*Case Report / Olgu Sunumu*

# Tracheal paraganglioma: a case report

## Trakeal paraganglioma: Olgu sunumu

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Paragangliomas are neuroendocrine tumors that originate from extra-adrenal chromaffin cells. The trachea is an unusual site for paragangliomas. A 29-years-old female was admitted to our clinic with complaints of recurrent hemoptysis and dyspnea, and a 1.5x1.3 cm mass in the posterior wall of the trachea, which was subsequently diagnosed as a paraganglioma. In this report, we presented the 11th case of tracheal paraganglioma, a rare benign tracheal tumor, and reviewed the medical literature.

*Key Words:* Paraganglioma; recurrent hemoptysis; tracheal mass.

Paragangliomalar ekstra-adrenal kromafin hücrelerden köken alan nöroendokrin tümörlerdir. Trakea ise paragangliomalar için alışılmadık bir bölgedir. Yirmi dokuz yaşında bir kadın hasta tekrarlayan hemoptizi ve dispne yakınmaları ve trakea arka duvarında, daha sonra paraganglioma tanısı konan 1.5x1.3 cm'lik bir kitleyle kliniğimize başvurdu. Bu yazıda, trakeanın nadir bir bening tümörü olan trakeal paragangliomaya ilişkin 11. olgu sunuldu ve medikal literatür gözden geçirildi.

*Anahtar Sözcükler:* Paraganglioma; reküren hemoptizi; trakeal kitle.

Paragangliomas are neuroendocrine tumors originating from extra-adrenal chromaffin cells. The vast majority of head and neck paragangliomas arise from paraganglionic systems, such as the carotid bifurcation, middle ear and ganglion nodosum of the vagus.[1] The trachea is an unusual site for paragangliomas.[2] In this report, we present the 11th case of tracheal paraganglioma, a rare benign tracheal tumor.

### CASE REPORT

A 29-year-old female was admitted to our clinic with complaints of hemoptysis and dyspnea. She had hemoptysis two years ago and recurrent hemoptysis for the last two months. She was referred to our clinic upon detection of a tracheal mass on computed tomography (CT) performed in another clinic. On the sagittal CT section of the neck and axial CT section of the thorax, a mass with a size of 1.5x1.3 cm narrowing the lumen was observed in the posterior wall of the trachea (Figures 1, 2). Direct laryngoscopy revealed a polypoid mass originating 4-5 cm below the vocal cords. Owing to the significant bleeding tendency of the mass, a biopsy was avoided. The red-purple appearance and bleeding tendency of mass led us to consider tumors like hemangioma and paraganglioma in the differential diagnosis. No abnormality was

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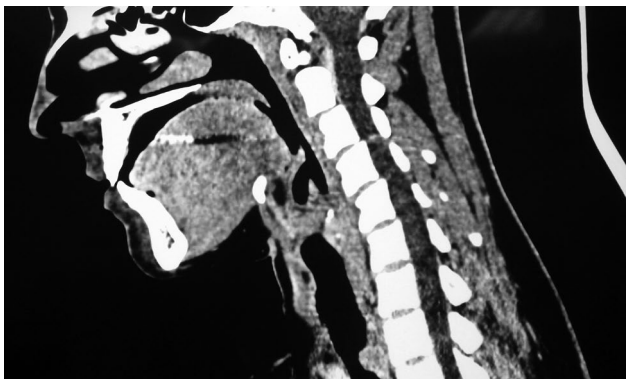

***Figure 1.*** *A mass originating from posterior wall of the trachea on sagittal computed tomography sections of the neck.*

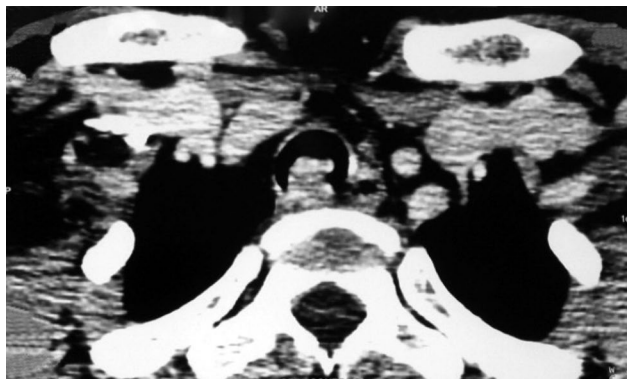

***Figure 2.*** *A mass with a size of 1.5x1.3 cm narrowing the lumen on axial computed tomography sections of the thorax.*

detected in the blood tests of the patient. The urine vanillylmandelic acid (VMA) was within the normal range. Dissection of the wide-based polypoid mass was achieved through a tracheofissure from the posterior wall of the trachea and the base of the lesion was cauterized with diathermy (Figure 3). The sections prepared in the pathology laboratory were stained with hematoxylin-eosin and reticulin; immunohistochemical analyses were performed using S100, chromogranin, and synaptophysin. On histologic examination, nests of tumor cells with eosinophilic cytoplasm (Zellballen pattern) surrounded by fibrovascular stroma were observed (Figure 4).

## DISCUSSION

The tumors originating from the chromaffin cells of the adrenal medulla are referred to as pheochromocytomas.[3] The adrenal medulla is composed of chromaffin cells derived from the neural crest and their supporting sustentacular cells. Chromaffin cells release catecholamines in response to stimulation by sympathetic preganglionic nerve fibers. Similar cells are also located in the extra-adrenal paraganglion system. Non-functional tumors arising from the extra-adrenal paraganglionic system are referred to as paragangliomas, whereas functional tumors are termed extra-adrenal pheochromocytomas.[3] Paragangliomas of the head and neck are innervated by the parasympathetic fibers of the vagus or glossopharyngeal nerves and rarely secrete catecholamine.[1,4]

Paragangliomas may arise along the great vessels in the head and neck and thorax, nerves and vessels of the abdomen and pelvis, visceral organs and sites in which paraganglionic cells

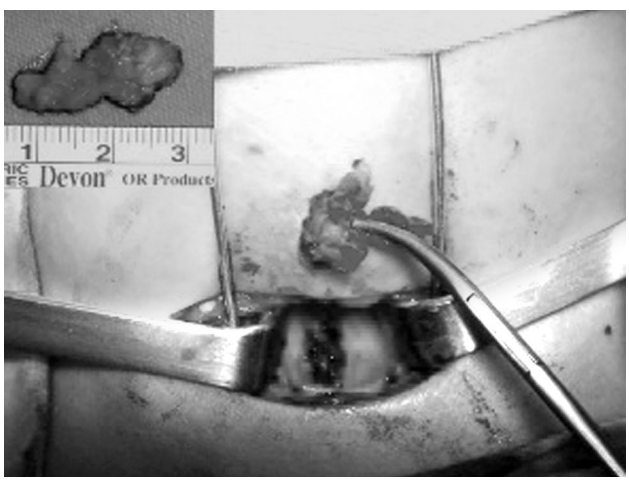


***Figure 3.*** *Wide-based polypoid mass excised from the posterior wall of the trachea by performing tracheofissure and the base of the lesion that cauterized using diathermy.*

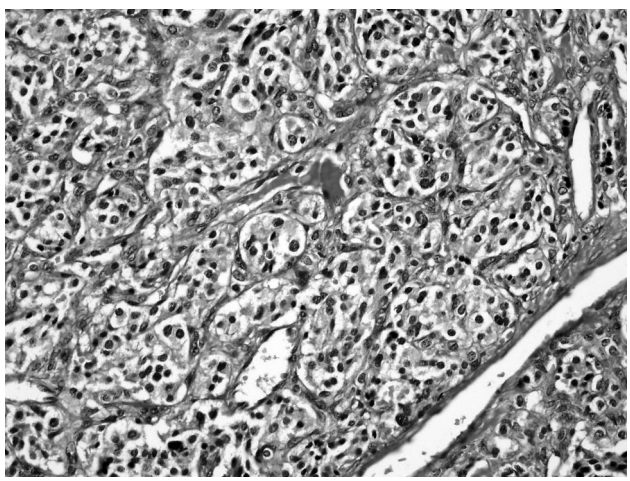

***Figure 4.*** *On histologic examination of tumor cells with eosinophilic cytoplasm and a Zellballen pattern surrounded by fibrovascular stroma (H-E x 400).*

are present.[3,5-8] Paraganglionic cells regulate the vascular system by secreting neuropeptides such as epinephrine, norepinephrine and dopamine.[3,5] Tumors secreting these active neuropeptides are functional tumors causing symptoms such as hypertension, tachycardia and flushing. Such tumors are generally diagnosed by urinary elimination of VMA (catecholamine metabolites). While the tumor cells in the abdomen tend to be functional, tumors in the head and neck region are generally non-functional and are diagnosed by imaging methods and a negative urinary VMA test. However, pathologic examination is required for definitive diagnosis.[3]

The classification of the extra-adrenal paraganglion system according to their anatomic distribution (branchiomeric, intravagal, aortico-sympathetic, and visceral autonomic paraganglioma) has gained wide acceptance.[1] Laryngeal paragangliomas are of branchiomeric origin.[4,9] Supraglottic tumors arise from the superior laryngeal glomus body, whereas subglottic tumors arise from the inferior laryngeal glomus body.[1,9] Tracheal paragangliomas are considered to originate from the main paraganglionic tissue in the trachea.[1]

The definitive diagnosis of a paraganglioma is established by histopathologic and immunohistochemical analyses.[1,9] The microscopic appearance of paraganglioma is characterized by chief cells (type 1) surrounded by sustentacular cells (type 2) which are referred to as a Zellballen pattern. Chief cells contain at least one of the neuroendocrine markers, such as neuron-specific enolase, synaptophysin or chromogranin, enabling differentiation from the other tumors.[9] Paragangliomas almost always have a benign character and <2% of them show malignant features.[10] There are no reliable histologic criteria indicating the malignant potential of a paraganglioma. Pleomorphism, nuclear hyperchromatism and vascular invasion are also observed in benign tumors; however, only metastasis is considered a reliable criterion for malignancy.[1]

Tracheal paragangliomas are extremely rare tumors of the trachea.[1,3,4,10] In 1956, Zeman[6] reported the first carotid body tumor of the trachea. The present case is the 11$^{th}$ case in the medical literature. Although initial publications on this topic reported the age range to be 45-67 years,[1] in recent years tracheal paragangliomas were mostly encountered in the second decade of life.[10] Moreover, while previous reports have indicated a 3-4-fold higher rate of tracheal paragangliomas in females,[9,10] the gender distribution of 11 cases is as follows: six females, four males and one undefined. This condition arises from the rare occurrence of paragangliomas. Therefore, it is not straightforward to limit the female-to-male ratio and the age distribution.

As summarized in table 1, eight patients were admitted to hospital with a complaint of hemoptysis, two patients with a complaint of dyspnea, and one patient with a complaint of cough. Although hemoptysis is the most common symptom, additional symptoms such as hoarseness, dysphagia, stridor and cough have also been reported.[1,3,10] The tumor originated from the posterior wall of the trachea in four cases, the left lateral wall of the trachea in three cases and the right lateral wall in two cases.[3,9,10] In one case, the tumor originated from the anterior wall of the trachea just above the carina[2] and from the bronchus intermedius in the other case.[5]

A defined treatment is not available for endotracheal paragangliomas. While surgical excision is the most commonly performed procedure, various methods such as removal of the tumor by cup forceps and cauterization of the base, segmental excision of the trachea, excision of the mass through a tracheal fissure, removal of the tumor with a Nd:YAG laser, removal of the mass by a flexible bronchoscopy using endobronchial electrocautery and gold probe have also been performed.[3,4,10] The most common complication was intraoperative bleeding in the previous reports and one patient died due to hemorrhage during surgery.[3,4] Therefore, the lesion can be devascularized by preoperative embolization prior to the excision as described by Michaelson et al.[9] In our case, open surgery was preferred because the tumor base had a wide pedicle and no postoperative complications were observed.

Maintaining the patency of the airway is the first goal of treatment and if needed, tracheotomy can be performed prior to surgery.[3] Jones et al.[4] described three alternative methods: *(i)* tracheal intubation following induction of general anesthesia (but the most significant disadvantage is detachment and subsequent aspiration of the tumor cells or hemorrhage); *(ii)* insertion of needle

**Table 1.** Case reports of paragangliomas of the airway

| Reference | Age/gender | Location | Symptoms | Treatment | Recurrence |
|---|---|---|---|---|---|
| Zeman[6] | 45/M | Left wall of trachea just distal to vocal cords | Hemoptysis | Removed with cup forceps. The base of the lesion was cauterized | No (at 3 years) |
| McCall and Karam[7] | 67/F | Right vocal cord extending down trachea to carina | Dyspnea, hoarseness, difficulty in swallowing | Bronchoscopy Tracheostomy, severe tumor hemorrhage, patient died during the operation | – |
| Horree[8] | 56 Not slated | Posterior wall of upper trachea | Dyspnea on exertion, stridor | Surgical excision, marked hemorrhage during biopsy | No (at 6 weeks) |
| Liew et al.[1] | 55/F | Posterior wall of trachea, 2nd to 5th tracheal ring | Hemoptysis | Surgical excision | No (at 6 months) |
| Gallimore and Goldstraw[2] | 55/F | Anterior wall of trachea immediately proximal to carina | Hemoptysis | Segmental resection, notable hemorrhage with biopsy | No (at 12 months) |
| Sandur et al.[5] | 37/F | Bronchus intermedius | Cough, wheezing | Nd: YAG laser, photo resection with wallstent placement | No (at 18 months) |
| Jones et al.[4] | 41/M | Right posterolateral aspect of subglottis | Hemoptysis | Tracheostomy Tumor removed by avulsion | No (at 12 months) |
| Wilson et al.[3] | 86/M | Left anterolateral wall of trachea immediately distal to vocal cord | Hemoptysis, dyspnea, stridor | Bronchoscopy Biopsy Removal with endobronchial electrocautery gold probe | No (at 3 months) |
| Michaelson et al.[9] | 50/F | Subglottic mass. 1-2 cm involving the left paratracheal region, just inferior to the thyroid cartilage | Hemoptysis coughing episode respiratory distress | Tracheostomy. Biopsy. The mass excised with three tracheal rings after preoperative embolization. Primary tracheal anastomosis | No (at 3 months) |
| George et al.[10] | 22/M | Posterior wall of the trachea | Respiratory difficulty intermittent hemoptysis | Tracheostomy tumor was dissected off by a tracheofissure. The base of the lesion was cauterized by diathermy | – |
| ***Current case*** | 29/F | Posterior wall of the trachea | Intermittent hemoptysis dyspnea | Tracheofissure. Surgical excision. The base of the lesion was cauterized by diathermy | No (at 6 months) |

cricothyrotomy following induction of general anesthesia followed by formal tracheostomy (preferred in patients who are not eligible for local tracheotomy); and *(iii)* excision of the tumor with general anesthesia and direct laryngoscopy following local tracheostomy (the author used this method). However, we performed intubation with a thin intubation tube under direct vision following induction of general anesthesia, exposed the trachea by passing through the skin layers with a horizontal incision, after which the patient was extubated. A transoral endoscope was introduced into the trachea, and a needle was inserted from the anterior wall of the trachea to check for its level according to the endoscope light, after that a tracheofissure was performed. The intubation tube was temporarily placed at this level and tumor resection was performed. After bleeding control was achieved, oral intubation was performed and suturing closed the defect. The patient was kept intubated in the intensive care unit on postoperative day one. The patient was then extubated and she was kept in the intensive care unit for a further two days due to the possibility of bleeding. She was discharged on postoperative day five.

In conclusion, the purpose of the present case report was to draw attention to the presence of

rare tumors in cases with diagnostic difficulties. If a paraganglioma is suspected, biopsy should be avoided considering the highly vascular nature of such tumors. If biopsy is to be performed, the surgeon should be prudent for the risk of bleeding. The most appropriate method should be selected during the operation considering the tumor size and its localization.

## REFERENCES

1. Liew SH, Leong AS, Tang HM. Tracheal paraganglioma: a case report with review of the literature. Cancer 1981;47:1387-93.
2. Gallimore AP, Goldstraw P. Tracheal paraganglioma. Thorax 1993;48:866-7.
3. Wilson MA, Judson MA, Silvestri GA, Ravenel J. A case of primary tracheal paraganglioma. J. Bronchol 2003;10:125-8.
4. Jones TM, Alderson D, Sheard JD, Swift AC. Tracheal paraganglioma: a diagnostic dilemma culminating in a complex airway management problem. J Laryngol Otol 2001;115:747-9.
5. Sandur S, Dasgupta A, Shapiro JL, Arroliga AC, Mehta AC. Thoracic involvement with pheochromocytoma: a review. Chest 1999;115:511-21.
6. Zeman MS. Carotid body tumor of the trachea: glomus jugularis tumor, tympanic body tumor, nonchromaffin paraganglioma. Ann Otol Rhinol Laryngol 1956;65:960-2.
7. McCall JW, Karam FK. Chemodectoma of the trachea. AMA Arch Otolaryngol 1958;67:372-3.
8. Horree WA. An unusual primary tumour of the trachea (chemodectoma). Pract Otorhinolaryngol (Basel) 1963;25:125-6.
9. Michaelson PG, Fowler CB, Brennan J. Tracheal paraganglioma presenting with acute airway obstruction. Otolaryngol Head Neck Surg 2005; 132:661-2.
10. George M, Ayyappan AP, Cherian R, Kurien M. Tracheal paraganglioma: a rare vascular neoplasm. AJR Am J Roentgenol 2006;187:W231-2.