A Rare Case of Marginal Zone Lymphoma Presenting with Bilateral Ear Erythema and Swelling

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Marginal Zone Lymphoma (MZL) accounts for between 5% and 17% of all non-Hodgkin lymphomas (NHL). We report a rare case of marginal zone lymphoma presenting with bilateral ear erythema and swelling. The case was a 79-year-old man who suffered from progressive bilateral ear erythema and swelling since 2 years ago. Physical examination revealed for erythematos, edematous and non-tender auricles bilaterally which affected both lobules. He was scheduled for a punch biopsy and pathology report revealed some atypical cells with large vesicular nuclei, prominent nucleoli, with clear cytoplasm (monocytoid B-cells) in mid-dermis with extension to deep dermis favoring low grade B-cell lymphoma, marginal zone subtype. Computed tomography of the chest revealed lymphadenopathy in both hilar, subcarina, aortopulmonary and retrocaval space greater than 10 millimeter. In conclusion, involvement of the ears is an unusual finding in malignancy and therefore tissue biopsy must be performed, even if it occurs bilaterally.

Keywords: Cancer risk, Compound BTEX, Automobile manufacturing, Painters
Introduction

Marginal Zone Lymphoma (MZL) accounts for between 5% and 17% of all non-Hodgkin lymphomas (NHL) in adults depending on the series. Extranodal marginal zone lymphoma (EMZL) of mucosa-associated lymphoid tissue (MALT) lymphoma is a mature B-cell neoplasm that typically follows an indolent clinical course. Most MALT lymphoma patients present with an indolent disease with good performance status, absence of B-symptoms, and no adverse biological prognostic factors such as high lactate dehydrogenase (LDH) or β2-microglobulin levels. Dissemination of the disease occurs either to other mucosal sites or, more often, by extension on a mucosal site to a non-mucosal site such as spleen, bone marrow, or liver. Bone marrow involvement is detected in 20% of cases. Risk of dissemination is significantly higher for non-gastrointestinal tract lymphomas.

Herein, we report a rare case of marginal zone lymphoma presenting with bilateral ear erythema and swelling.

Case Report

The patient was a 79-year-old man who suffered from progressive bilateral ear erythema and swelling since two years ago. He denied any trauma, fever, chills, night sweating and significant weight loss; he also had good performance status. His past medical history was unremarkable except for chronic hypertension, for which he took Atenolol 100 mg twice a day. He was referred to an otolaryngology clinic for evaluation. Physical examination revealed erythematous, edematous and non-tender auricles bilaterally which affected both lobules (Figure 1). Nasal deformity was not found and physical examination of the cardiopulmonary system was unremarkable. He did not have peripheral lymphadenopathy, but his spleen was palpable 5 cm below the ribs on the midclavicular line. He was scheduled for a punch biopsy and pathology report revealed some atypical cells with large vesicular nuclei, prominent nucleoli, with clear cytoplasm (monocytoid B-cells) in mid-dermis with extension to deep dermis favoring low grade B-cell lymphoma, marginal zone subtype (Figure 2). Ki-67 proliferation index showed 20% of tumor cells with positive nuclear stain. Immunohistochemistry (IHC) staining was positive for CD20 in tumoral cell and CD3 in some cells. Complete blood count (CBC) showed a normocytic anemia (hemoglobin level of 9.4 g/dL) with counts of white blood cells and platelets within normal limits. Bone marrow aspiration and biopsy were performed according to consultation with a hematologist and showed normocellular marrow with increased plasma cell. Computed tomography of chest revealed lymphadenopathy in both hilar, subcarina, aortopulmonary and retrocaval space greater than 10 mm. Similarly, there was a prominent lymph node in hilar region of liver and spleen and left paraaortic space. Spleen was 190 mm in size. The patient was referred to an oncologist for further evaluation and treatment, but the patient refused medication.

Discussion

Malignant lymphoma can spread to all areas of the body, they account for approximately 5% of all malignant neoplasms of head and neck and may involve nodal or extranodal sites. The head and neck region is the second most frequent anatomical site of extranodal lymphomas (after the gastrointestinal tract). Two articles have reported extranodal lymphoma with middle ear and auditory canal involvement. Involvement of the temporal bone as part of generalized lymphoma has been reported; however, clinical evidence of temporal bone or ear involvement is unusual. Malignancy can involve auricles,
but it rarely occurs bilaterally; also, MZL presenting with bilateral ear inflammation is a rare condition\textsuperscript{11}. Our case had MZL, presenting with bilateral ear involvement. Although clinical signs and symptoms are typically enough for a diagnosis of auricular inflammation causes, a biopsy may be required for a definitive tissue diagnosis\textsuperscript{11}. Our patient complained of progressive bilateral ear erythema and swelling. Therefore, these complaints can be symptoms of malignancy such as lymphoma of the ear. Definitive diagnosis of non-Hodgkin lymphoma needs incision or excisional biopsy, flow cytometry, IHC and molecular tests\textsuperscript{12}.

**Conclusions**

Ear involvement is a rare condition in marginal zone lymphoma, and bilateral involvement of the ears is a

![Image of patient’s auricles](image.png)

**Figure 1**: Image of patient’s auricles (A) Right side view (B) Front view
very rare presentation. If bilateral ear swelling is observed, malignant causes such as lymphoma should be considered and therefore tissue biopsy should be done.

**References**


**Figure 1.** (A) Monocytoid B cells with clear cytoplasm and plasma cells (x100), (B) Diffuse infiltration of the mid and deep dermis by a relatively monomorphic population of medium size lymphoid cells (x40), (C) Positive immunohistochemical reaction of tumor cells by CD20(x40)