LARYNGEAL PARAGANGLIOMA - A CASE REPORT
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Abstract: Laryngeal Paraganglioma are benign slow growing tumours with symptoms resembling squamous cell carcinoma. Hoarseness and dysphagia are the commonest presenting symptoms and usually they present as a sub mucosal mass on laryngoscopy. 90 percent of the tumours occurs in supraglottis and the rest in glottis and subglottic region. Functional activity is seen in a few (2.9 percent). None are associated with para neoplastic syndromes. A 43 year old female presented with complaints of change in voice and difficulty in breathing for 2 months. MRI demonstrated a well defined homogenous moderately enhancing lesion seen arising from the posterior wall of epiglottis and microscopy confirmed it to be a paraganglioma. Pre operative tracheostomy was done. The tumour was excised by lateral pharyngotomy approach.

INTRODUCTION:
Head and neck para ganglioma are rare tumours arising from the neural crest. They are highly vascularised tumours involving the walls of the blood vessels or specific nerves. In the larynx they arise from the superior or inferior laryngeal paraganglia and are seen in supraglottis in 90% of cases. Since these tumours are neural crest-derived cells of the parasympathetic nervous system, they usually appear adjacent to nerve structures, most commonly the superior laryngeal nerve or the recurrent laryngeal nerve. The paraganglioma of larynx show a female preponderance compared to other neuro endocrine tumours. Other neuro endocrine tumours differ in their biologic behaviour, management and prognosis compared to paragangliomas. Paraganglioma of larynx are benign sub mucosal lesion and the management includes a conservative resection.

CASE REPORT:
A 43 year old female patient presented with history of change of voice for the past 1 year & progressively increasing difficulty in breathing for past 2 months. She has no history of throat pain, dysphagia, aspiration, neck swelling, hemoptysis, hematemesis and no other significant medical or surgical illness in the past.

ON CLINICAL EXAMINATION: General examination findings were all normal. On Indirect laryngoscopy (FIG 1) there was a smooth globular swelling arising from the laryngeal surface of epiglottis extending to the left aryepiglottic and obscuring the view of laryngeal inlet.

INVESTIGATIONS:
MRI Neck – T1 hypo(FIG 2A), T2 hyper intense (FIG 2B). Well defined homogenous moderately enhancing lesion seen arising from the posterior wall of epiglottis seen effacing the oro laryngo pharyngeal junction. Pre-epiglottic fat appears normal and paralaryngeal structures normal. Provisionally the pre operative diagnosis was adenoma arising from posterior wall of left epiglottis. All other blood investigations were normal.

FIG 1 videolaryngoscopy showing smooth globular mass
FIG 2A-MRI Neck axial view showing homogenous moderate enhancing lesion
FIG 2B MRI NECK Sagittal view showing homogenous enhancing lesion

TREATMENT:
Pre-operative tracheostomy (FIG. 3) was done. Boyle-Davis mouth gag was introduced & under endoscopic guidance biopsy was taken from the lesion. Biopsy report came as Paraganglioma (FIG. 4) composed of nests of polygonal chief cells (zellballen) enclosed by trabeculae of sustentacular cells. To know whether it is secreting or not, 24 hour urinary Vanillyl Mandelic Acid was done which was normal. The patient was operated by Lateral Pharyngotomy approach, making a horizontal incision (FIG 5A) at the level of thyrohyoid membrane that followed a mid neck skin crease. Sub platysmal flaps raised and plane of dissection was carried deep to the sub mandibular gland. Pharyngotomy revealed a mass lesion (FIG 5B) in posterior wall of epiglottis which was dissected submucosally and removed in toto and nasogastric tube was inserted. Wound closed in layers. Final HPE report also confirmed paraganglioma. Post operative period was uneventful (FIG 6A & 6B). Nasogastric tube was removed on the seventh post operative day and oral feeding started. Patient weaned from tracheostomy tube. Patient is on regular follow up and there is no recurrence.

Fig 5 A Picture showing horizontal incision

FIG 3 Clinical picture showing pre operative tracheostomy

FIG 4 showing Histopathology report-suggestive of paraganglioma

FIG 6A Post op picture

FIG 6B post operative picture

DISCUSSION:
LARYNGEAL PARAGANGLIOMA:
Less than 80 cases of laryngeal paragangliomas are reported in literature. Mean age of presentation 40-50 years. Mean duration of symptoms is 2 years.

SYMPTOMS AND SIGNS:
Dysphonia is the most commonly reported symptom, followed by stridor, dysphagia, and dyspnoea. Unlike other neuroendocrine tumors of the larynx, there is a definite female preponderance (3:1). In the larynx, paragangliomas are seen as a well circumscribed smooth submucosal mass. Hence laryngeal paraganglioma should be considered in the differential diagnosis of all such masses. Of the reported cases, 90% occurred in the supraglottic larynx, but they might arise in the glottic and subglottic region. Rarely laryngeal paraganglioma is being functionally active (2.9%). Recurrence rate was 17%. Paragangliomas can infrequently have focal necrosis, mitoses, and vascular invasion. These features are not synonymous with malignant behavior.

ORIGIN
The migration of the neural crest cells take place along the paired superior and recurrent laryngeal nerves. The superior paraganglia are situated in the anterior false cord while the inferior one is found between the cricoid cartilage and the first tracheal ring.

DIFFERENTIAL DIAGNOSIS:
- Carcinoid tumors
- Granular cell tumors
- Epithelioid papilloma
- Squamous cell carcinoma
- Tumors of salivary glands

CONCLUSION:
Paragangliomas are benign tumors that mimic symptoms of malignancy. Suspicion is more towards paraganglioma when a solid swelling in the larynx is mucosa covered. Pre operative imaging is mandatory if there is clinical suspicion of paraganglioma. Direct laryngoscopy and biopsy particularly under local anaesthesia may lead to troublesome intraoperative bleeding that can be catastrophic for the patient.

REFERENCES:


