Case Report of a Rare Tumour in an Unusual Site

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Abstract: Laryngeal paragangliomas are rare tumours of neuroendocrine origin with a reported incidence of fewer than 100 cases in world literature. We report the case of a 62 year old gentleman with supraglottic paraganglioma.

Keywords: Larynx; paraganglioma; supraglottic

1. Background

Paragangliomas are rare neuroendocrine tumours which account for 0.6% of neoplasms in the head and neck¹. The four primary sites in this region include the carotid body, along the course of vagus nerve, the jugular foramen and within the middle ear cavity, Larynx being an unusual site.

2. Case Report

We report the case of a 62year old gentleman who presented to our hospital with hoarseness of voice of 2 months duration. Video laryngoscopy (Figure 1) revealed a smooth mass arising from right lateral pharyngeal wall partially blocking the laryngeal inlet. Radiograph of neck, AP view (Figure 2) showed an oval homogeneous radio-opacity displacing the tracheobronchial air column to the left. Lateral view (Figure 3) depicts the lesion in supraglottic larynx obliterating the air column. From the radiographs, lesion was localized to right supraglottis. Grey scale ultrasound scan of neck (Figure 4) revealed a well circumscribed hypoechoic lesion involving the larynx closely abutting the right side of thyroid lamina. Colour Doppler evaluation (Figure 5) demonstrated significant internal vascularity with feeders from ipsilateral external carotid artery. For superior delineation of site, extent and characterization of lesion, we proceeded with contrast enhanced CT scan of the head and neck. Plain and contrast enhanced CT scan (Figure 6) revealed a well circumscribed intensely enhancing lesion measuring 2.4x3x3.1cm in supraglottic larynx on right side. The lesion was seen involving the right half of epiglottis, right aryepiglottic fold and false cord, reaching up to the pyriform fossa, displacing the epiglottis to left with asymmetrical airway narrowing. Prominent feeders from right external carotid artery seen piercing the thyrohyoid membrane and supplying the lesion. True vocal cords and infraglottic larynx unremarkable. No evidence of extra-laryngeal extension. A diagnosis of right supraglottic paraganglioma was made. The tumour was successfully resected by means of midline laryngofissure with mucosal preservation. Histopathologic examination (Figure 7) showed nests of round to oval cells surrounded by delicate vascular septae. Tumour cells have abundant clear to granular eosinophilic cytoplasm and uniform round to ovoid nuclei, some with vesicular nuclei, confirming the diagnosis of paraganglioma.

3. Discussion

Laryngeal paragangliomas are rare neuroendocrine tumours with a female predominance², female-to- male ratio of 3:1. Age at presentation is between 4th -6th decade. 90% of cases arise from the superior laryngeal paraganglia. CT and MR depict these highly vascular soft tissue masses equally well and serve as the main imaging modalities for preoperative diagnosis. Lesions demonstrate intense and homogeneous enhancement following contrast administration owing to the lesion's intrinsic hypervascularity. 2% of these tumours are claimed to be malignant³. Ultrasound examination is useful in the follow up of small paragangliomas. Major differential diagnosis include typical and atypical carcinoid, vascular leiomyoma.



Figure 1

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Figure 2, 3



Figure 4



Figure 5



Figure 6



Figure 7

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