CASE REPORT

AN EARLY PRESENTATION OF SUPRAGLOTTIC LARYNGEAL PARAGANGLIOMA: CASE REPORT AND REVIEW OF LITERATURE

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A thirty-year-old male presented with progressive hoarseness and dysphagia for solids. Fibre optic laryngoscopy showed a right supraglottic mass. Subsequent CT imaging showed the location and extent of the mass. Biopsy revealed a Paraganglioma with cytokeratin negative and chromogranin positive cells. The mass was subsequently removed without complications. Follow up also showed no complications. Hoarseness and dysphagia resolved following tumour excision.

Keywords: Hoarseness; Dysphagia; Paraganglioma; Cytokeratin; Chromogranin

Citation: Zaidi SAH, Raza SS, Ahmad D, Medhat N, Haq IU, Ayaz T, *et al*. An early presentation of supraglottic laryngeal paraganglioma: case report and review of literature. J Ayub Med Coll Abbottabad 2021;33(3):532–4.

INTRODUCTION

Neuro-endocrine neoplasms of the larynx such as laryngeal Paragangliomas are rare. This case is an earlier than usual presentation of a rare tumour, in the non-dominant demographic.

CASE PRESENTATION

A thirty-year-old male presented with complaints of progressive change in voice for the past one year and dysphagia for solids for the past five to six months. There was no associated weight loss or fever. No significant past medical, surgical and family history was present. Sleep and appetite were normal. Evaluation of all systems yielded no abnormal findings. The socioeconomic status of the patient was satisfactory as he could bear the cost of treatment. After initial evaluation, CECT Neck revealed supraglottic mass and indirect laryngoscopy confirmed a right supraglottic mass, a normal tongue and a normal palate [Figure-1]. No obvious lymphadenopathy was found. Direct laryngoscopy and biopsy were then planned and undertaken under general anaesthesia. There was some difficulty with endotracheal intubation prior to general anaesthesia. The patient required stabilization of blood pressure before this procedure. During the procedure, a tissue sample was taken for biopsy. Minor bleeding was controlled through pressure packing. Tracheostomy was done post-procedure. After the procedure there was a brief period where oxygen saturation was not satisfactory. This resolved after pulmonology consultation and subsequent management. Two weeks after direct laryngoscopy and biopsy, lateral pharyngotomy was done in order to completely remove the tumour [Figure-2]. There was no complaint of dysphagia or voice loss by the patient after lateral pharyngotomy. The patient began oral intake of liquids twenty-four hours later and solid food three days after the procedure. Follow up was at one week and one month after the procedure. Biopsy revealed Paraganglioma [Figure-3] The presenting complaints of hoarseness and progressive dysphagia resolved after complete surgical excision of the tumour.

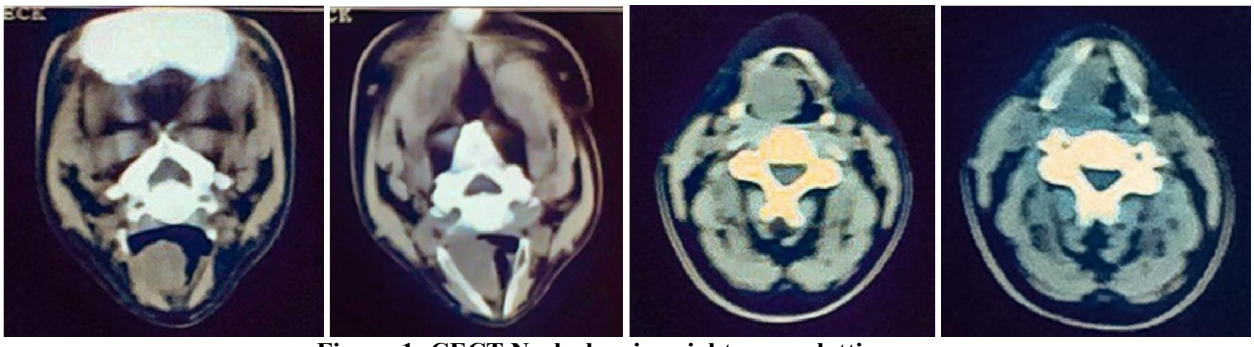

Figure-1: CECT Neck showing right supraglottic mass

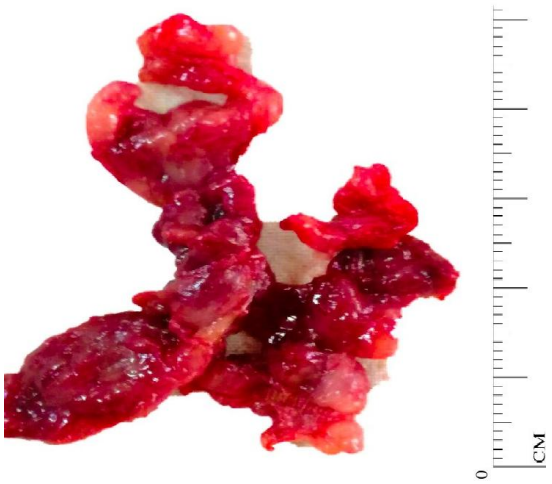

Figure-2: Surgery specimen

DISCUSSION

Neuroendocrine tumours of the larynx have no uniform classification system, as such; there is a lot of confusion, which could lead to improper classification.[1,2] It is crucial to diagnose correctly since the management and prognosis vary for all the types of laryngeal neuroendocrine neoplasms.[1] Broadly, they can be classified as primary epithelial and non-epithelial. The epithelial neoplasms include typical carcinoid, atypical carcinoid, and small and large cell neuroendocrine neoplasms.[3]

Typical carcinoid is a rare neuroendocrine tumour of the larynx. It has the potential to metastasize. The atypical carcinoid is an aggressive lesion with distant metastases and poor survival. Both are treated surgically with elective neck dissection being preferred for atypical carcinoid.[1,3] In order to distinguish between Paragangliomas and carcinoids, the most helpful biomarker is cytokeratin.[3,4] Small cell neuroendocrine carcinoma (SCNC) has a 5-year survival rate of about 5 percent. It is also a very aggressive lesion with high potential for malignancy.[3,5] Radiation and adjuvant chemotherapy confer the greatest survival benefit in treating SCNC. Surgery is not recommended as a treatment option.[6]

Large cell neuroendocrine carcinoma has been classified by WHO as an independent entity.[7] It is also a very aggressive lesion that usually leads to death in the two years following diagnosis.[8] Medullary thyroid carcinoma is also a differential diagnosis for laryngeal neoplasms. In very rare instances, this can extend to the larynx.[3] The biomarker TTF-1 is used in differentiating between it and laryngeal neoplasms.[9]

Laryngeal Paraganglioma poses a diagnostic challenge. This is due to some overlap in immunohistochemistry with other neuroendocrine lesions of the larynx[10]. Although other neuroendocrine tumours can be positive for chromogranin, laryngeal Paraganglioma is mostly cytokeratin negative.[9] The characteristic histologic growth pattern (Zellballen), the benign nature of the mass, the supraglottic location, resolution of presenting complaints post-resection, chromogranin positivity and cytokeratin negativity all indicated a diagnosis of supraglottic laryngeal Paraganglioma. Out of many, one treatment option is lateral pharyngotomy. This procedure is used for submucosal tumour excision in the larynx.[11,12] Laryngeal Paraganglioma is one such lesion.

The laryngeal Paraganglia are part of the branchiomeric extra adrenal Paraganglia. This system originates from the neural crest cells. In the larynx, the superior and inferior pairs of Paraganglia are found along with occasional accessory Paraganglia.[13]

Laryngeal Paragangliomas are more frequent in females with a ratio of almost three to one compared with males. The incidence peaks in the forties to sixties. In about 80–90% of cases, supraglottic Paragangliomas are found.[2,13,14] In our case, the patient was a male who presented at age thirty. The location as described above was right supraglottic. Cancers of the larynx are classified as rare.[1] Thus far, less than a hundred verified cases of laryngeal Paraganglioma can be found in the literature.[2,15] The problem with verification of reported cases is the lack of a universal classification system. For example, it is thought that reported cases of malignant laryngeal Paragangliomas might actually be atypical carcinoids.[3,16] To our knowledge, this might be the first reported case of a supraglottic laryngeal Paraganglioma from Pakistan.

This tumour has a benign course and excellent prognosis. However, some sources caution against labelling it as benign. The metastatic rate is about 2% in Paragangliomas of the larynx.[2,17] It usually presents with mass effects. The most common presenting symptom is hoarseness. Our patient presented with a history of hoarseness for almost a year. Other common presenting symptoms include dysphagia, dysphonia, dyspnoea, cough, stridor, etc. In this particular case, the patient also complained of dysphagia for solids. The mean duration of symptom(s) is a little over 2 years.[2,13,18]

Surgical rather than endoscopic excision is the recommended treatment for laryngeal Paraganglioma. The risk of local recurrence is about 17%, especially if excision was done inadequately.[1,13,15] In the present case, after confirmation of diagnosis, surgical excision was done through lateral pharyngotomy. This resulted in the resolution of symptoms and follow up after one week and one month was also clear.

CONCLUSION

Hoarseness, especially if prolonged, should increase the suspicion for cancer of the larynx. Therefore, Precise diagnosis of laryngeal neuroendocrine tumours should be made using histology and immunohistochemistry. Weigh the risks and benefits of biopsy before excision surgery of a local laryngeal tumour is necessary in order to improve morbidity. If a diagnosis of laryngeal Paraganglioma is confirmed, complete surgical excision should be the treatment of choice. Chemotherapy and radiation therapy have no role in management.

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Submitted: July 31, 2020	*Revised: --*	*Accepted: December 13, 2020*

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