Abstract. Background: More than 90% of laryngeal tumors are squamous cell carcinomas. Primary hematopoetic neoplasms of the larynx are rare, being mainly extramedullary plasmocytoma and non-Hodgkin’s lymphoma (NHL). These are mainly located in the supraglottic and glottic area, with only a few reported in the subglottic region. Case Report: We report on a 58-year-old man, who presented at our clinic with severe dyspnea. On microlaryngoscopy, a subglottic stenosis at the level of the cricoid cartilage was found. The biopsy revealed the diagnosis of a MALT-type lymphoma (marginal zone B-cell lymphoma). The tracheostomy was followed by locoregional radiotherapy. Conclusion: This is the first report of a subglottic MALT-type lymphoma causing a subglottic tracheal stenosis. The preferred treatment is locoregional radiotherapy including the draining lymph nodes.

More than 90% of all malignant tumors of the larynx are squamous cell carcinomas, whereas primary malignant manifestations of the hematopoetic system in the larynx are extremely rare. Most of these cases are extramedullary plasmocytomas (EMP) or non-Hodgkin’s (NHL) lymphomas (17). The majority of these cases are B-cell lymphomas, whereas laryngeal T-cell lymphomas are uncommon (5, 20). A special form of the extranodal NHL is the marginal zone B-cell lymphoma (MALT-lymphoma). This disease usually occurs in late adult life and occasionally in childhood (17, 26). Hoarseness is often one of the first symptoms, whereas dyspnea is only found in advanced glottic or subglottic cases (26). Dysphagia is also a common complaint. Most of the described cases affect the supraglottic region (17). To date, only one case of a glottic MALT-lymphoma has been published in the literature (4). We here report on the first case of a subglottic marginal zone B-cell lymphoma (MALT-lymphoma) causing a subglottic tracheal stenosis.

Case Report

Anamnesis. In November 2003, a 58-year-old male patient presented with a two-month history of progressive dyspnea. The patient, suspected of having a subglottic tracheal stenosis, was sent to our hospital for treatment by his ENT-specialist. Clinical assessment showed no evidence of airway damage due to long-term intubation, a preceding trauma, an operation of the thyroid gland or a tracheostomy. The medical history revealed a left-sided seminoma, which had been treated in 1998 by surgical semi-castration followed by a postoperative radiotherapy.

Clinical findings. The direct laryngoscopy showed redness of the subglottic mucosa and a subglottic stenosis. Both vocal cords and the supraglottic region showed no pathological findings (Figure 1). Standard laboratory tests including anticytoplasmatic antibodies (ANCA’s) were within normal range. Lung function tests showed characteristic signs of upper airway stenosis. Computer tomography of the neck revealed a subglottic tumor in the area of the posterior wall of the cricoid cartilage. This mass of 1.9 cm x 1.5 cm showed soft tissue characteristics, narrowing the tracheal lumen from dorsal (Figure 2). There were no enlarged cervical lymph nodes. Microlaryngoscopy and bronchoscopy confirmed the subglottic tracheal stenosis in the area of the cricoid cartilage with lumen stricture of approximately 70%. The vocal cords were slightly swollen without any evidence of pathological findings (Figure 1).

Histological findings. Histology from a biopsy of the subglottic area showed a diffuse submucosal infiltration of monomorphous lymphoid cells, with small to medium-sized nuclei of a so-called centrocyte-like morphology with very few interspersed blastoid cells, devoid of reactive follicles. Immunohistochemical staining revealed positivity for CD20, CD79a and Bcl-2, with monotypic expression of kappa light chain and negative expressions of CD3, CD5, Cyclin D1,
CD10, CD30 and Bcl-6 (Figures 3-5). These features indicated a marginal zone B-cell lymphoma (MALT-lymphoma).

**Therapy.** The patient was treated by local radiotherapy including the regional lymph nodes, based on the clinical classification stadium IE. The follow-up after 12 months showed no evidence of local recurrence of the lymphoma. The tracheostoma was closed after 8 months.

**Discussion**

Early in the eighties, Isaacson et al. introduced the “MALT-concept” representing “mucosa-associated lymphoid tissue” of epithelial, mainly gastrointestinal organs, secondarily acquired through infections or immunological stimuli. This concept...
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The first MALT-lymphoma of the larynx was described by Diebold et al. in 1990 (11), and subsequently only a few cases of primary MALT-lymphomas of the larynx have been published (9, 17, 20). Seventy-five % of these cases occurred on the left side of the larynx, predominantly located in the supraglottic area, in particular in the epiglottic and aryepiglottic fold (17). Kutta et al. were able to demonstrate, in cadavers, that MALT-tissue of the larynx (LALT) is predominantly located in the supraglottic fold, detectable in 100% of all children and in 90% of all adults examined (23). Additionally, with increasing age a decreasing frequency could be proved. Interestingly, no LALT-tissue could be detected in the subglottic region. This could be a possible explanation for the fact that all previously described primary laryngeal MALT-lymphomas were found in the supraglottic region. To date, the trachea as a primary localization of a B-cell lymphoma has been described in only a few cases (21). A typical macroscopic finding of a laryngeal MALT-lymphoma is submucous swelling without ulceration (11).

For the treatment of MALT-lymphomas, the various characteristics of the disease have to be considered: the localization of the lymphoma and the status of the disease as localized or disseminated. At the time of first diagnosis, 60% to 70% of all patients are staged I or II (2, 29). The therapy of first choice in localized stage (I, II) is involved-field-radiotherapy (25-30Gy). Advanced stages with disseminated organ affection (Stage III and IV) or recurrent disease are treated with chemotherapy, occasionally in combination with radiotherapy (5, 22, 25, 26, 31). Other therapy options used in clinical trials are the local application of IFN α2a (32), the systemic application of the chimeric monoclonal anti-CD20-antibody Rituximab or the radio-labelled anti-CD20-antibody (6, 7, 13). According to Zinzani et al., a combination of radiotherapy and chemotherapy provided the best results in most cases, with only a few patients benefiting from locally applied IFN α2a (32). The therapeutic impact of chemotherapy has yet to be tested by appropriate studies (7). Determinative for prognosis is the clinical stage (26). The overall prognosis of MALT-lymphomas, compared to other types of B-cell lymphomas, is good, because of the satisfactory response to therapy (8, 32). Bone marrow involvement results in a worse prognosis (32).

In summary, we have described the first primary MALT-lymphoma of the larynx located in the subglottic region causing a subglottic tracheal stenosis. The therapy of first choice is local radiotherapy including the draining lymph nodes, combined with tumor debulking in the case of airway constriction. Only in the case of systemic disease is a systemic therapy indicated. Compared to other B-cell lymphomas, the prognosis is very good, with a 5-year survival-rate of 80%.
References


