

Untreated Cleft Palate a Unique Surgical Contraindication in Carcinoma of the Pyriform Fossa

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ABSTRACT

Background/objectives: Swallowing problems following total laryngectomy are significantly lesser compared to other partial laryngectomies as a permanent separation between the trachea and esophagus is seen in the earlier. Sometimes, the patient complains of difficulty in propelling bolus through the oral cavity and pharynx as a result of the loss of hyoid bone, which is the anchor for the tongue. The increased pressure in the narrowed pharyngoesophageal segment following laryngectomy requires the tongue to move with greater force.

Setting: Department of Head and Neck Oncosurgery, Kidwai Memorial Institute of Oncology, Bengaluru.

Case report: A 52-year-old man with history of difficulty and pain while swallowing and diagnosed as squamous cell carcinoma of the right pyriform fossa extending to the pharyngoepiglottic fold. Incidentally, the patient had an old untreated cleft palate. Assessment of the pharyngeal wall involvement on direct laryngoscopy proved that pectoralis major myocutaneous flap patch was needed to reconstruct the pharyngeal lumen. Cleft palate proved a contraindication for surgery as the oral swallowing phase would be affected.

Conclusion: Current management protocols of hypopharyngeal cancers include multifactorial assessment of anatomical and functional outcomes and rehabilitation. Even though no literature is found stating cleft palate as the contraindication for total laryngectomy, chemoradiation was preferred citing swallowing problems.

Keywords: Cleft palate, Total laryngectomy, Deglutition Oral phase.

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INTRODUCTION

Hypopharyngeal malignancies are on an increasing trend in incidences in the Indian subcontinent as compared with the Western and European nations.¹⁻³ Most of these malignancies present in their advanced stages with extensive submucosal spread, high-risk of regional lymphatic involvement and relatively high propensity of distant metastases.⁴

Management modalities includes consideration of stage of the malignancy, site, age of the patient, performance status, personal and hospital protocols.⁴ Advanced hypopharyngeal cancers are usually treated surgically and

later followed by adjuvant postoperative radiotherapy (PORT) to maximize the locoregional control and improve overall survival.⁵ Recent organ preservation protocols have come up with definite chemoradiation to preserve laryngeal and pharyngeal form and function, without compromising survival.⁶

The overall results in advanced hypopharyngeal cancer with primary chemoradiation are suboptimal and inferior compared to primary surgery and adjuvant therapy.⁷⁻¹⁰ The results with the commonly advocated surgery and PORT will provide a baseline for future comparisons in the era of evolving organ-preservation strategies and intensified adjuvant therapies.⁴

Here, we present a case of carcinoma of the right pyriform fossa stage III who was treated with definitive chemoradiation with humanized monoclonal antibody for epidermal growth factor receptor (EGFR) as he had an untreated cleft palate which was a relative contraindication for total laryngectomy. The complex critical laryngeal functions during swallowing are elevation of the larynx and protection of the airway.¹¹ Initially, the larynx elevates and moves anteriorly under the base of tongue moving away from the path of the bolus and to assist in cricopharyngeal sphincter opening.¹¹ Later, it closes the airway at three levels, the epiglottis, false vocal folds and true vocal folds, hence, protecting the airway from aspiration.¹¹

Aspiration while swallowing is seen when surgery on the larynx compromises this closure especially of the true vocal folds.¹¹ Swallowing problems following total laryngectomy are significantly lesser compared with other partial laryngectomies as a permanent separation between the trachea and esophagus is seen in the earlier.¹¹ Sometimes the patient complains of difficulty in propelling bolus through the oral cavity and pharynx as a result of the loss of hyoid bone, which is the anchor for the tongue.¹¹

Usually the increased pressure in the narrowed pharyngoesophageal segment following laryngectomy requires the tongue to move with greater force.¹¹ Other factors associated are stricture at the anastomotic site causing narrowing and reduced bolus flow through the pharynx and pseudoepiglottis, a postsurgical fold of tissue from the pharynx at the level of the base of tongue, may serve as a mechanical barrier to efficient bolus flow and trap food in its pocket.¹²

CASE REPORT

A 52-year-old man with history of difficulty and pain while swallowing since 5 months was referred to the outpatient department. The patient had nasal regurgitation problem earlier and now since 5 months he has swallowing problems deep down in the throat. No history of difficulty in breathing.

On examination, the patient had a left-sided cleft lip repaired at the age of 14 and the cleft palate was not repaired. The cleft was complete with an alveolar defect (Fig. 1). Indirect laryngoscopic examination was done with a mass involving the right pyriform fossa with aryepiglottic fold involvement and the lateral wall of the pyriform fossa and pharyngoepiglottic fold with hemilarynx fixation. Left-sided vocal cords movements were normal. A right-sided level II lymph nodes were enlarged. A diagnosis of right-sided pyriform fossae lesion T3N2bMx.

X-ray lateral view of the neck showed an irregular soft tissue opacity in the region of the right pyriform fossa extending into the aryepiglottic fold. Complete blood picture revealed Hb% -12.3 gm/dl, white blood cells $7.7 \times 10^9/l$, platelets $266 \times 10^9/l$. Random blood glucose was 83 mg/dl, blood urea nitrogen was 9.5 mg/dl, serum creatinine was 0.7 mg/dl. A normal liver function test revealed total bilirubin 0.2 mg/dl, total protein 7.7 gm/dl, serum albumin 3.2 gm/dl, alkaline phosphatase 160 U/l, AST 20 U/l, ALT-40 U/l, LDH 210 U/l. Thyroid profile reported T3-1.34 ng/ml, T4- 8.5 µg/dl, TSH-5.57 µU/ml and electrolytes serum sodium 140 mEq/l, potassium 3.4 mEq/l.

Endolaryngeal biopsy revealed squamous cell carcinoma grade III. Contrast enhancing CT neck identified a soft tissue mass moderately enhancing lesion in the base of the right pyriform fossa extending to the pharyngoepiglottic fold. Right-sided level Ib and IIa lymph nodes were enlarged.

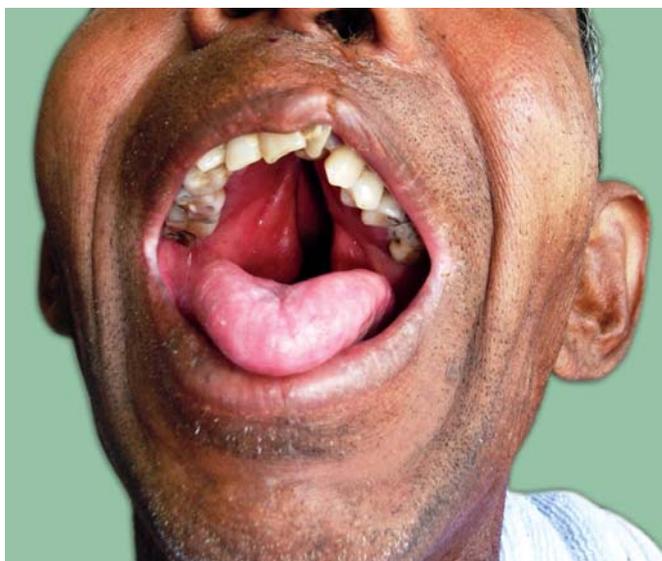


Fig. 1: Complete cleft palate seen with alveolar defect

Left-sided maxillary and ethmoidal sinuses were congested and left-sided inferior turbinate hypertrophy was seen. Complete cleft palate with the left maxillary segment hypoplastic was seen. Cardiac status was normal with 2D ECHO and color Doppler showing normal left ventricular function and no regional wall abnormalities. Electrocardiography was normal.

Assessment of the pharyngeal wall involvement on direct laryngoscopy proved that pectoralis major myocutaneous flap patch was needed to reconstruct the pharyngeal lumen. The patient had cleft palate and so postlaryngectomy swallowing difficulties were seen as contraindication for surgery. The patient was treated with concurrent chemotherapy and radiotherapy along with humanized monoclonal antibody for EGFR. The therapy included intensity modulated radiotherapy (linear accelerator 3DCRT) 50 Gy/28 fractions and 18 Gy/9 fractions with concurrent 5 cycles of intravenous cisplatin (60 mg) and 7 cycles of intravenous nimotuzumab (200 mg).

DISCUSSION

Hypopharyngeal cancer is a head and neck neoplasm with one of the most unfavorable prognosis.¹³ In developing countries including India hypopharyngeal cancers are relatively more common constituting 20 to 35% of all the head and neck cancers.¹⁴ They usually present as a advanced disease with extensive submucosal spread, high-risk of regional lymphatic involvement and relatively high propensity of distant metastases.^{5,15}

Current management protocols include multifactorial assessment of anatomical and functional outcomes and rehabilitation.¹⁶ It includes complete disease clearance and organ preservation determined by tumor extension, the amount of tissue remaining and the later plan for reconstruction.¹⁷ Surgery for pyriform fossa malignancies include removal of the total larynx and the part of the pharyngeal wall on which the tumor invaded onto and clearance of the draining interjugular lymph node levels.¹⁷

Aspiration causing swallowing problems are not seen after total laryngectomy as airway and pharyngoesophagus are physically separated.¹⁸ The greatest difficulty with deglutition is in propulsion of the bolus through the oral cavity and pharyngoesophageal segment where the cleft in the palate in our case formed a technical contraindication for the surgical procedure.¹⁸

Patients after surgery experience a mild change in tongue control of the bolus during chewing and during the oral stage of the swallow because of the loss of the hyoid bone as the foundation for the tongue.^{19,20} Also, literature indicate an increased pressure in the pharyngoesophagus after total

laryngectomy, requiring the tongue to increase the pressure it generates to propel the bolus.^{19,20}

So the oral preparatory phase dysfunction interfere in the performance of the subsequent phase by changes in swallowing biomechanical events causing stasis in the mouth and hypopharynx.¹⁷ Strictures along the pharyngo-esophagus narrows the food channel and can cause collection of food in the pharyngo-esophagus above the stricture.^{19,20} This is common when more of the pharyngeal mucosa are excised and primary closure done without attempts at reconstruction.^{19,20} If the remaining pharyngeal mucosa is smaller closure should be done as a 'T closure' technique or the flap harvested from the pectoralis major myocutaneous skin pedicle.²¹

Anatomical changes cause dysphagia in 10 to 58% of the patients and also depends on the technique used to close the pharyngeal mucosa.^{20,21} Regurgitation back into the mouth or into the nose due to the stricture is possible when the patient attempts to drink large amounts of liquids.²² An entity called pseudoepiglottis which is a fold of mucous membrane and sometimes scar tissue coming from the lateral pharyngeal wall into the base of the tongue sometimes impairs swallowing.^{23,24} The pseudoepiglottis can be diagnosed on lateral X-ray soft tissue neck but mirror examination misses it as it rests hidden below the base of the tongue.²²

But the pseudoepiglottis tends to be pulled toward the posterior pharyngeal during swallowing opening a large pocket between the tongue base and the pseudoepiglottis causing dysphagia.²² This pocket sometimes organically obstructs as it becomes difficult to pass food around it and through the pharyngo-esophagus.²² The mere presence of pockets or anatomical alteration does not indicate dysfunction.²⁵ Some remain asymptomatic for longer time, but symptomatic ones leads to regurgitation problems.¹⁷

Inability to clear these residues consequently increase the pharyngeal transit time.²⁵⁻²⁸ The lack of coordination caused by the adaptation of neopharynx constricting muscles after total laryngectomy is regarded as the important cause of dysphagia.¹⁷ The average pharyngeal transit time doubles and there is the need for a strong thrust force in order to overcome pharyngeal resistance.²⁵ Longer pharyngeal transit time were seen on videofluoroscopic evaluation with inadequate pharyngeal movement.¹⁷ Although there are no literature stating that the cleft palate as the contraindication for total laryngectomy, we gave the option to the patient explaining him about the complications about swallowing postoperatively. As the laryngeal function was compromised with hemilarynx fixation total laryngectomy with partial pharyngectomy with bilateral neck dissection with adjuvant

radiotherapy with or without concurrent chemotherapy depending on the creatinine clearance.

The preservation of laryngeal form and function is possible in a sizeable proportion of patients with hypopharyngeal cancers with primary nonsurgical approach is now an accepted option.⁴ Accurate patient selection criteria are needed to optimize treatment outcomes regarding organ preservation.⁴

Optimal organ preservation nonsurgical approach like induction chemotherapy or concurrent chemoradiotherapy, choice of drugs in these induction and concurrent regimens and radiotherapy fractionation still remains an area of active clinical trials.⁴ Also seen is a significant reduction in resting pressure and of maximal contraction in relaxation extension and movement coordination in the upper esophageal sphincter after total laryngectomy.¹⁷ Partial resection of the pharyngeal constrictor muscle and the rupture of the pharyngeal plexus, which innervates the pharyngeal constrictor muscle and the cricopharyngeal are responsible for pressure changes after surgery.²⁹

The patient shows a complete response at the primary with a 4 years disease free period and no neck nodes recurrence so far. He did not have much swallowing problems after chemoradiotherapy with 6 monthly follow-up.

CONCLUSION

Current management protocols of hypopharyngeal cancers include multifactorial assessment of anatomical and functional outcomes and rehabilitation. Even though no literature is found stating cleft palate as the contraindication for total laryngectomy, chemoradiation was preferred as difficulties in the oral deglutory phase was expected. Accurate patient selection criteria are needed to optimize treatment outcomes regarding organ preservation protocols for hypopharyngeal cancers.

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