

Laryngeal Lymphoepithelial Carcinoma: Which Specificity?

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Abstract

Undifferentiated carcinoma of nasopharyngeal type (UCNT) is mainly known in its nasopharyngeal localization. Nevertheless, it can be found in other sites of the upper and lower aerodigestive pathways. The aim of this work is to study the characteristics and therapeutic management of UCNT located outside of nasopharynx; particularly at the larynx; in the light of the medical observation of a 65-years-old who presented a laryngeal lymphoepithelial carcinoma admitted to the department of otorhinolaryngology and head and neck surgery at the specialties hospital, Mohamed V university, Rabat-Morocco.

Keywords: Lymphoepithelial carcinoma, nasopharynx, larynx, radiotherapy.

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INTRODUCTION

Malignant tumors of the nasopharynx are classified according to WHO into three histological subtypes: squamous cell carcinoma (Type I); differentiated non-keratinizing carcinoma (Type II) and undifferentiated carcinoma of nasopharyngeal type (UCNT) (Type III). The latter one presents between 65 % (North America) and 95% (China) of cases [1].

Nasopharyngeal type undifferentiated carcinoma (UCNT) occurs as a mixed, undifferentiated epithelial and lymphocytic proliferation. Only the epithelial component is neoplastic. It is well known in nasopharyngeal localization. Nevertheless, undifferentiated carcinoma is reported in other sites of the upper and lower aerodigestive pathways [1].

We report in this work a case of laryngeal UCNT diagnosed at the department of otorhinolaryngology and head and neck surgery at the specialty hospital-Mohamed V University, Rabat-Morocco.

CASE REPORT

We report a case of a 65-year-old patient with a history of 40 pack /year active smoking, stopped 10 years ago with high blood pressure and type II diabetes.

The history of his disease goes back to 9 months by the installation of dysphonia of progressive aggravation, associated since 5 months with dry cough, effort dyspnea and odynophagia. The patient was admitted for severe dyspnea that required salvation tracheotomy.

Physical examination found a sensitive and firm cervical anterior swelling in front of the thyroid cartilage. There were bilateral, mobile and firm cervical lymphadenopathies at IIb level measuring 2 to 3cm.

Pan-endoscopic examination found a voluminous ulcerated bulge ranging from epiglottis laryngeal face to the left area of the three fold and ventricular band with extension into homolateral piriform sinus. The left hemilarynx was fixed. The nasopharynx was free.

A cervical computed tomography showed a left glotto-supraglottic tissue process, poorly circumscribed, invading the homolateral piriform sinus associated to several bilateral lymphadenopathies of metastatic appearance on upper jugulo-carotid territory (figure 1). There was no tumor in the nasopharynx (figure 2).

After two negative laryngeal biopsies under general anesthesia, the patient underwent a median

thyrotomy with biopsy of the tumor. Histo-pathological examination objectified an undifferentiated carcinoma of the larynx. Tumor cells expressed cytokeratin on immunohistochemistry and the screening for EBV was negative.

The assessment of extension including, computed tomography (CT) of the chest and abdomen

were negative. The patient was staged T3N2cM0 of larynx and the therapeutic decision was to perform a total laryngectomy with bilateral functional neck dissection followed by adjuvant radiotherapy.

The patient refused any therapeutic modality and was discharged against medical advice. Three weeks later the patient died.

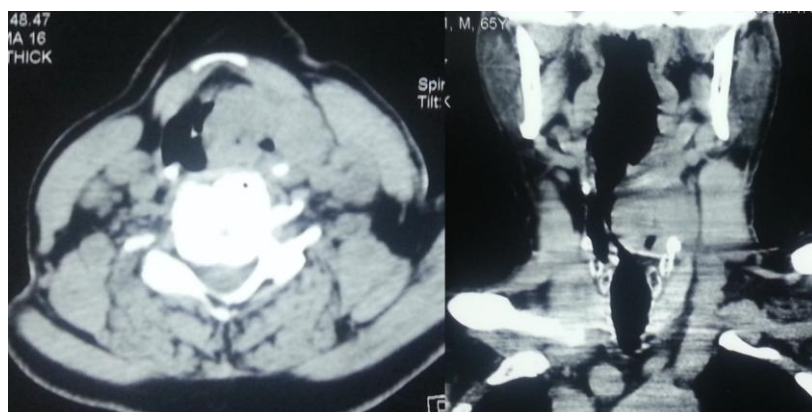


Fig-1: Images of a cervical computed tomography in axial and coronal sections showing a glotto-sus-glottic bulge of the left hemilarynx, invading the homolateral piriform sinus with the presence of bilateral upper jugulo-carotid lymphadenopathies

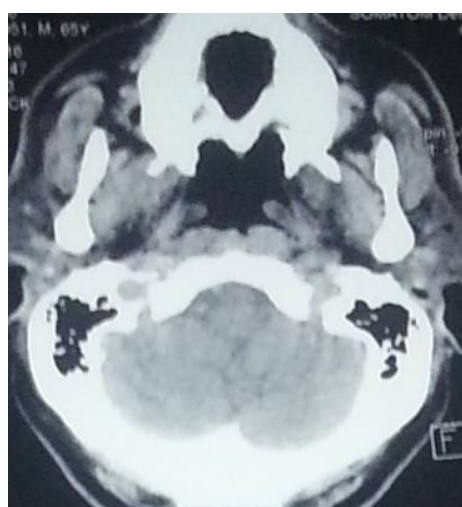


Fig-2: Scanning image in axial section objectifying absence of suspicious lesion at the level of the cavum with reliefs respected

DISCUSSION

Undifferentiated carcinoma type of nasopharyngeal (UCNT) is primarily diagnosed in nasopharynx. Characterized by its association to the Epstein-Barr virus (EBV) in more than 90% of cases but the causality has never yet clearly established [1].

Other sites have been reported outside the nasopharynx, including nasal cavities, maxillary sinus, tongue base, parapharyngeal area, tonsil, and thymus. Laryngeal localization is extremely rare. It represents between 0.06 to 0.2% of laryngeal neoplasms [2, 3].

Although a variety of appellation, such as undifferentiated carcinoma of non-nasopharyngeal type, undifferentiated carcinoma with lymphoid stroma, lymphoepithelial-like carcinoma, lymphoepithelioma, and LEC have been used for non-nasopharyngeal LEC, the term lymphoepithelial carcinoma (LEC) is the one approved by World Health Organization since 2017 [4].

According to literature, laryngeal LEC occurs mostly in older adults (mean 62 years) and predominates in males (76%) [5].

The etiology of laryngeal and hypopharyngeal LEC remains controversial [3]. Smoking and alcohol has been reported as major contributing factors. However, the cause-effect relationship was never established [6, 7]. Our patient had also a history of chronic smoking. Laryngeal LEC is less likely associated with EBV than its nasopharyngeal counterpart. Only 20% of cases seem to be related to EBV [5, 6].

Clinical diagnosis of laryngeal LEC is usually difficult. It originates mainly at hiding sites, as a submucosal bulging tumor without ulceration of covering mucosa, or with ulceration in case of advanced tumor [8].

That was the cases in our patient because we had to do a median thyrotomy in order to access the tumor and access the biopath diagnosis.

Among the common characteristics, undifferentiated carcinomas of the larynx and nasopharynx have a significant potential for early loco-regional and distant metastases regardless of the tumor size. Lymph node metastases at diagnosis are present in 75% and systemic metastases are present in 29% of cases [8]. Sites of distant metastases included the mediastinum, lung, and abdomen [7]. Our patient was so far M₀.

Undifferentiated carcinoma is a highly radiosensitive tumor. Nevertheless, there is no therapeutic consensus regarding the laryngeal localization of an undifferentiated carcinoma [9, 10]. According to cases reported and series, treatment was based on radiotherapy alone for localized tumors. Surgical treatment associated to radiotherapy was indicated in advanced tumors. The value of chemotherapy is not yet demonstrated [8, 7, 9, 10].

In our case, the therapeutic choice was to perform a surgery followed by radiotherapy. Unfortunately, the patient refused any therapeutic modality and he died three weeks later. However, our case confirms the high grade of malignancy of this particular localization

CONCLUSION

The LEC brings together a heterogeneous family of pathologies whose common point are: histology, sex ratio, radio-sensitivity, frequency of ganglion and distant metastases. The responsibility for infection with the EBV virus, widely incriminated in the genesis of the LEC of nasopharynx is much discussed outside this location.

The rarity of laryngeal LEC makes it difficult to define therapeutic modalities. That is why more studies are needed to establish guidelines for the treatment of laryngeal LEC according to TNM staging.

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