Online Journal of Health and Allied Sciences
Peer Reviewed, Open Access, Free Online Journal
Published Quarterly : Mangalore, South India : ISSN 0972-5997
Volume 10, Issue 1; Jan-Mar 2011

Case Report:
Undifferentiated Carcinoma of Larynx of Nasopharyngeal Type

Sandeep Bansal, Amit Shankar, Ashok K Gupta,
Department of Otolaryngology, Head and Neck Surgery, Post Graduate Institute of Medical Education and Research, Chandigarh, INDIA.

Address for Correspondence:
Dr. Sandeep Bansal,
Assistant Professor,
Department of Otolaryngology,
PGIMER, Sector 12,
Chandigarh – 160012, India.
E-mail: drsandeepb@rediffmail.com

Citation: Bansal S, Shankar A, Gupta AK. Undifferentiated Carcinoma of Larynx of Nasopharyngeal Type. *Online J Health Allied Scs*. 2011;10(1):24
URL: http://www.ojhas.org/issue37/2011-1-24.htm
Open Access Archives: http://cogprints.org/view/subjects/OJHAS.html and http://openmed.nic.in/view/subjects/ojhas.html

Submitted: Jan 31, 2011; Accepted: March 29, 2011; Published: April 15, 2011

Abstract:
Undifferentiated carcinoma of nasopharyngeal type arising in the larynx is unusual. This type of carcinoma-which occurs almost exclusively in nasopharynx-is very infrequent in the larynx (0.2%). Till date only 17 cases are reported in the medical literature. We present the clinical and histopathological findings along with the management of one additional case of undifferentiated carcinoma of nasopharyngeal type in the larynx which was managed successfully with radiotherapy.
Key Words: Nasophryngeal type of carcinoma larynx; Chemotherapy; Rare tumour

Introduction:
In 1978 the World Health Organization (WHO) classified carcinomas of the nasopharynx into three histologic subtypes[1]: squamous cell carcinoma (Type A), non-keratinizing carcinoma (Type B), and undifferentiated carcinoma of nasopharyngeal type (Type C). The WHO Type C corresponds to the lymphoepithelial carcinoma (lymphoepithelioma) Schmincke-Regaud, characterized by various degrees of lymphocytic infiltration. This tumor is essentially encountered in the nasopharynx, where surface epithelium contains indigenous lymphoid tissue. Undifferentiated carcinoma of the nasopharyngeal type is very rare in the hypopharynx and larynx. To our knowledge only a few have been reported in the English literature.[2] We present the clinical and histopathologic findings of a case of undifferentiated carcinoma of nasopharyngeal type in the larynx.

Case Report:
A 62 years old man was referred with a six months history of progressive hoarseness and occasional dysphagia and odynophagia. He complained of a progressive cough but denied dyspnea, hemoptysis and weight loss. He had a 60 pack/year history of tobacco use and a significant history of alcohol abuse. Indirect laryngoscopy showed mucosa covered growth involving left aryepiglottic fold, left arytenoid, left false cord, left true cord, and medial wall of left pyriform fossa with restricted left vocal cord mobility. There was left level II 2x3 cm hard mobile non tender lymph node. Fine needle aspiration cytology from the neck node revealed squamous cell carcinoma metastatic. The results from metastatic work up, including a CT scan of the chest and abdomen were negative. A computed tomography of neck confirmed the location of the mass and also showed involvement of the preepiglottic space and thyroid cartilage invasion (Fig 1). Endoscopy under general anaesthesia was then performed and findings were confirmed. Biopsy was taken from the representative sites which revealed undifferentiated carcinoma of nasopharyngeal type (anaplastic variant) (Fig 2). The primary tumor received a total dose of 68.4 Gy in 38 fractions of 1.8 Gy each, which were delivered over 54 elapsed days. The draining regional cervical lymphatics received a dose of 50.4 Gy in 28 fractions of 1.8 Gy, which were delivered over a similar period of time. The patient tolerated therapy with modest xerostomia and altered taste. Four months from completion of therapy, fiberoptic evaluation of the larynx showed near complete regression of the submucosal mass in the medial aspect of the left pyriform sinus. There is modest edema of the supraglottic laryngeal structures. The left hemilarynx remains fixed. There is no clinical evidence of regional nodal recurrence or any clinical evidence of distant metastases at the time of last follow-up.

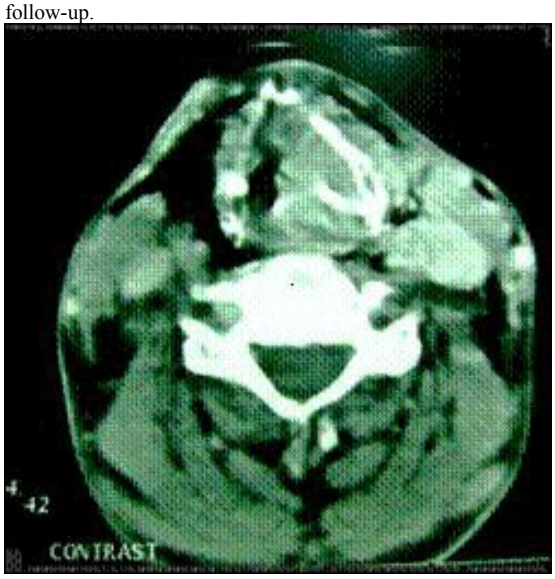

Fig. 1: Computed tomography showing left hemilarynx submucosal tumor mass

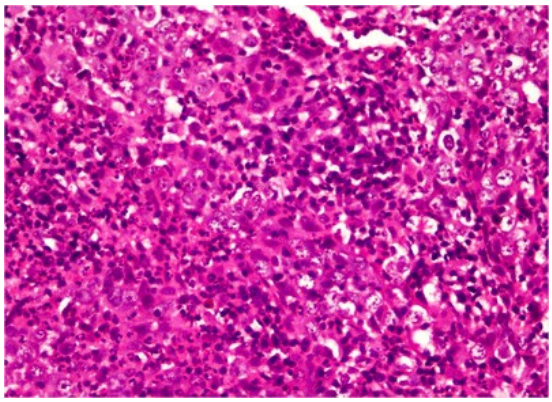

Fig. 2: H& E staining (original magnification x 25) showing pleomorphic tumor cells surrounded by mononuclear inflammatory cells.

Discussion:

Lymphoepithelioma is diagnosed most often in the nasopharynx. It compromises 40% of all nasopharyngeal tumors and is associated with Epstein - Barr virus (EBV).[3] EPV genomes are found in over 90% of nasopharyngeal carcinomas.[4] It is postulated that EBV causes lymphocytes to undergo blastic transformation, and a typical cellular arrangement of anaplastic cells dispersed among a lymphocyte infiltrate is seen. Although uncommon, lymphoepithelioma has also been reported in the nasal fossae, maxillary sinus, tongue base, parapharyngeal area, tonsil, and thymus.[3] Lymphoepithelioma of the larynx is extremely rare. Micheau et al, [2] noted a 0.2% occurrence rate from all tumors of the larynx. laryngeal lymphoepitheliomas is unknown; although it may arise from lymphatic tissue in the laryngeal ventricle. In an attempt to evaluate the modes of invasion of laryngeal and pharyngeal carcinomas, Micheau et al[2] observed a single or double laryngocele in 70% of the 2430 laryngectomy and pharyngolaryngectomy surgical specimens. Microscopic evaluation of the laryngoceles showed cylindrical or squamous epithelium with organized lymphoid tissue, typical of the histology seen in lymph nodes and the lymphoid structures of Waldeyer's ring. This laryngeal lymphoepithelial tissue has been proposed to be a true tonsil of the larynx and is potentially the site of origin of lymphoepithelioma of the larynx[1]. Alternatively, Toker and Peterson[5] postulated that these lesions may arise from active basal epithelium in the larynx, which is similar to epithelium found in tonsillar crypts.

Although smoking is not considered to be a risk factor for lymphoepitheliomas of the nasopharynx, it may play a role in the larynx. In 1921, Regaud and Schmincke each described, the pathology of lymphoepithelioma separately[3]. Regaud described nests of nonkeratinizing squamous cells embedded in a lymphoid stroma, whereas Schmincke noted isolated transitional cells scattered in lymphoid tissue which resembled a sarcoma.[3] In general, lymphocytes are believed to be a nonneoplastic component of a lymphoepithelioma. When metastasis occurs, only the epithelial component of the tumor is found at the distant site. Diagnosis can be difficult because the tumors may arise from hidden, submucosal sites. Micheau et al[2] described a patient who underwent two negative biopsies before a supraglottic laryngectomy was performed. Pathology showed a lymphoepithelioma in an intramural diverticulum without ulceration of the mucosa.' Laryngeal lymphomas may also present as submucosal masses. In a summary of 18 cases of malignant lymphoma confined to the upper aerodigestive tract, Friedberg and Has[6] reported four cases in which the lesions occurred submucosally in the hypopharynx. Most lymphomas of the larynx present as smooth supraglottic masses, usually involving the epiglottic or aryepiglottic fold.[7] Lymphomas and lymphoepitheliomas may be difficult to distinguish clinically. Lymphoepitheliom of the larynx share many characteristics with those of the nasopharynx. Both have significant potential for early regional and distant metastases. Sites of distant metastases included the mediastinum, lung, and abdomen.

Lymphoepithelioma is a highly radiosensitive tumor. Stanley et al[8] used primary radiation therapy in four patients with lymphoepitheliomas of the larynx with good local control. Ferlito[3] also reported a case of lymphoepithelioma of the larynx treated solely with radiation therapy in which his patient died in less than one year as a result of distant metastases. Our patient has had a favorable clinical and radiographic response to radiation therapy and currently shows no evidence of locoregional recurrence or distant metastases. Adjuvant chemotherapy may be useful. The value of neoadjuvant or concomitant chemotherapy for lymphoepithelioma is still unknown.

Conclusion:

Lymphoepithelioma of the larynx, although is rarely found in the larynx, it is essential to distinguish this lesion from squamous cell carcinoma. Radiation therapy is recommended as the sole treatment modality for local disease. Chemotherapy may also play a role in patients with advanced disease. Carcinoma nasopharyngeal type of the larynx must be considered as a distinct 'clinicopathological entity' and close cooperation between the clinician and the pathologist is essential for the correct diagnosis of these tumour as regards to the correct classification and therapy.

References:

1. Shanmugaratnam K, Sobin LH. Histologic typing of upper respiratory tumor, World Health Organization, Geneva. 1978.
2. Micheau C, Luboinski B, Schwaab B et al Lymphoepitheliomas of the larynx (undifferentiated carcinomas of nasopharyngeal type). *Clin Otolaryngol* 1979;4:43-48.
3. Ferlito A. Primary lymphoepithelial carcinoma of the hypopharynx. *J Laryngol Otol* 1977;91:361-367.
4. Chapman KE, Patton LL. Nasopharyngeal carcinoma: A 15-year retrospective study. *Special Care in Dent* 1993;13:163-170.
5. Toker C, Peterson DW. Lymphoepithelioma of the vocal cord. *Arch Otolaryngol* 1978;l04:161-162 .
6. Friedberg SA, Hass GM. Malignant lymphoma presenting as primary pharyngeal lesion. *Presbyt St Lukes Hosp Med Bull* 1962;1:2-7.
7. Friedman M, Toriumi D. Chapter 40. In Paparella M, Shumrick D, Gluckman J, et al (eds): Otolaryngology, Philadelphia, PA; Saunders. 1980. p 2512.
8. Stanley RJ, Weiland LH, DeSanto LW et al. Lvmphoepithelioma (undifferentiated carcinoma) of the laryngohypopharynx. *Laryngoscope* 1985;95:1077-1081.