

CASE REPORT

Youngest Case of Small Cell Anaplastic Cancer of Larynx in India

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ABSTRACT

Introduction: Small cell anaplastic carcinoma of larynx is a very rare disease. Hoarseness is the usual complain and almost many of the cases die due to spread of the disease. In this study we have tried to present the youngest case of small cell anaplastic carcinoma of larynx in India.

Case report: A patient aged 30 years presented with hoarseness of voice. 70 degree endoscopy found it to be a supraglottic swelling. Microlaryngoscopic surgery was done and specimen sent for biopsy and immunohistochemistry. Report claimed it to be small cell anaplastic cancer of larynx. MRI of the neck showed extensive involvement and metastasis to other laryngeal structures. Patient was planned for radiotherapy and chemotherapy and is on continuous follow up.

Conclusion: The patient is the youngest detected female non-smoker case of small cell anaplastic carcinoma of larynx in India.

Keywords: Youngest, small cell anaplastic carcinoma, larynx

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INTRODUCTION

Small cell anaplastic cell carcinoma of larynx is a very rare disease with mostly involving the adult age group with a complain of hoarseness. The most common association is found in smokers who are generally males.

The association of small cell carcinoma with squamous cell carcinoma both in the larynx and hypopharynx is reported.² In our case report we have tried to present a contradictory finding co-relating with small cell neuroendocrine carcinoma who is a female of 30 years. No documentation anywhere in the literature was found about such early involvement either in male or female patients in India.

CASE REPORT

Out of 585 diagnosed cases of head and neck neoplasms presented during the study period (Jul 14 – Jun15) 147 cases were of laryngeal cancers. A total of 18 cases of glottic cancer were followed up with histopathological examination after biopsies. Out of these two cases were diagnosed with neuroendocrine carcinoma. One of which was of age 58 years and did not turned up for further treatment. The other patient, a 30 year old female, presented to the out patient department, Department of Otolaryngology and Head and Neck Surgery, Assam Medical College, Dibrugarh, Assam, India, with a chief complain of hoarseness of voice since last 4 months. Her blood pressure was 110/70 mm of Hg. Laboratory data were as follows: Hb 10.2mg/dl, white blood counts 8,400 per mm³, ESR 30 mm AEFH, Differential count N₈₆L₀₉M₀₃B₀₀E₀₂, random blood sugar 113 mg/dl, urea 16mg/dl, creatinine 0.6 mg/dl, ALT 30 IU, AST 19 IU. Nasopharyngolaryngoscopy and 70 degree endoscope showed a polypoidal growth over the right false vocal cord extending to the anterior commissure, left false vocal cord, right pyriform sinus, pre-epiglottic space and right true vocal cord associated with pyriform sinus asymmetry.

TREATMENT MODALITIES

Pre-operative care

The patient was properly planned for operative procedures and pre-operative investigations included all routine investigations, liver function test, renal function test, chest x-ray, ECG, serology for infectious diseases etc

Intraoperative and anaesthetic care

A haemodynamically stable patient, during and after surgery, is the main goal of anaesthetists. The patients showed the following findings

	Blood Pressure	Pulse rate	SPO ₂
Baseline	110/70 mm Hg	98	100%
After Induction	120/76 mm Hg	88	100%
After Intubation	130/78 mm Hg	88	100%
Intra-operative	110/70 mm Hg	72	100%

Premedication was done with polonasetron, glycopyrolate, tramadol, pantoprazole and induction was done with propofol. Muscle relaxant for maintenance was done by atracurium. Reverse and extubate with neostigmine and glycopyrolate when patient was awake and responding to verbal commands.

Surgical approach

Figure: 1-2, The patient was operated by microlaryngoscopy. We can see here that a polypoidal growth is over the right false vocal cord extending to the anterior commissure, left false vocal cord, right pyriform sinus, pre-epiglottic space and right true vocal cord associated with pyriform sinus asymmetry. The suspected growth was excised and sent to laboratory for histopathological examination. During the whole operative period there was no complications.

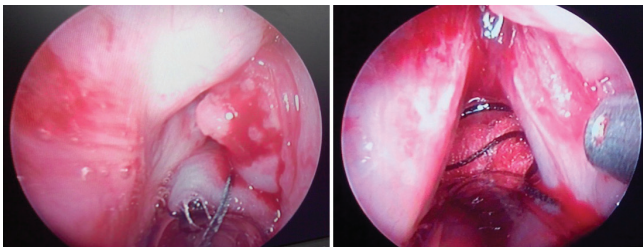


Figure-1,2: Microlaryngoscopic surgery showing polypoidal growth of glottis and post-operative.

Post-Operative Care

Post-operatively she was kept on complete voice rest. The histopathological examination report showed it to be small cell neuroendocrine anaplastic carcinoma. Immunohistochemistry was positive for chromogranin, synaptophysin and neuron-specific enolase. CEMR IMAGING of the neck showed bulkiness with T2FS hyperintense signals noted in the right false vocal cord extending to the anterior commissure, left false vocal cord, right pyriform sinus, pre-epiglottic space and right true vocal cord associated with pyriform sinus asymmetry. The lesion showed moderate heterogeneous enhancement. Few bilateral level II and level III neck lymph nodes were positive. PET CT SCAN of the head

and neck region showed minimal to mild thickening of the right vocal cord with mild enhancement of the right vocal cord, mild fat stranding in the right paraglottic region. Few bilateral level II and level III neck lymph nodes were positive. (FIG:3-4)

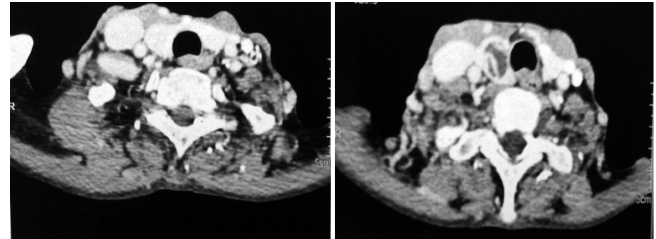


Figure-3,4: CT scan of head and neck region

Outcome

The patient by that time had already developed multiple lesions scattered throughout the liver, largest upto 12.5 x 6.2 mm in segment 4 of liver. The patient was further planned for chemotherapy and radiotherapy. The patient is on continuous follow-up.

DISCUSSION

Neuroendocrine tumors most often present in the 6th and 7th decades in men who are heavy cigarette smokers. Neuroendocrine carcinomas represent a heterogeneous group of malignant neoplasms with divergent differentiation along epithelial and neuroendocrine cell lines. They are of three variants – well differentiated, moderately differentiated and poorly differentiated. The poorly differentiated variant of neuroendocrine cancer is small cell carcinoma. Although second most common but represent fewer than 1% of all laryngeal malignant tumours. Patients with laryngeal small cell carcinoma are mainly elderly male smokers (Figure-5,6). The tumor is composed of small cells arranged in undifferentiated sheets and nests. Occasionally, ribbons of cells may be seen, as may rare neural rosette-like structures.¹

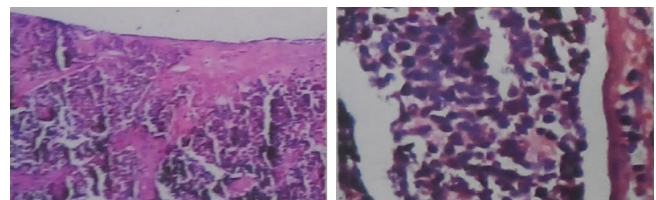


Figure-5,6: Poorly differentiated (small cell) neuroendocrine carcinoma of larynx. Undifferentiated small tumor cells with scant indistinct cytoplasm and nuclei with fine stippled chromatin and generally inconspicuous nucleoli.

The neoplasm is hypercellular with hyperchromatic,

pleomorphic, oval to spindle-shaped nuclei, increased nucleus to cytoplasm ratio, nondescript cytoplasm, and indistinct cell borders. Study done in Japan showed statistically significant difference between two groups of typical (well differentiated) carcinoid and atypical (moderately differentiated) variety was evident only in a few items such as rates of metastases, positive CEA, and multisecretory activity, while a significant difference between the carcinoid groups and SCC/OCC group was demonstrated in numerous areas, among others, e.g. 1) in the overall rates of metastases and at the sites of involvement, 2) in Grimeliusargyrophilia, 3) in immunohistochemical demonstration of positive chromogranin, CEA, calcitonin and multisecretory activity, and 4) in the 5-year survival rates. Such a definite difference between the carcinoid groups and SCC/OCC group suggested an apparently different characteristic nature present between these two series of neoplasms.³ The treatment of choice is nonsurgical and includes systemic chemotherapy and therapeutic irradiation. Small cell neuroendocrine carcinoma is a highly lethal tumor with aggressive malignant behavior. Metastases are commonly seen to regional lymph nodes in a majority of patients (60%-90%)¹, as well as to liver, lung, bone, and brain. The prognosis is poor with 2-year survival of 16% and 5-year survival of 5%.^{1,4}

CONCLUSION

Neuroendocrine small cell carcinoma of larynx is very rare. The carcinoma although known to occur mainly in elderly male smokers can also be seen in young female non-smokers. In our report we have tried to present a contraindicatory condition. However, the management and the line of treatment still remains the same. The treatment of choice is nonsurgical and includes systemic chemotherapy and therapeutic irradiation. The prognosis is also poor.

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