CLINICAL IMAGE

Richter transformation of chronic lymphocytic leukemia presenting as otitis externa

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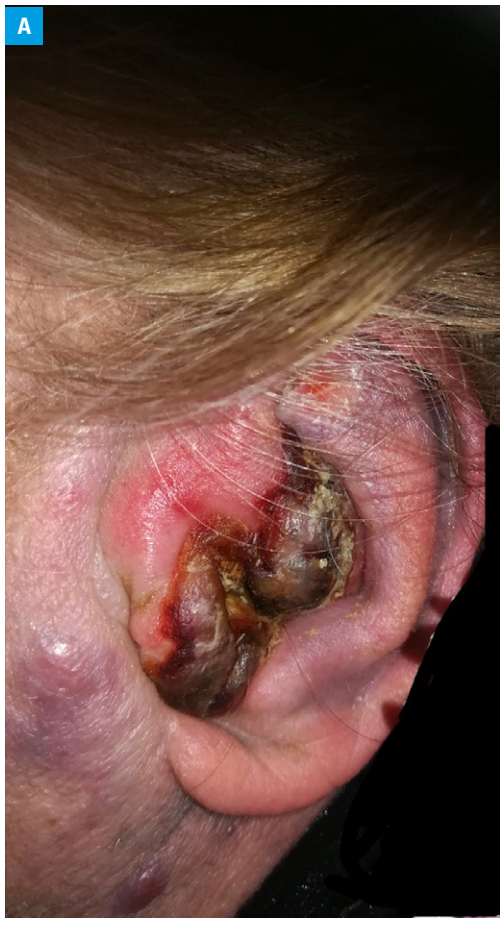
Acute otitis externa is a common condition which generates a high volume of urgent ear, nose, and throat (ENT) consultations, also during the COVID-19 pandemic. In this setting, a 75-year-old woman was referred for an outpatient ENT consultation due to ear pain and hearing loss and diagnosed with acute otitis externa. The patient had been diagnosed with chronic lymphocytic leukemia (CLL) 3 years earlier (Rai stage I, Binet stage A). Following a sharp increase in lymphocyte number, chemoimmunotherapy with fludarabine, cyclophosphamide, and rituximab was initiated 1 month previously. Despite the administration of combined systemic antimicrobial and topical treatment, the patient deteriorated (FIGURE 1A) and was referred for hospitalization. Meanwhile, she developed marked regional lymphadenopathy.



Diagnostic workup on admission to hospital did not identify the specific pathogen and the patient received empiric antimicrobial treatment with meropenem and clindamycin. Biopsy of the external auditory canal revealed diffuse infiltration of the cutaneous lamina propria by neoplastic lymphoid cells with histologic and immunohistochemical characteristics compatible with high-grade non-Hodgkin lymphoma (NHL) (FIGURE 1B). The patient developed sepsis and died within 3 weeks of hospitalization.

Numerous conditions may be misdiagnosed as otitis externa, and special considerations apply to immunocompromised patients.[1] Chronic lymphocytic leukemia is the most frequently encountered type of leukemia in Western countries. It affects predominantly the elderly and is characterized by a highly heterogeneous prognosis. Transformation of CLL to an aggressive lymphoma (termed *Richter transformation*) is rare, with a cumulative 5-year incidence of 2.8% and a dire prognosis.[2,3] Reports of skin or ear infiltration are scant; prompt recognition may be key to prolong survival.[4]

FIGURE 1 A – erythema, extensive oedema, and skin thickening causing obstruction of the left external auditory canal. Skin necrosis developed in the course of hospitalization.

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Received: February 20, 2021.
Revision accepted: March 22, 2021.
Published online: March 26, 2021.
Pol Arch Intern Med. 2021;
131 (5): 469-470
doi:10.20452/pamw.15898

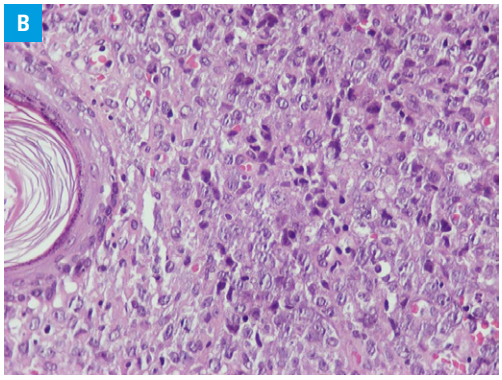

FIGURE 1 **B** – histological examination of the external auditory canal showing diffuse infiltration of the cutaneous lamina propria by neoplastic lymphoid cells of medium to large size, with high nucleocytoplasmic ratio, ovoid nuclei, and large nucleoli (hematoxylin and eosin staining, magnification × 400)

Localization of NHL in the external auditory canal is generally a rare occurrence and misdiagnosis as otitis externa is a common pitfall.[5] Although the clinical presentation further depends on the extent of disease and infiltration of adjacent anatomical structures, the most common clinical symptoms include severe otalgia, otorrhea with exudative discharge, erythema and oedema of the ear canal, sensorineural hearing loss, difficulty with mastication, and regional lymphadenopathy.[5] The main therapeutic approach is immunopolychemotherapy; however, the management differs by NHL subtype, among other factors.

ARTICLE INFORMATION

PATIENT CONSENT The patient provided informed consent to publish the photographs.

CONFLICT OF INTEREST None declared.

HOW TO CITE Pilalas D, Daios S, Fotiadou A, et al. Richter transformation of chronic lymphocytic leukemia presenting as otitis externa. Pol Arch Intern Med. 2021; 131: 469-470. doi:10.20452/pamw.15898

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