

Burkitt lymphoma with unusual granulomatous reaction. A case report

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SUMMARY

Formation of epithelioid histiocytic cell granulomas has been described in the past in various neoplasms, hematologic malignancies included. Among lymphoproliferative disorders such changes are commonly found in Hodgkin lymphoma and T-cell non-Hodgkin lymphomas (NHL), but are rarely described in B-NHL, like Burkitt lymphoma. This report presents a case of sporadic Burkitt lymphoma accompanied by a sarcoid-like reaction without any clinical, laboratory or histological evidence of microorganisms nor sarcoidosis. Using *in situ* hybridization and polymerase chain reaction the presence of the Epstein-Barr virus (EBV) was detected in the analyzed lymphoma cells. EBV demonstrated latency I phenotype as defined by the lack of immunohistochemical positivity of latent membrane protein 1 (LMP1). Cytogenetic investigation using fluorescence *in situ* hybridization uncovered c-MYC mutation and provided indirect indication for the MYC/IgI fusion gene. The lack of EBV positivity in histiocytes indicated the reactive character of the granulomatous reaction in relation to the neoplasm. The role of the granulomatous reaction in the biology and prognosis of Burkitt lymphoma and the function of EBV infection in its development remain to be established.

Key words: Burkitt lymphoma - granulomatous reaction - Epstein-Barr virus - EBV

Burkittov lymfóm s nezvyčajnou granulomatóznou reakciou. Kazuistika

SÚHRN

Výskyt granulomatózne reakcie sprevádzajúcej rôzne druhy nádorov bol už v minulosti opísaný. Spomedzi lymfoproliferatívnych chorôb, vykazuje tvorbu granulómov predovšetkým Hodgkinov lymfóm a T-bunkové non-Hodkinove lymfómy (NHL). V B-bunkových NHL, ako je Burkittov lymfóm, sú granulomatózne reakcie zriedkavé. V tejto práci predstavujeme prípad sporadického Burkittovho lymfómu sprevádzaného tvorbou granulómov pripomínajúcich sarkoidózu. Klinické, laboratórne, ani histologické vyšetrenie nepreukázalo prítomnosť infekčných agens, resp. nesvedčilo pre diagnózu sarkoidózy. *In situ* hybridizáciou a polymerázovou reťazovou reakciou sa v tkanive dokázala prítomnosť Epstein-Barrovej vírusu (EBV). Negativita imunohistochemicky stanovovaného vírusového latentného membránového proteínu 1 (LMP1) spolu s predchádzajúcimi vyšetreniami svedčí v tomto prípade pre typ I latentnej EBV infekcie. Cytogenetická analýza s použitím fluorescenčnej *in situ* hybridizácie odhalila mutáciu génu c-MYC spolu s naznačenou fúziou MYC/IgI. Absencia EBV v histiocytoch podporuje skôr reaktívny charakter granulomatózne reakcie na nádorový proces. Biologický a prognostický význam vzniku týchto granulómov sprevádzajúcich Burkittov lymfóm a úloha EBV infekcie v tomto vzťahu zostáva však nejasná.

Kľúčové slová: Burkittov lymfóm – granulomatózna reakcia - Epstein-Barrovej virus - EBV

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Tumor-related tissue reactions as the formation of epithelioid cell granulomas have been reported in association with several neoplasms (1). Such a sarcoid-like reaction may occur in solid tumors, particularly keratinizing squamous cell carcinoma and seminoma (2). In sarcomas the presence of such histological changes is extremely rare (3). Formation of granulomas has been also described in hematologic malignancies (1), mainly in Hodgkin lymphomas. The incidence of granulomatous reaction in patients with Hodgkin disease is reported to be 13.8% (4), whereas the frequency in non-Hodgkin lymphomas (NHL) ranges from 3.6% (5) to

7.3% (3). From the latest group, sarcoid-like granulomas most commonly occur in T-cell-derived non-Hodgkin lymphomas (6), including a marked number of cutaneous lymphomas like mycosis fungoides and subcutaneous panniculitis-like T-cell lymphoma. Epithelioid cell granulomas are less frequent in B-cell NHL. They have been reported in follicular center cells lymphomas, small lymphocytic lymphomas, large cell lymphomas (5) and in a few cases of sporadic Burkitt lymphoma (6-8). The appearance of these coincidental granulomas has been described in regional lymph nodes either involved or uninvolved by the tumour, in sites of distant metastases and in non-involved organs, respectively (2).

The presented paper reports a case of a patient with sporadic Burkitt lymphoma accompanied by a sarcoid-like reaction, which emerged in lymph nodes nearby the ligamentum hepatoduodenale.

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