL) levels resulted within normal ranges.

The surgical incision followed the anterior border of the right sternocleidomastoid muscle and revealed the tumor lying next to the common carotid artery and external carotid artery. All major surroundings were identified and addressed to maintain hemostasis. The right recurrent laryngeal nerve was not affected during surgery.

The tumor comprised monomorphic cells (chief cells), with round-to-oval nuclei and contained many basophilic granules. Most tumor cells are arranged into trabeculae, islets, surrounded by fibrovascular tissue. There were no images of cystic differentiation, mucinous degeneration, necrosis, mitosis, or amyloid deposition.

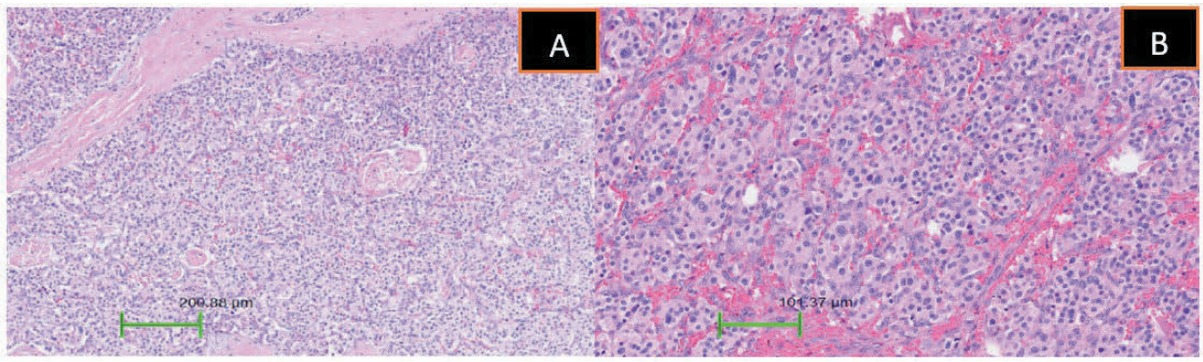

Figure 3. Hematoxylin and PAS staining (A) and (B) with different magnifications demonstrated tumor cells arranged in a well-defined nest (zellballen) pattern surrounded by a thin fibrovascular stroma.

II. DISCUSSION

Head and neck paragangliomas arise from the parasympathetic paraganglia in the neck and are therefore rarely functional (secrete catecholamines). Consequentially, the symptoms of laryngeal and thyroid paragangliomas depend on their size and location causing mass effect, which generally includes dysphonia, dysphagia, stridor, or a foreign body sensation, or as incidental findings on other imaging studies [4,5].

Paragangliomas in the head and neck account for 0.03% of all tumors and 0.6% of all head and neck tumors, estimated incidence is between 0.3 and 1 per 100,000 [8]. In that group, laryngeal paragangliomas are uncommon. It is estimated that approximately 90% of laryngeal paragangliomas are located in the supraglottic area, with 12% in the subglottic region, and 3% in the glottis [2]. Given their origin from the neural crest cells linked to the parasympathetic nervous system, they often associate with either the superior or recurrent laryngeal nerves [9].

Neck ultrasound often shows solid hypoechoic masses with increased vascularity, making them frequently indistinguishable from other solid hypervascular thyroid tumors such as papillary carcinoma [10], [11]. Contrast-enhanced CT of the neck with contrast is useful for suggesting this diagnosis when surrounding abnormal vascularity is present. MRI is advantageous in detecting small tumors or screening concurrent paragangliomas. T1 MRI with contrast is a preferred MRI sequence for laryngeal paragangliomas because the tumor typically presents with avid contrast enhancement.

Differentiating paragangliomas from other pathologies hinges on a combination of histological, immunohistochemical, and clinical features. Paragangliomas predominantly consist of chief and sustentacular cells [5]. The chief cells exhibit characteristic uniform polygonal nuclei and a pale eosinophilic cytoplasm, often arranged in a zellballen pattern [12]. Sustentacular cells with spindle-shaped nuclei appear around the tumor's periphery. Chief cells are typically stained for chromogranin, synaptophysin, neuron-specific enolase, and CD56, while S-100 and SOX10 stains are indicative of the sustentacular cells [13].

In exceedingly rare functional cases, FNA for these patients may be contraindicated without biochemical

screening for catecholamine secretion or pre-FNA alpha-adrenergic blockade [14]. This is another avenue for imaging to guide clinical practice by suggesting the possibility of a thyroid paraganglioma on imaging before FNA to initiate serum catecholamine testing.

Therapeutic techniques include laryngoscopic excision and open resection using a transcervical approach. Endoscopic techniques are indicated in small lesions, however, they are associated with high rates of recurrence. In contrast, open techniques allow excellent access to dissection in the submucosal plane, favoring better exposure and less damage, as well as direct control of the vascular supply. Complete excision while preserving the surrounding structures is essential, primarily focusing on maintaining voice and swallowing functions [15].

Preoperative embolization is often performed to reduce intraoperative blood loss and is recommended in tumors >2.5 cm in size [9], [16]. While the ascending pharyngeal artery is often the primary feeding vessel in more typical paragangliomas such as carotid body and vagal paragangliomas, laryngeal and thyroid paragangliomas are more often supplied by the superior and inferior thyroidal arteries and their branches [2], [17]. There is variability in the vascular supply to the thyroid and larynx in the degree of supply from the superior versus inferior thyroidal arteries, and the inferior thyroidal arteries may arise from either the thyrocervical trunk or the subclavian artery. Knowledge of these anatomical variations is essential when performing selective angiography for preoperative embolization.

III. CONCLUSION

In summary, the diagnosis and management of paragangliomas originating in the laryngeal paraganglia, whether thyroid or laryngeal, demands a multidisciplinary approach. This involves advanced imaging, meticulous histopathological examination, and an understanding of molecular genetics. Proper differentiation from other pathologies ensures accurate treatment, which, when combined with diligent follow-up, can result in favorable patient outcomes.

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Correspondent: Nguyen Thi Thu Hang. Email: lephuocdat99@gmail.com

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