CASE STUDY

Primary MALT Lymphoma of the Larynx

Linfoma tipo MALT primario de laringe

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Clinical Case

A 76 year old woman was referred to our unit after presenting with intermittent dysphonia of 1 year duration. There was neither a relevant medical history nor did she have any toxic habits such as consumption of alcohol or tobacco. There were no B symptoms.

Clinical nasopharynx, Waldeyer ring, and cervical examinations all showed normal results; the nasofibroscopy performed revealed a single laryngeal polyp in the left vocal cord (Fig. 1) with a preservation of laryngeal movement. Findings from a blood test and a general clinical examination were normal. Laryngeal microsurgery with the use of suspension laryngoscopy according to the Kleinasser technique was indicated and the before-mentioned polyp was extirpated. The subglottic region, the supraglottis, and the hypopharynx were disease free. A bronchoscopy was also performed which also resulted normal.

The biopsy from the anatomopathological study showed a lymphoid infiltration of the tissue with characteristics which are typical of MALT type extraganglionar B cell lymphoma (Fig. 2) and the immunohistochemical staining confirmed B lymphocytes (CD20 + CD79 + BCL + BCL6, CD5, CD3, CD23, D1 cyclin). The proliferation index (MIB-1) was low. PCR expansion of IGH reordering, in accordance with BIOMED-2 concerted action protocol in tumor sample showed a monoclonal B cell population. Resection margins were tumor free.

As a result of these findings a computed tomography (CT) scan was requested of the cervical and chest area which resulted negative for cervical adenopathies and/or mediastinal tumors; we also consulted with members of the digestive disease unit who perform esophagogastroscope for the confirmation of gastroesophageal reflux disease symptoms. Gastric biopsies were performed which resulted
negative for MALT lymphoma; chronic interstitial gastritis and Helicobacter pylori (H. pylori) infection was confirmed.

The patient was diagnosed with MALT Ann Harbor stage IE MALT lymphoma in the vocal cord.

Eradication therapy for the H. pylori was initiated with Clarithromycin, Amoxicillin, and Omeprazol, according to protocol.

No evidence of recurrence was observed after an intense follow up of 3 years.

Discussion

Mucosa associated lymphoid tissue (MALT) is a type of non-Hodgkin lymphoma. The staging of it assesses the level of spread (I–IV), and the presence of systemic symptoms: (A) absent and (B) any of the following: fever lasting 1–2 weeks, weight loss, perspiration (E) tumor located in an organ or tissue which does not form part of the lymphatic system (S) tumor in the spleen. The importance of this classification is its value in prognosis. It is one of the most frequent lymphomas in adults and accounts for between 5% and 10% of gastric neoplasias. Despite its representation of just 2%–3% of lymphomas, its location in the gastric system represents 70% of extraglandular locations; other MALT lymphoma sites are: lung, head and neck, thyroid glands, skin, breast, and other areas of the digestive tract.

Primary MALT lymphoma is a rare disease which represents below 1% of malignant tumors of this location. At least 25 cases have been described in literature from its initial appearance in literature in 1990 with supraglottic location being 77.3%, subglottic 18.2%, and glottis less than 5%. It occurs most frequently in middle age (50 years of age), there are no gender differences, and symptoms which appear may vary depending on location and size. The great majority are associated with heliobacter pylori gastroesophageal reflux.

The etiology of this type of lymphoma relates to a background of chronic inflammatory disease or autoimmune disease; for this reason, MALT lymphoma is associated with Sjögren’s syndrome, systemic lupus and particularly erythematous, Hashimoto thyroiditis, and gastritis associated with H. pylori.

Physiopathologically, the H. pylori gastric tissue causes inflammation of the mucosa which stimulates lymphocytoid aggregation and in some cases this process may become chronic and degenerate into MALT type lymphoma.

Cytogenetic studies have shown serious anomalies in these tumors with translocations (11; 18) (q21, q21) those most frequently found in approximately 50% MALT lymphomas and which result as positive in immunohistologica studies for CD20, CD21, CD35, and IgM; and negative for CD5, CD10, CD23, and D1 cyclin (−). Histopathologically, MALT lymphomas are heterogeneous and cover a cytological spectrum which may lead to centrocyclic cells, smaller lymphoid cells or monocytoid cells. One major characteristic is the presence of lymphoepithelial tumors formed by the invasion of lymphocytes to different tissues in this case the glottis mucosa tissues.

Almost all cases of primary MALT lymphoma of the larynx show a disease history of gastroesophageal reflux and/or H. pylori infection.

There is no specific treatment for gastric MALT lymphomas. The majority of them are treated with disease targeted radiotherapy and with disease disseminated or systemic chemotherapy. The most frequently applied treatment in the larynx is chemotherapy and
radiotherapy (28–50 Gy). In the latest studies and reviews F-fluorodesoxyglucose positron emission tomography/computed tomography (F-FDG-PET/CT) has proven to be not merely useful in the diagnosis and staging of this disease but also in providing information on response to radiotherapy when it is indicated with doses under 28 Gy.

Three cases have been described in literature where tumor removal and eradication of H. pylori were the only treatments used, but none were located in the glottis.

In our case, after complete resection of the laryngeal tumor, there was no evidence of systemic compromise; eradication of the H. pylori was confirmed and the gastroesophageal reflux was successfully managed. After excision there was no evidence of recurrence. In general, low grade MALT lymphomas have a high response to treatment and recurrence is rare. We therefore believe that conservative treatment combined with treatment of the disease with gastroesophageal reflux and eradication of the H. pylori disease could be an option, provided that complete surgical extirpation is possible.

Conflict of Interests

The authors have no conflict of interests to declare.

References


