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CASE REPORT

Laryngeal Paraganglioma: Diagnostic Challenge of an Unusual Case

Laringeal Paraganglioma: Nadir Bir Vakanın Tanısal Zorluğu

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ABSTRACT

Paragangliomas are frequently observed in the head and neck region, while laryngeal paragangliomas are extremely rare, but are seen far more frequently among women than men. Their immunohistochemically features require experience for proper pathological diagnosis, and preoperative assessment is important due to the risk of hemorrhage during treatment. This article reports on the observations made during the diagnosis and treatment of a 67-year-old female patient, and makes a review of previous literature.
Keywords: Paraganglioma, Laryngeal; Challenge; Immunohistochemistry.

ÖZET

Paragangliomalar baş boyun bölgesinde sıkça görülmesine rağmen laryngeal paragangliomalar son derece nadir görülür. Kadınlarda erkeklere oranla daha yüksek oranda görülmektedir. Patolojik tanıda immunohistokimyasal özellikleriyle tecrübe gerektirir. Tedavide hemoraji riskinden ötürü preoperatif değerlendirme önem taşımaktadır. Biz bu yazıda 67 yaşında kadın hastanın tanı ve tedavi sürecinde yaşanan tecrübeyi ilgili literatür eşliğinde tartıştık.
Anahtar kelimeler: Paraganglioma, Laringeal, Zorluk, Immunohistokimya.

Paragangliomas are tumor masses that arise out of the neuroendocrine structures of autonomous nervous system paraganglia (1). Head and neck paragangliomas account for 0.6 percent of all head and neck tumors. In the absence of any anatomopathological criteria regarding such tumors to indicate malignancy, metastasis is the only tangible criterion for malignancy (2). The most frequent localization are the carotid body, jugular bulb, tympanic plexus and vagal ganglions. Laryngeal paragangliomas are rare (3), although they appear most commonly in the larynx as non-squamous tumors (4). While they are generally observed as single lesions, 10 percent of sporadic tumors and 40 percent of familial tumors may appear as multiple lesions (5). The average age of onset is between 40 and 60, while cases involving women outnumber those involving men by a ratio of 3:1. The larynx has two paraganglia, one superior and the other inferior, and the localization of tumors is most frequently supraglottic (82% of cases) (6).
This article reports on the difficulties experienced in the diagnosis and treatment of a laryngeal paraganglioma case with a supraglottic localization that was observed in a particularly rare case.

CASE REPORT

A 67-year-old woman was admitted with complaints of snoring, difficulty swallowing and the sense of having something stuck in her throat that had been ongoing for two years. The patient's complaints had gradually worsened over time, and a laryngoscopic examination of the patient identified a round mass with a regular surface and a benign appearance originating from the side of the right aryepyglottic fold, facing the priform sinus. The mass limited the view of the right vocal cord and arytenoid. The results of other examinations were normal. The coronal, axial and sagittal images on a contrast computerized tomography of the patient showed a contrast-retaining mass at the identified localization (Figure 1).

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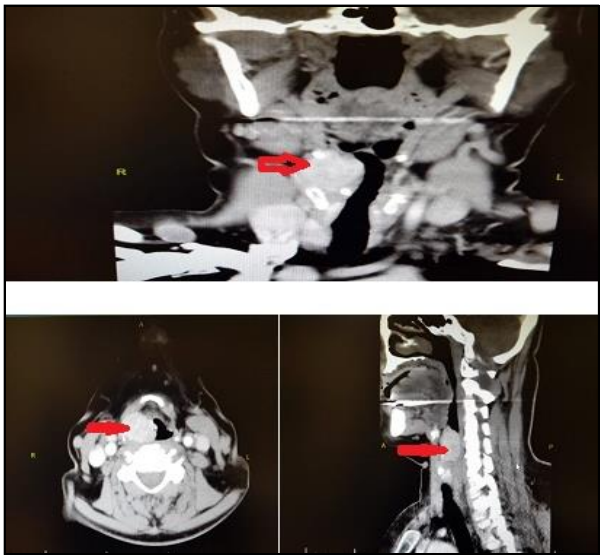

Figure 1. The contrast-enhanced right supraglottic mass can be seen in the coronal, axial and sagittal sections of the computerized tomography.

No pathologies were observed in the preepiglottic area, vallecula or other laryngeal structures. The decision was taken to perform a laryngoscopic biopsy on the patient, and the direct laryngoscopy revealed a mass originating from the lateral mucosa of the right aryepiglottic plica pressing against the lateral wall of the priform sinus, partially blocking the view of the larynx. After the biopsy was taken, the excised piece was sent for a frozen section examination due to its predisposition to bleeding. The mass was presumed to be benign due to its regular mucosal surface, which was an assumption that was supported by the frozen section results. Accordingly, the decision was taken to make a total excision by opening a tracheotomy, and the patient's family was informed of this decision. The mass excision and bleeding control was carried out using a laryngofissure and endoscopic laryngoscopy. In a pathological examination, the mass stained negative for pan-cytokeratin and positive for S100, synaptophysin and CD56 (Figure 2).

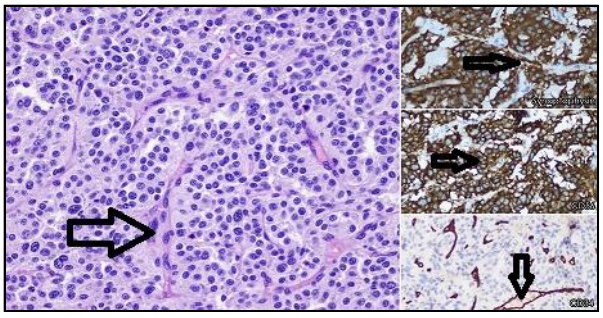

Figure 2. Laryngeal Paraganglioma, comprising islands of Zellballen nests. The tumor showed diffuse staining with neuroendocrine markers such as synaptophysin and CD56, and had a notably widespread vascular network (H&E, X200; Immunoperoxidase, X200).

The mass was hence diagnosed as laryngeal paraganglioma.

DISCUSSION

Laryngeal paraganglioma was defined for the first time by Blanchard and Saunders in 1955 (7). The larynx constitutes a very rare localization for paragangliomas, and is generally not associated with the multi-centric and familial types. Most laryngeal paragangliomas are observed as supraglottic submucosal masses (8). Depending on the size and localization of the tumor, patients may present with non-specific symptoms such as dyspnea, dysphagia, snoring or a sense that something is stuck in their throat (9). This case involved a smooth-surfaced mass arising from the lateral surface of the supraglottic aryepiglottic fold, and the patient was admitted with symptoms of dysphagia and the sense of having something stuck in her throat. During diagnosis, there are several challenges to obtaining and evaluating a biopsy. The biopsy must be taken deeply, but since as a vascular pathology, the risk of hemorrhage must be considered. In a pathological diagnosis, two types of cell series stand out, namely the principal cells and the sustentacular cells, although there is also a characteristic polygonal-shaped Zellballen pattern with round nuclei and eosinophilic cytoplasm. There are no anatomopathological criteria for the malignancy of paragangliomas, with the only clinical assessment criterion being the presence or absence of metastasis. Only two percent of laryngeal paragangliomas are malignant, and are frequently diagnosed as atypical carcinoids (10). Paragangliomas exhibit positivity to neuroendocrine markers such as synaptophysin, chromogranin, CD56 and neuron-specific enolase, while showing no staining with epithelial markers such as cytokeratins, CEA and EMA. In addition, among the tumor cells, the sustentacular cells react with S-100. Immunomarkers can be used to distinguish between typical carcinoid tumors and atypical ones, which is the main differential diagnosis. Paragangliomas release the regulator neuropeptide galanin, which is absent in carcinoid tumors. In addition, atypical carcinoid tumor cells exhibit a higher nucleus-cytoplasm ratio than paragangliomas (11).

A local resection is sufficient in most cases, and no additional treatment is required, such as neck dissection or radiotherapy. Various surgical options have been tried in literature, including elective supraglottic laryngectomies, tracheotomies and laryngeal reconstruction, all of which were applied without performing neck dissection (12). Furthermore, if there are no pathological lymph nodes on the neck, a surgical excision through the cervical approach (lateral pharyngolaryngectomy) without dissection may be preferred for laryngeal paragangliomas (2, 13, 14), while a preoperative embolization may also be applied (2). While endoscopic excision under a suspension laryngoscopy may be opted for in some cases, this is generally not recommended due to the risk of uncontrolled bleeding

(15-17). To control excision and bleeding in the present case we resorted to a tracheotomy, the laryngofissure laryngeal approach, and the use of endoscopic vision-supported excision under laryngoscopy. No recurrence was identified at the sixth postoperative month of the patient.

Laryngeal paragangliomas can be difficult to diagnose and treat. As a highly vascularized pathology, despite having a benign regular surface, there can be severe bleeding, even during a biopsy under direct laryngoscopy. Furthermore, due to its regular surface, a pathological diagnosis requires a deep submucosal biopsy, which further increases the risk of bleeding. For these reasons, preoperative imaging and, in case of suspicions, angiographic embolization may be employed instead. A surgical excision with the proper approach is also of vital importance.

Conflict of interest: All authors declared no conflict of interest.

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