Extranodal marginal zone lymphoma (MALT) of the larynx and orbit

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Marginal zone lymphomas are a subtype of B-cell lymphomas, most often of indolent course. There are three main subtypes of marginal zone lymphoma: extranodal or MALT (mucosa-associated lymphoid tissue), nodal, and splenic. MALT lymphomas arise in non-lymphoid organs after prolonged antigenic stimulation, e.g. H. pylori in the stomach, Chlamydia in the ocular adnexa, Borrelia in the skin and in the context of HCV infection. One of the rarest reported sites of MALT lymphoma is the larynx. We present the case of a 71-year-old woman who presented for ENT evaluation due to a voluminous lesion of the epiglottis and the left palatine tonsil, described as an incidental finding on orbital MRI performed for the swelling of the left upper eyelid. On ENT fiberendoscopic examination, a rounded, smooth-surfaced lesion measuring 1.5 cm was seen on the lingual surface of the epiglottis, extending to the left lateral wall of the hypopharynx, filling the vallecula and overhanging the laryngeal inlet; the vocal cords showed changes consistent with Reinke's edema. The neck was painless and without palpable lymph nodes. An indurated lesion of the upper left eyelid measuring 3 cm was also noted. Additional radiological evaluation demonstrated an extensive submucosal solid tissue process involving the left side of the larynx, filling the paraglottic space and protruding medially into the ventricular fold. Since the eyelid lesion exhibited the same radiological characteristics as the laryngeal lesion, fine-needle aspiration of the left upper eyelid was performed, followed by biopsy. Cytological analysis of the aspirate and flow cytometry, as well as histopathological examination of the eyelid biopsy, confirmed an extranodal marginal zone lymphoma (B-NHL, EMZL). The patient underwent immunochemotherapy combined with radiotherapy of the left eyelid. The treatment achieved clinical regression of the disease, confirmed by PET/CT, with the regression of both the laryngeal and eyelid lesions. During therapy, the patient was regularly followed by an otorhinolaryngologist to assess treatment response and airway patency. Maintenance therapy with rituximab was planned. The aim of this report is to highlight the rare manifestation of MALT lymphoma in the larynx, which should be considered in the differential diagnosis of laryngeal tumors.

Keywords: MALT, larynx, orbit