

Laryngeal Paraganglioma Causing Airway Obstruction: A Case Report

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Hava yolu obstrüksiyonuna neden olan larengeal paraganglioma: olgu sunumu

Paragangliomalar sıklıkla adrenal bezde görülen nöroendokrin tümörlerdir. Adrenal bez dışı dokulardan en sık kaynaklanan paraganglioma karotid cisimciğidir. Paragangliomalar larenkste oldukça nadiren görülürler. Bununla beraber larengeal kitlelerin ayırıcı tanısında paragangliomaları da düşünmek gereklidir. Bu çalışmada 63 yaşında, 18 aydır ses kısıklığı, son zamanlarda da solunum sıkıntısı şikayetiyle kliniğimize başvuran ve larengeal kitle tanısıyla opere edilen bir olgu sunulmuştur. Laringofissur yöntemiyle opere edilen hastanın histopatolojik sonucu paraganglioma olarak geldi. Postoperatif dokuzuncu aydaki kontrolde nükse rastlanmadı. Bu olguda olduğu gibi büyük boyutlu paragangliomalar laringofissur yöntemiyle eksize edilebilir. Bununla beraber eksizyon sırasında oluşabilecek ciddi kanamalardan dolayı tetikte olunmalıdır.

Anahtar Sözcükler: Paraganglioma, larenks, nöroendokrin tümör, vasküler tümör.

Abstract

Paragangliomas are neuroendocrine tumours commonly seen in the adrenal gland. The commonest paraganglioma that arises from extra-adrenal sites is carotid body. Paragangliomas are rarely seen in the larynx. It is, however, important to consider paragangliomas of the larynx in the differential diagnosis of the laryngeal masses. This study introduces a 63-year-old man who has been suffering from hoarseness for 18 months and has begun to experience dyspnea in recent times and who has had an operation due to the diagnosis of a laryngeal mass. The histopathologic result of the patient who was operated using laryngofissur procedure turned out to be paraganglioma. There was no recurrence in the postoperative 9th month. As in our case, considerably large-sized paragangliomas may be excised with laryngofissur procedure. However, surgeons have to be alert due to the serious bleeding risk during the local excision.

Key Words: Paraganglioma, larynx, neuroendocrine tumour, vascular tumour.

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Introduction

Approximately 90% of paragangliomas developing from non-chromaffin cells are seen in the adrenal gland. The remaining 10% arise from extra-adrenal sites, including abdomen, thorax, head and neck.¹ The commonest of the paragangliomas of the head and neck is carotid body, followed by jugulotympanic paragangliomas and vagal paragangliomas. Other sites

include sinonasal, orbit, trachea, aortic body, lung and mediastinum.^{2,3}

This condition may be rarely encountered in the larynx. It is usually seen in the supraglottic region of the larynx, and these tumors are rarely functional.^{1,4,5} Although paragangliomas usually have a benign nature, they are solely seen as malign tumors.^{3,6} Because of diagnosis and treatment difficulties, we present here a case of paraganglioma of the supraglottic larynx.

Case Report

A 63-year-old woman was admitted in our clinic due to hoarseness that had been continuing for 18 months. In the examination of the patient, it was observed that she had dyspnea due to laryngeal obstruction. The oral cavity, nasal cavity and pharynx were observed to be normal in routine examination. There were no palpable lymph nodes in the cervical region. Endoscopic examination with a rigid endoscope of the larynx revealed a smooth mass (reddish-blue) in the area of laryngeal side of the epiglottis covering the right aryepiglottic fold, false fold and ventricle. Chest X-ray was normal. Computed tomography (CT) scan indicated a laryngeal

mass, obliterating priform sinus in the level of right supraglottic (Figure 1). Magnetic resonance imaging (MRI) of the neck showed a supraglottic mass as being 4x5 cm in diameter with hyoid bone invasion, obliterating right priform sinus (Figure 2A). On contrast enhanced images, the mass showed intense contrast enhancement (Figure 2B). Because of the dense vascularity of the mass in the color Doppler ultrasound, it was postulated that the mass might be a vascular tumor. Subtraction angiography of the right common carotid artery showed that the mass was prominently vascularized, and its blood supply was primarily from branches of the superior thyroid artery. The laboratory tests did not indicate any neurosecretory activity, and the other laboratory tests were within the normal ranges. The rest of her examination was unremarkable. After the tracheotomy, punch biopsy was performed under the general anaesthesia in order to find out diagnosis of the mass. Bleeding appeared during punch biopsy. But this bleeding was within acceptable limitations. Unfortunately, as a result of the repeated punch biopsies (three times), histopathological diagnosis was not possible. The mass was excised using laryngofissure procedure. Serious bleeding appeared during the local

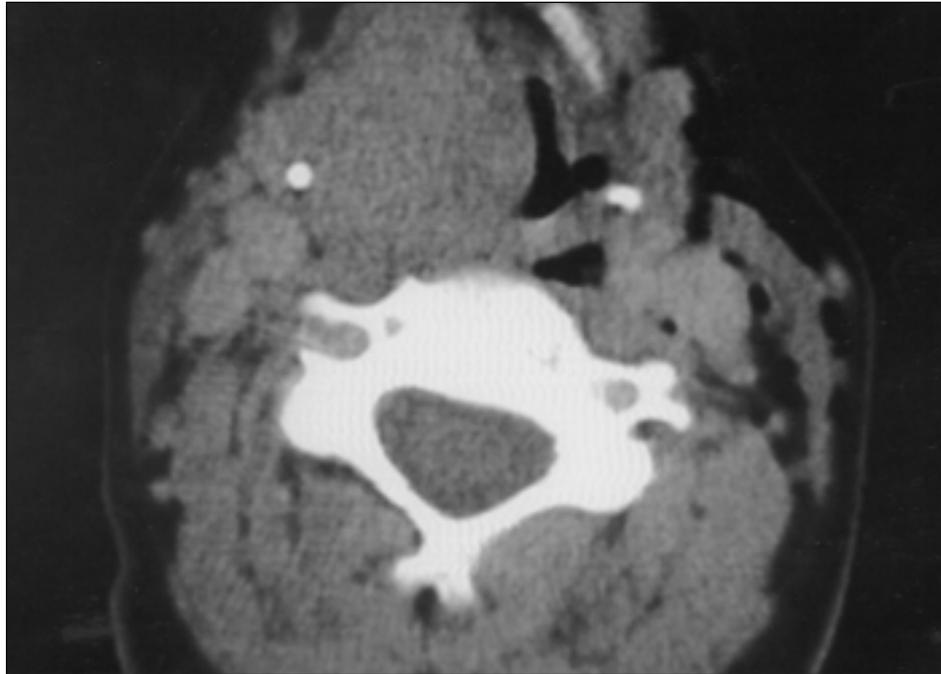


Figure 1. Computed tomography (CT) scan shows a right supraglottic laryngeal mass obliterating piriform sinus with invasion of the hyoid bone on the right.

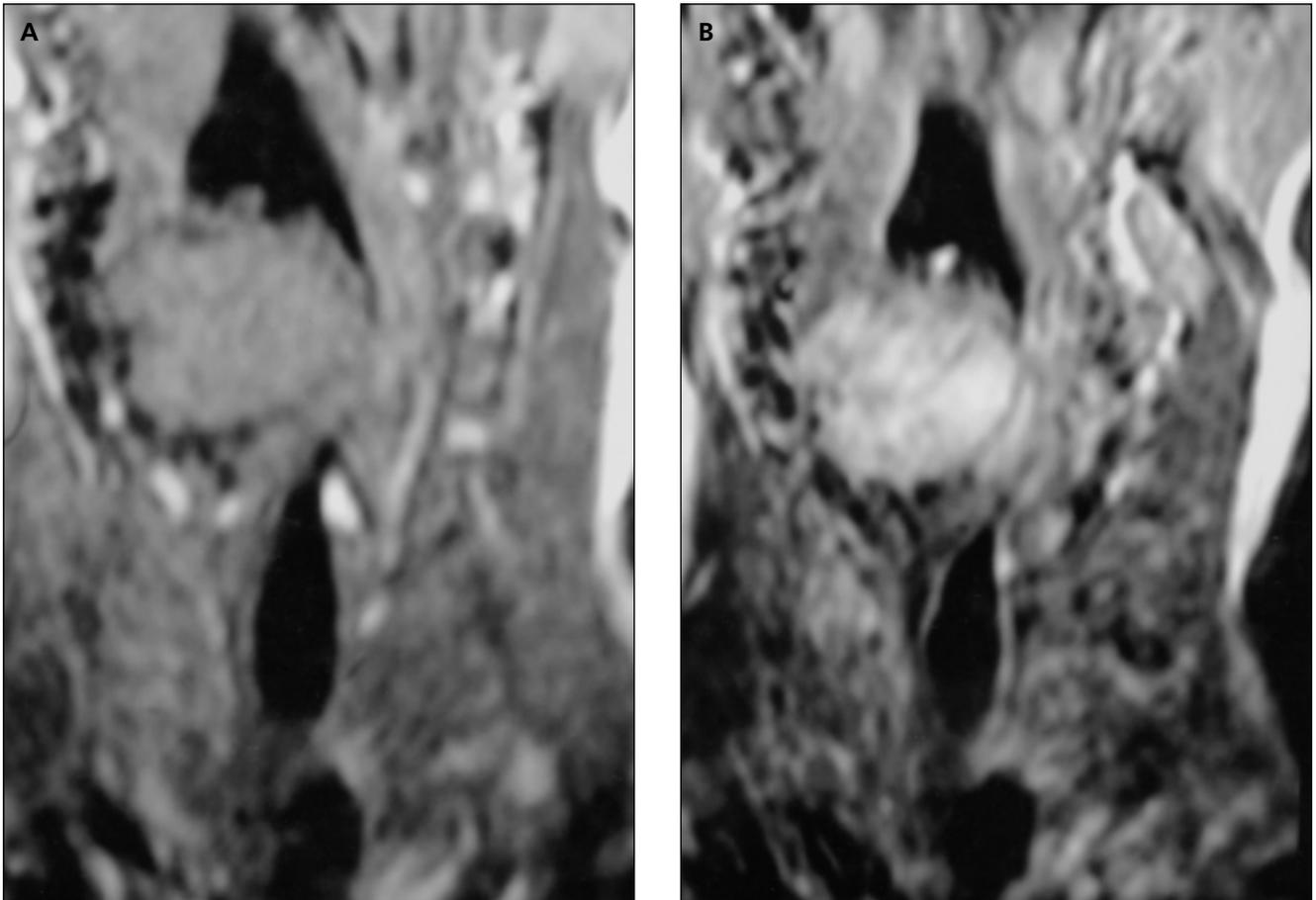


Figure 2. On coronal plain T1-weighted image (TR/TE, 500/20), the laryngeal lesion has similar intensity with muscles and internal vascular flow-voids (A). There is also prominent enhancement on contrast enhancing T1-weighted image (B).

excision of the mass, and two unites of blood had to be transfused. In the examination of the histological sections, it was observed that the neoplasm was made up of chief cells arranged in clusters and round cell nests surrounded by delicate stroma containing numerous vascular channels (Figure 3A). These cells had abundant eosinophilic cytoplasm, prominent nucleoli, and vesicular nuclei. Mitoses, necrosis and vascular invasion were not present. Vascular spaces were prominent. A reticulin stain accentuated the organoid pattern (Zellballen configuration) (Figure 3B). The tumor was argyrophil-positive and argentaffin-negative. Glandular structure was absent. The tumor contained no mucin. Immunohistochemical investigations of the tumor showed positive reactions of the chief cells for neuron-specific enolase, chromogranin and synaptophysin (Figure 3C). The S-100 protein stained the sustentacular

cells. The immunohistochemical reactions were negative for calcitonin, bombesin, cytokeratin, CEA, desmin and GFAP. These findings support our diagnosis of paraganglioma of the larynx. Swallowing and respiratory functions of the patient were normal in the postoperative ninth month.

Discussion

Paragangliomas are neuroendocrine tumors with a neural origin. They are rarely seen in the larynx. The other neuroendocrine tumours are carcinoid tumour, atypical carcinoid tumour and small cell neuroendocrine carcinoma. But unlike paragangliomas, these tumors are with an epithelial origin.⁷ Although neuroendocrine tumors of epithelial origin may show malignant behavior, it is a controversial subject in the

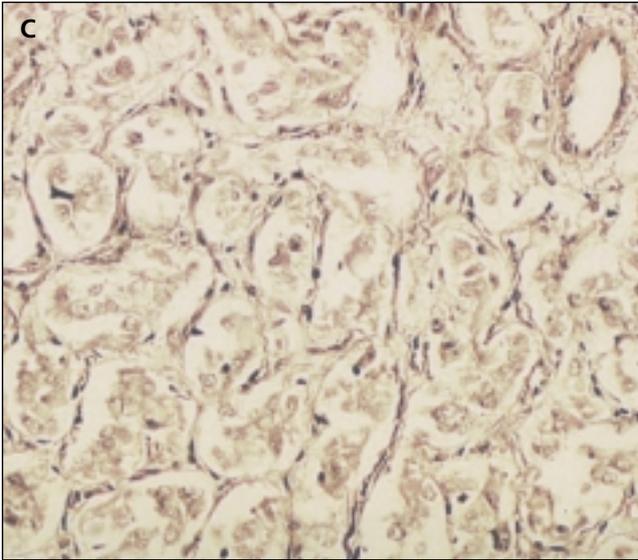
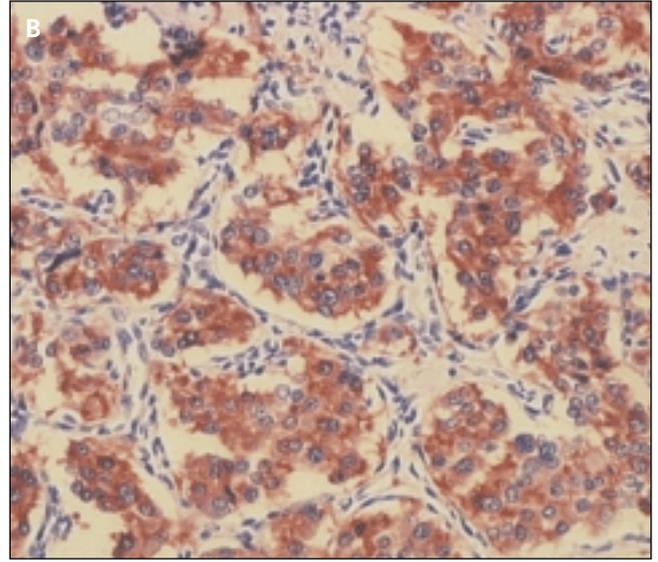
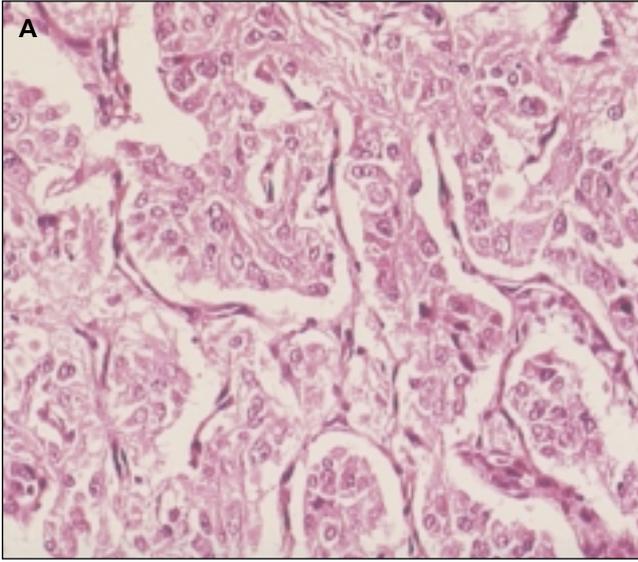


Figure 3. Paraganglioma is composed of chief cells and sustentacular cells (HE **x200**) (A), organoid nests (Zellballen) separated by thin fibrovascular septa (reticulin stain, **x100**) (B). Reactivity of tumour cells (synaptophysin, **x200**) (C).

literature whether paraganglioma have malignant behavior or not in the field of otorhinolaryngology-head and neck surgery.^{4,6-8}

Histopathologically, it is difficult to find out precise frequency of these tumors due to diagnosis difficulties. For atypical carcinoids may be confused with paragangliomas or typical carcinoids, and small cell neuroendocrine carcinoma may be confused with undifferentiated carcinomas.⁷ Immunohistochemical studies are important in order to be able to establish the correct

diagnosis. The chief cells generally express all general neuroendocrine markers (neuron-specific enolase, chromogranin and synaptophysin) and are usually negative for epithelial stains (cytokeratin, EMA, CEA). They are not stained with bombesin or calcitonin.⁷ Several investigators have stated that the cytokeratin marker is the most useful one for distinguishing paraganglioma and neuroendocrine carcinoma.⁹

Hoarseness, dyspnea, dysphagia and laryngeal pain sometimes induced by swallowing, hemoptysis and foreign body sensation are the known symptoms of laryngeal paraganglioma.⁹⁻¹¹ There was no complaint except hoarseness and dyspnea in our case having a considerably large supraglottic mass.

CT scan, MRI, ultrasonic scanning with Doppler flow measurement and arteriography are of important value in the diagnosis of paragangliomas. On contrast enhanced CT scan and MRI, a prominent contrast enhancement of the lesion in the larynx is an important finding in differential diagnosis of paraganglioma.¹² The value of arteriography in the evaluation of paragangliomas of the head and neck is to establish their vascular nature, to determine the size, blood supply and extent of the lesion. Nonlaryngeal paragangliomas, often poorly circumscribed, have a variable blood sup-

ply and are frequently multicentric, especially with a positive family history. By contrast, the supraglottic paraganglioma is well circumscribed, receives its blood supply via the superior thyroid artery, and has a low incidence of multicentricity.¹³ Besides, preoperative embolization of tumour blood supply may be achieved during the arteriography.^{12,14} The other diagnostic procedure is the histopathological examination of the punch biopsy made under the direct laryngoscopy. The precise diagnosis of the tumors is essential to the histopathological examination as all physicians know. Biopsy must be made carefully due to profuse bleeding risk in the cases of paraganglioma. Malignant paragangliomas are diagnosed on the basis of their clinical behavior and not solely on the basis of their histological appearance.¹⁵ Biologically, paragangliomas are defined as malignant when there are regional and/or distant metastases. Histologic criteria which, by themselves, reliably identify those paragangliomas which possess a capacity for metastasis have not yet been generally agreed upon. Clinical findings remain the most reliable criteria for malignancy. Laryngeal paraganglioma are, in fact, almost invariably benign. The incidence of metastases from acceptable cases of laryngeal paraganglioma appears to be less than 2%.¹⁶ Despite the repeated (three times) punch biopsies, we did not observe any serious bleeding in our case. However, precise diagnosis of the disease was not accomplished with the histopathological examination of the samples. This condition may be due to the fact that biopsies are not taken from the depth. Because of the submucosal localization of these tumors, a deep biopsy is always necessary to obtain a representative tissue sample.¹⁰ The treatment of paraganglioma consists of adequate local excision, and elective neck dissection is not necessary.⁷

Radiotherapy and chemotherapy are of no value in the treatment of laryngeal paraganglioma.^{13,17} Our case was treated with local excision via laryngofissur procedure. The patient was free of the disease in the 9th month after the operation.

Consequently, it is important to consider paraganglioma of the larynx in the differential diagnosis of the

laryngeal masses, especially located in the supraglottic area. Even considerably large-sized paragangliomas as in our case may be excised using laryngofissur procedure. However, surgeons have to be alert due to serious bleeding risk during the local excision.

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