EXTRANODAL NON-HODGKINS LYMPHOMA OF LARYNX

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ABSTRACT: Non-Hodgkins lymphoma is found in the older age group with extranodal involvement more commonly seen than in Hodgkins lymphoma. It is usually of B-cell type which has a better prognosis than T-cell type. Extranodal Non-Hodgkin’s lymphomas of larynx are rare. They can present as isolated lesions in larynx or associated with multiple involvement. They are usually found in the supraglottic region of the larynx. We present a case of 70-year-old female with extranodal Non-Hodgkins lymphoma of epiglottis with metastasis in the liver.

Keywords: Non-Hodgkins lymphoma; Mucosa associated lymphoid tissue (MALT).

INTRODUCTION
Marginal zone B-cell lymphoma of MALT-type makes up to 8% of Non-Hodgkins lymphoma found in body. These small cell lymphomas are present in extranodal sites. It was previously considered small lymphocytic lymphoma or pseudolymphoma. Currently they are grouped under the heading ‘MALTOMAS’. Among the extranodal sites, larynx is rare. Here we present a case of Marginal zone-B-cell lymphoma of epiglottis-supraglottic larynx.

CASE REPORT
A 70 year old female presented to the ENT & Head & Neck Surgery Department, Medical College, Baroda & Sir Sayaji General Hospital with complaints of difficulty in swallowing for one year; change in voice and difficulty in breathing for three months. All complaints were progressive in nature. Patient was unable to take solids and semisolids orally and developed distress on lying down in supine position. There was a past history of flexible oesophagoscopy done with a biopsy taken from a small suspicious area 12 cm from upper incisor which showed only moderate dysplasia, following which patient had not taken any form of treatment nor had she gone for follow-up oesophagoscopy.

On indirect laryngoscopic examination, a smooth, globular mass arising from laryngeal surface of epiglottis and left ary-epiglottic fold was seen which was compromising the laryngeal inlet (as seen in fig.-1). These findings were later

Fig. 1. Clinical endoscopic picture of lesion seen overhanging the laryngeal inlet.

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confirmed by flexible fibreoptic laryngoscopy. Routine hematological investigations were within normal limits. CT-scan was done to see extent of lesion, which revealed a 3 x 2.8 x 3.3 cm. size polypoid soft tissue mass arising from epiglottis, left ary-epiglottic fold and displacing left pyriform sinus. Mass was causing significant obstruction of laryngeal inlet (as seen in fig.-2).

A planned preoperative tracheostomy was done which was followed by excision of the mass. It was sent for histopathological examination which revealed diagnosis of extranodal indolent Non-Hodgkins lymphoma (fig.-3), confirmed by immunohistochemistry as marginal zone B-cell type of NHL with positive CD20, CD45 & Be12 protein with light(lambda) chain restriction. Later CT-chest and abdomen and bone marrow study was carried out. CT abdomen revealed multiple cystic lesions in both lobes of liver (fig.-4). Definitive treatment in form of chemotherapy was planned and patient is currently undergoing CHOP regime (A total of 6 cycles given 3 weekly).

CHOP regime: - Cyclophosphamide 750 mg/m²
Doxorubicin 50 mg/m²
Vincristine 1.4 mg/m²
Prednisolone 60 mg/m²

3 weeks after 1st cycle of chemotherapy; repeat flexible fibreoptic laryngoscopy was done which showed no residual mass and patient was successfully decannulated.

DISCUSSION
Extranodal marginal zone B-cell lymphoma of MALT type may occur in stomach, orbit, intestine, lung, thyroid, salivary-gland, skin, soft-tissues, bladder, kidney & CNS. Only a few have been reported till date as arising from larynx. It may present as a new mass; be found on routine imaging studies; or be associated with local symptoms (abdominal discomfort in gastric lymphomas and upper airway symptoms in laryngeal lymphomas). These lymphomas may be localized to a particular organ; however distant metastasis can occur, particularly with transformation to diffuse large B-cell lymphoma. Many patients will have an auto-immune or inflammatory process such as Sjogren’s syndrome (salivary gland lymphoma), Hashimoto’s thyroiditis (thyroid associated lymphoid tissue) or Helicobacter Pylon gastritis (gastric mucosa associated lymphoid tissue). Diagnosis must be confirmed by immunohistochemistry & thorough evaluation must be done for distant metastasis. When localized to one site, local therapy in form of radiation or surgery can effect cure. Patients who present with more extensive disease are treated with chemotherapy.

Fig. 2. Axial cut of a contrast study showing a contrast enhancing lesion arising from the epiglottis and left ary-epiglottic fold intruding into the laryngeal inlet.

Fig. 3. The slide photograph shows effacement of lymphnodal architecture with proliperation of monotonous lymphoytes of inter mediate size.

Fig. 4. Axial cut of delayed post contrast study showing multiple lesions in liver