

Unusual Presentation of Follicular Lymphoma with the Involvement of Bilateral Ear Helices and Lobes

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ABSTRACT

Follicular lymphoma is a type of systemic lymphomas which constitutes approximately 30-35% of all Non-Hodgkin lymphomas. It typically presents itself in the form of generalized lymphadenopathy, hepatomegaly, splenomegaly and bone marrow involvement. Cutaneous involvement of follicular lymphoma generally appears as skin-coloured to red, violaceous papules or nodules most commonly involving the scalp, trunk and head&neck region. Herein, we would like to present an unusual case of follicular lymphoma which appears as skin-coloured papules prominent upon the both ears and trunk.

Keywords: follicular lymphoma, ear, skin involvement.

INTRODUCTION

Cutaneous involvement of B-cell lymphomas may present itself in the form of indurated, confluent papules and nodules involving the scalp, trunk and ears. We would like to share an extraordinary case of follicular lymphoma (FL) presenting as skin-coloured, firm papules prominent upon the both ears.

CASE PRESENTATION

A 70-year old man with a history of diabetes mellitus and hypertension, was consulted to our outpatient clinic with the complaint of asymptomatic, flesh-

coloured papules involving bilateral ears, the back and left upper arm. He had realized the lesions three months ago and new infiltrated papules continued to show up subsequently since then. No systemical symptoms such as fever, weight loss, fatigue or diaphoresis were accompanied. He was using metformin hydrochloride, gliclazide, linagliptin, insulin and doxazosin for diabetes mellitus and hypertension with no recent medicine initiation. Dermatological examination revealed pink to flesh-coloured, shiny, confluent papules on the bilateral ear lobules, discrete papules involving the bilateral ear helices, left upper arm and the back (Figure 1). Pseudolymphoma, primary cutaneous

B-cell lymphoma, generalized eruptive histiocytosis and cutaneous amyloidosis were considered as differential diagnosis. A 4 mm punch biopsy was taken from the left upper arm and showed lymphoid infiltration in a diffuse pattern, leaving a grenz

zone under the epidermis progressing up to the subcutaneous fat tissue in focal areas. The lymphoid infiltrate consisted of small-medium size, narrow cytoplasm, coarse chromatin pattern and irregular nuclear contour and large nuclear slit/notched cells

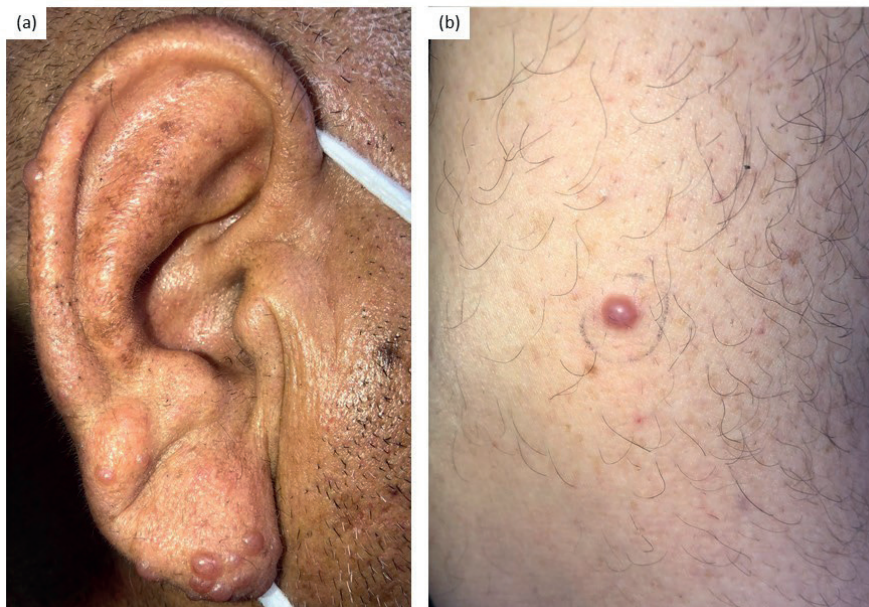


Figure 1. Flesh-coloured, pinkish, discrete papule on the ear helix and confluent papules on the ear lobule (a), discrete, shiny papule on the left upper arm (b).

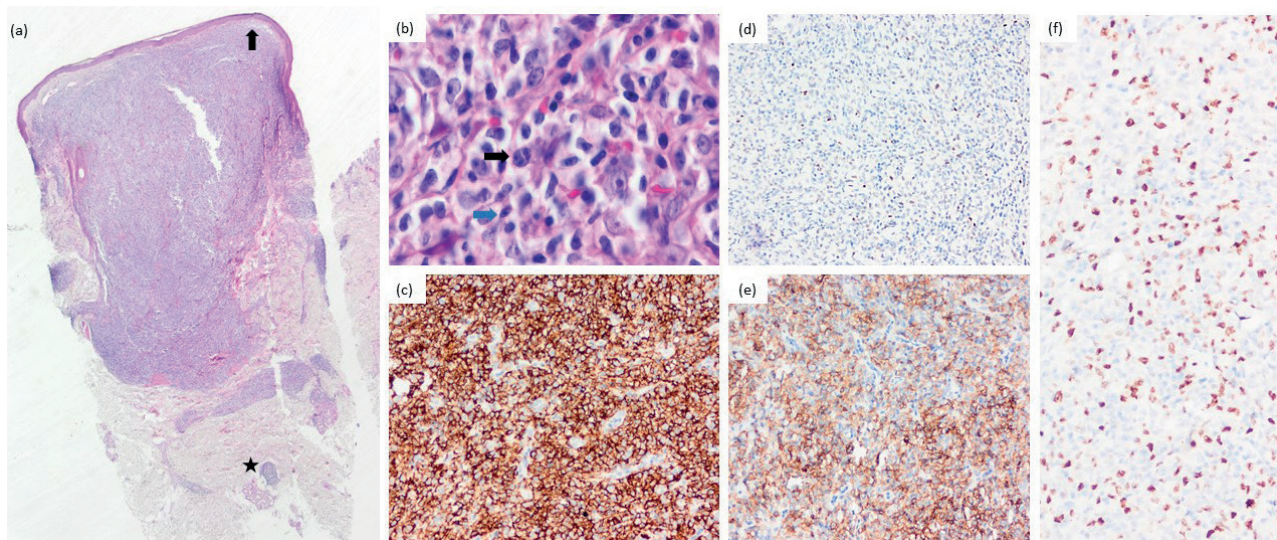


Figure 2. Lymphoid infiltration in a diffuse pattern, leaving a grenz zone (arrow) under the epidermis and progresses up to the subcutaneous fat tissue (asterisk) in focal areas (H&E, x20) (a). The infiltration consists of a polymorphic population. In addition to cells with small-medium size, narrow cytoplasm, coarse chromatin pattern and irregular nuclear contour (centrocytes- black arrow), there are also large nuclear slit/notched cells and cells with smooth nuclear contours in centroblast (blue arrow) morphology with multiple nucleoli (H&E, x1000) (b). In the immunohistochemical studies, the infiltrate is positive with CD20 (CD20, x200) (c), Bcl-2 MUM1 and IgM. Focal weak staining was observed with Bcl-6 (Bcl-6, x200) (d). CD10 and p63 were negative. Expanded diffuse dendritic network is stained with CD23 (CD23, x200) (e). Ki-67 proliferation index was about 30% (Ki-67, x200) (f). Although with solely immunophenotypic findings, diffuse large B-cell lymphoma “leg type” could also be included in the differential diagnosis, with the morphology and the indolent clinical course this entity was excluded and the case was interpreted in favor of a lymphoma of follicular origin.

in centroblast morphology with multiple nucleoli (Figure 2). The immunohistochemical results are shown in Figure 2. Positron Emission Tomography and Computed Tomography (PET-CT) showed F-18 fluorodeoxyglucose uptake prominent in only cervical, axillary, inguinal and abdominal lymph nodes. The leukocyte/lymphocyte and neutrophil count were within normal limits, only iron deficiency anemia was present. Since the PET-CT detected multiple lymph node involvement, he was diagnosed with systemic FL and rituximab therapy was planned.

DISCUSSION

FL is an indolent type of B-cell lymphoma which is derived from germinal center B-cells, centroblasts and centrocytes [1]. It comprises 35% of all Non-Hodgkin lymphomas and 70% of all indolent, low-grade lymphomas [2]. The chromosomal translocation t(14;18)(q32;q21) which leads to the overexpression of BCL2 is generally seen in patients with FL [1]. Cutaneous involvement of systemic lymphomas most commonly appear as pink papule, plaque or nodule formation involving the scalp, trunk, head and neck [3]. Talebi-Liasi et al. [3] presented a case of disseminated FL in the form of an ill-defined reticular patch over the scalp and forehead. Gordon et al. [4] reported another case of systemic B-cell FL, which showed ear involvement in the form an erythematous plaque. Bilateral ear involvement of primary cutaneous marginal zone

lymphoma associated with rheumatoid arthritis, was also reported by Yildirim et al. [5]. Adnexal tumors, histiocytosis, cutaneous deposition disorders such amyloidosis and chronic tophaceous gout may all be considered in the differential diagnoses of the cutaneous lesions of the external ear.

CONCLUSION

We would like to emphasize that cutaneous involvement of systemic B-cell lymphomas should always be considered in the differential diagnoses of asymptomatic papules/nodules of unknown origin presenting upon the ears, especially in elderly patients.

Author contribution

Study conception and design: EB, AK, EÖ, and SA; data collection: EB, EÖ, and SA; draft manuscript preparation: EB and EÖ. All authors reviewed the results and approved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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