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Article type: Clinical image

Received: February 20, 2021.

Accepted: March 22, 2021.

Published online: March 26, 2021.

ISSN: 1897-9483

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Richter transformation of chronic lymphocytic leukaemia presenting as otitis externa

Short title: Richter transformation of CLL presenting as otitis externa

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Conflict of interests: none declared.
Acute otitis externa is a common condition which generates a high volume of ear, nose and throat (ENT) urgent consultations even during the COVID-19 pandemic. In this setting, a 75-year-old female patient was referred for an outpatient ENT consultation for ear pain and hearing loss and diagnosed with acute otitis externa. The patient had been diagnosed with chronic lymphocytic leukaemia (CLL) 3 years ago (Rai stage I, Binet stage A).

Following a sharp increase in lymphocyte number, chemoimmunotherapy with fludarabine, cyclophosphamide and rituximab was initiated one month ago. Despite administration of combined systemic antimicrobial and topical treatment, the patient deteriorated (Figure 1A) and was referred for hospitalization. Meanwhile, the patient developed marked regional lymphadenopathy.

The inpatient workup did not identify a pathogen and the patient received empiric antimicrobial treatment with meropenem and clindamycin. An external auditory canal biopsy 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 three weeks from hospitalization.

Numerous conditions may be misdiagnosed as otitis externa, while special considerations apply to immunocompromised patients [1]. CLL is the most frequently encountered leukaemia in western countries affecting predominantly the elderly and is characterized by a highly heterogeneous prognosis. Transformation of CLL to an aggressive lymphoma termed Richter transformation (RT) 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].

NHL localization in the external auditory canal in general is 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 and signs include severe otalgia, otorrhea with exudative discharge, ear canal erythema and oedema, 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.

**Contribution statement:** GK conceived the paper, provided relevant data as the treating hematologist. DP and SD performed the literature search and wrote the first draft. AF provided Figure 1B and the relevant analysis. KM provided Figure 1A and was the lead physician during patient hospitalization. CS and GK authors provided critical feedback. All authors helped shape and approved the final manuscript. GK is the guarantor.

**Funding:** This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

**Patient consent for publication:** Obtained.
References


Figure 1. A. Erythema, extensive oedema and skin thickening causing obstruction of the left external auditory canal. Skin necrosis developed in the course of the hospitalization. B. External auditory canal biopsy with 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 (haematoxylin-eosin, 400x).