1.Title

Diffuse large B-cell lymphoma of the skin with colonization of a haemangioma mimicking intravascular large B-cell lymphoma.

2. Abstract Body (Word Count: 498 words)

Introduction

We describe an unusual case of primary cutaneous diffuse large B-cell lymphoma (DLBCL) occurring as a plaque on the head and neck region, with concomitant intravascular colonization of a hemangioma at a separate, non-contiguous anatomical site. The latter presentation mimics intravascular large B-cell lymphoma (IVBCL) both clinically and histologically, raising the alternative possibility of synchronous occurrence of both lymphomas. This posed a diagnostic challenge with both therapeutic and prognostic implications.

Case Report

A 75-year-old female presented with a rapidly growing nodule on her right cheek of 1 month duration. She was otherwise well with no other systemic features.

On clinical examination, there was a 4.3cm x 2.5cm ulcerated nodule on her right cheek. Incidentally, a 6mm erythematous nodule was noted over her right parietal scalp.

An incisional biopsy performed from the right cheek nodule revealed a dense, diffuse nodular dermal infiltrate of large tumour cells, which extended to the subcutis and surrounded dermal vessels. These tumour cells were diffusely positive for CD20, PAX-5, Mum-1, BCL-6 and BCL2 on immunohistochemistry staining. EBER in-situ hybridization was negative, with no light chain restriction on kappa and lambda stains. The histopathological features and immunophenotype were consistent with DLBCL.

Histological examination of the scalp nodule confirmed the presence of an IVBCL colonizing a hemangioma, characterized by a dermal proliferation of dilated thin-walled vessels (positive on CD31 stain), containing intra-luminal aggregates of large, atypical lymphocytes. Significantly, the immunophenotype of these neoplastic cells were like those from the right cheek nodule.

A staging positron emission tomography scan confirmed the presence of an FDG-avid lesion on her right cheek with no other radiological evidence of systemic disease. No neurological involvement was detected on magnetic resonance imaging of the brain and lumbar puncture.

An initial diagnosis of Stage IV IVBCL with concomitant cutaneous DLBCL was made. She received six cycles of R-CHOP (Rituximab, Cyclophosphamide, Vincristine and Prednisolone) and demonstrated a surprisingly favorable clinical response with complete resolution of her right cheek nodule both clinically and radiologically (on repeat imaging). Hitherto, her disease remains in complete remission two years post-treatment.

Discussion

Considering the similar immunophenotype shared between both tumours (indicating the same disease process), patient's paucity of systemic involvement and her favourable response to chemotherapy, we opined that this was likely an atypical case of cutaneous DLBCL demonstrating angiocentricity with intravascular colonization of a distant cutaneous haemangioma, rather than a coincidental synchronous occurrence of DLBCL and IVBCL (which conventionally presents with disseminated disease and is associated with poor response to chemotherapy). The colonization of haemangiomas by IVBCL is a well-documented phenomena that is not typically observed with DLBCL, and we hypothesize that neoplastic cells of DLBCL may also demonstrate expression of adhesion molecules which enable adherence to endothelium, thereby enabling extravasation from blood vessels and distant micro-metastasis. Whilst DLBCL and IVBCL have been regarded as separate entities with distinctive clinicopathological and prognostic features, the characteristics of this case may suggest the that both conditions are closely related and possibly constitute a phenotypic spectrum of a single disease.

3. **Learning Objective**

We describe an atypical case of diffuse cutaneous large B-cell lymphoma (DLBCL) with colonization of a cutaneous haemangioma at a separate anatomical site, which posed an initial diagnostic dilemma as to whether this was an unusual manifestation of DLBCL or if there was a coincidental synchronous occurrence of intravascular large B-cell lymphoma (IVBCL). Through this case, we hope to highlight to participants the important clinic-pathological features contributing to the final diagnosis, as well as the proposed pathomechanisms underlying this clinical observation.