Primary Malignant Hodgkin Lymphoma of the Larynx: A Case Report

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Abstract

Background: Primary malignant lymphomas (PHML) of the larynx are exceptional. Case: A 43-year-old man presented with dysphonia and pulmonary aspiration. The physical examination found bilateral cervical lymph nodes and the pan endoscopy revealed a bleeding budding tumor of the epiglottis. The head and neck scan showed a mass arising from the epiglottis and invading the vallecules, the right aryepiglottic fold, the two vocal cords and the anterior commissure with bilateral cervical lymph nodes. The patient inderwent a chemotherapy, followed by an irradiation of the involved nodal fields and the initial laryngeal tumor. At a follow-up time of 3.5 years, the patient remains in disease complete remission. Conclusion: We report the first case of laryngeal PHML. Given the lack of recommendations for this extremely rare type of tumor. The prognosis, even if it seems to be good, it is difficult to specify whether it matches that of other localizations or not. Keywords: Laryngeal Tumors, Malignant Hodgkin Lymphoma, head and neck, Chemotherapy, Radiotherapy

Primary Malignant Hodgkin Lymphoma of the Larynx: A Case Report

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Introduction:

Head and neck malignant lymphomas are rare histological entities, which represent 5% of all malignant neoplasms of head and neck. They affect preferentially Waldeyer's ring.

Primary malignant lymphomas of the larynx are exceptional, representing <1% of all laryngeal tumors, mainly non-Hodgkin malignant lymphomas (NHML) subtype (1).

To our knowledge, no case of laryngeal Hodgkin's malignant lymphoma (HML) has been reported in the literature.

We report the first case of an epiglottic primary Hodgkin's malignant lymphoma.

Case:

A 43-year-old man with no particular pathological history presented with dysphonia and pulmonary aspiration. The patient complained of pruritus without fever, sweat or weight loss. The physical examination found bilateral fixed cervical lymph node magmas affecting all levels on the right side measuring 10cm and the level II on the left side measuring 5.5 cm . Pan endoscopy revealed a bleeding budding tumor of the epiglottis invading the right aryepiglottic fold, the right side of the three folds, the right pharyngolaryngeal wall, the right piriform sinus and the right vocal cords .The ventricles, the glottic and subglottic plane, the esophageal mouth, the base of the tongue and the valleys were free from tumor invasion.

The biopsy confirmed the diagnosis of classical mixed cellularity Hodgkin Lymphoma. Immunoisthochemical analysis revealed that tumor cells expressed CD30, CD15, fascin, and focal CD20.

The blood count, the biochemical analysis and the serum lactico-deshydrogenase (LDH) were normal. The erythrocyte sedimentation rate was 106 in the first hour,.

The head and neck scan showed a mass measuring 28*31 mm arising from the epiglottis and invading the vallecules, the right aryepiglottic fold, the two vocal cords and the anterior commissure with bilateral cervical lymph nodes :at the levels II III and IV on the right side and at the level II on the left side (Figure 1).

An extension assessment including a bone marrow biopsy and a chest abdomen and pelvic scan was negative, so that the tumor was classified as stage IIEA according to the Ann Arbor classification.

The patient inderwent a chemotherapy, which consisted on ABVD regimen (doxorubicin, bleomycin, vinblastine and dacarbazine). After four cycles of ABVD, we noted a tumor volume reduction and a complete cervical lymph nodes regression with a radiological response of 82% according to cheson. An involved field radiotherapy was delivered at a dose of 30 Gy in 15 fractions with a boost of 6 Gy at the epiglottic residual lesion in 3-Gyfractions. This irradiation was well tolerated.

An epiglottic biopsy was performed six months after the end of treatment, no longer showing tumor infiltration. At a follow-up time of 3,5 years, the patient remains in disease complete remission (clinical,radiological, histological and biological) .

Discussion:

Malignant lymphomas represent the 3rd most frequent histological subtype of head and neck cancers (12%) after squamous cell carcinoma (46%) and thyroid carcinoma (33%).

Extranodal involvement represents 23% of NHML and 1-4% of HML of head and neck (2).

Four localisations are the most affected: the Waldeyer's ring, the sinuses and the nasal cavities, the oral cavity and the salivary glands (3).

Waldeyer's ring, as it is a lymphoid structure, is the site of the most affected by head and neck extra nodal malignant lymphomas (4–8)

1) NHML of the larynx:

Primary laryngeal malignant lymphomas are rare and mainly NHML. They predominantly arise from the supraglottic region (containing follicular lymphoid tissue). A meta-analysis published by Kim et al, reported 57 cases of laryngeal NHML (1). They occur preferentially in elderly men. Clinically they most often manifest as dysphonia, dysphagia, and stridor with or without general signs (9). The diagnosis is confirmed by a biopsy. According to the World Health Organization's classification for NHMLs, type B is the most common malignant cell phenotype (10, 11).

The initial assessment includes a chest, abdomen and pelvic scan cervico-thoraco-abdominal CT and a cervical Magnetic resonance imaging, a bone marrow biopsy and a serum LDH test. The 18-Fluoro-deoxyglucose positron emission computed tomography (18 FDG TEP) is currently recommended (12). Radiotherapy is the main treatment for indolent early-stage NHML (stage I, II) at a total dose of 24 GY in 12 fractions. It must be combined with a chemotherapy in case of presence of poor prognosis factors. As for aggressive NHML, treatment is based on chemotherapy: R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) followed by consolidating radiotherapy depending on tumor response (13,14). The prognosis is similar to lymph node NHML. Ten years overall survival at is 50 - 60% (11)

2) HML of the larynx:

Primary extra nodal HML remains extremely rare and even exceptional (1-4%) (2) .To our knowledge no case of a laryngeal HML has been reported in the literature. Therefore, this case represents the first one described. The clinical presentation is not specific and very heterogeneous depending on the anatomical localization. The diagnosis is confirmed by an histological and immunoisthochemical analysis (5). The monoclonal antibodies panel used includes CD15 (Leu M1), CD20 (L-26), CD30 (Ber H2), CD45 (LCA), CD45RO (UCHL-1), latent membrane protein type 1 (LMP-1 ; CS1-4) of Epstein-Barr virus (EBV) and polyclonal CD3 and fascin (4,5). The most common subtype is classical mixed cellularity lymphoma (2,4). However, Quinones-Avila et al (5) suggested that the classical nodular-sclerosis and lymphocyte-rich Hodgkin Lymphoma subtypes are more common. This could be explained by the fact that in previous studies lymphomas diagnosis was only based on the histomorphological characteristics and old malignant lymphomas classification systems were used.

Head and neck HML treatment consists in ABVD chemotherapy regimen followed by a radiotherapy at a total dose of 25 to 40 Gy, targeting the Waldeyer ring and the involved cervical lymphnodes fields (8).

This treatment provides excellent disease long-term local control with similar results to those reported in cases of lymph node HML. According to the results of a cohort of 34 patients, head and neck extra nodal HML treated with ABVD or MOPP regimens chemotherapy (mechlorethamine, vincristine, procarbazine, and prednisone) followed by radiotherapy at a total dose of 39.6 Gy, in the MD Anderson Cancer Center at the University of Texas between 1967 and 2007, the 10-year disease-free survival rate was 71% (2).

Conclusion:

Head and neck HML are exceptional and the laryngeal localization is unusual. Despite its exceptional nature, it is a diagnosis to be systematically evoked and confirmed by histological and immunoistochemical analysis. This case reveals that the treatment could be the same as the other HML localizations , given the lack of recommendations , but the prognosis , even if it seems to be good, it is difficult to specify whether it matches the other localizations or not .

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