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Case report

Diffuse large B cell lymphoma coexistence with systemic mastocytosis

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ABSTRACT

Systemic mastocytosis is a rare disease and characterized by excessive mast cell accumulation in one or multiple organs. One subtype of systemic mastocytosis is systemic mastocytosis-associated clonal hematological non-mast cell lineage disease (SM-AHMND), which indicates concurrent evolution of two separate clonal entities, one consisting of mast cells and one as a second hematological as well as non-mast cell origin disease. When SM-AHMND is diagnosed, bone marrow examination is essential for the initial approach, because marrow is almost universally involved in adult mastocytosis and it facilitates detection of a second hematological neoplasm. Myeloid neoplasm is reported to be the most prevalent associated clonal hematological non-mast cell disease. Treatment strategy and outcome for SM-AHMND is dependent on hematological non-mast cell lineage disease. Herein, we have presented a case report of diffuse large B cell lymphoma coexisting with systemic mastocytosis where the patient underwent successful chemotherapy leading to extended survival duration.

1. Introduction

Mastocytosis indicates a group of disorders characterized by excessive mast cell accumulation in one or multiple organs and is a type of myeloproliferative neoplasm disorder, according to the 2008 World Health Organization (WHO) classification of tumors of hematopoietic and lymphoid tissues Non-Hodgkin lymphoma.[1] Mastocytosis is heterogeneous and can involve multiple sites, ranging from skin lesions, which may spontaneously resolve to highly aggressive mast cell leukemia associated with multiple organ failure. It is a rare disease and its exact incidence is unknown. Subtypes of mastocytosis are recognized primarily through clinical presentation and distribution of disease. Mast cell infiltration confined to the skin is cutaneous mastocytosis, whereas systemic mastocytosis (SM) is regarded as involvement of at least one extra-cutaneous organ with or without the presence of skin lesions.[2] Clinically, SM is classified as indolent SM (ISM), aggressive SM (ASA), SM with associated clonal hematological non-mast cell lineage disease (SM-AHMND), and mast cell leukemia (MCL).[1] ISM, the most common subgroup among SM, usually refers to symptoms resulting from mast cell degranulation, mediator release, allergies or anaphylaxis and no evidence of extra-cutaneous organ dysfunction or failure,[3] while ASM indicates the presence of extra-cutaneous organ dysfunction or failure caused by mast cell infiltration.[4] On the other hand, SM-AHNMD is the second most common SM subgroup and myeloid neoplasm is reported most frequently as hematological non-mast cell lineage disease.[3] Bone marrow (BM) examination is suggested as part of the initial diagnostic work-up for SM, because BM is almost universally involved in adult mastocytosis. Furthermore, BM examination also allows for

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the detection of a second hematological neoplasm. Since SM is an uncommon disease, SM coexistent with lymphoid neoplasm is extremely rare. Here, we present an interesting case of SM coexistent with diffuse large B cell lymphoma (DLBCL).

2. Case report

A 51-years-old man visited our hospital presenting with a palpable mass over the right inguinal area which had persisted for 3 months. This lesion was approximately 3–4 cm, firm and had progressively enlarged. Laboratory data showed no remarkable anomaly except elevated lactate dehydrogenase (265 U/L). The patient denied experiencing any significant weight loss, poor appetite, night sweats, or sustained fever. After discussing his circumstances with the hematologist in our outpatient department, the patient agreed to undergo surgical excision biopsy. Subsequent histological analysis revealed DLBCL and he underwent further computed tomography with contrast, and BM examination for complete staging. The computed tomography showed right lumbar, iliac and right inguinal lymphoadenopathies, and his clinical Ann Arbor staging was designated as stage II. Further BM examination disclosed diffuse proliferation of large atypical CD20-positive lymphoid cells using hematoxylin and eosin staining (Fig. 1) and immune-histochemical stain (Fig. 2). This is compatible with diffuse large B cell lymphoma, and the patient's staging was upgraded to stage IV. However, at the low power field of 40X, multiple well-circumscribed aggregates of polygonal cells with pale cytoplasm were observed in the bone marrow. At the high power field designation of 200X, these cells were oval to spindle-shaped and contained a moderate amount of pale cytoplasm (Fig. 3), which showed positive for tryptase (Fig. 4) and CD25 (Fig. 5) through immune-histochemical evaluation. The picture was compatible with systemic mastocytosis with associated clonal hematological non-mast cell lineage disease. Thus, the final diagnosis for this male patient was DLBCL, with right lumbar, iliac and right inguinal lymphoadenopathies and BM involvement, Ann Arbor stage IVA, International Prognostic Index 3, coexisting with SM. In March 2013 he began a course of immune chemotherapy with rituximab, cyclophosphamide, vincristine and prednisolone (R–CHOP), which was the standard treatment for DLBCL. The treatment consisting of 6 courses of R–CHOP was completed in September 2013, and a follow-up integrated Positron emission tomography and computerized tomography (PET-CT) scan revealed nearly complete remission of DLBCL. The port-A was removed in April 2015, as scheduled, and the patient was followed-up regularly in the outpatient department presenting in routine stable condition.

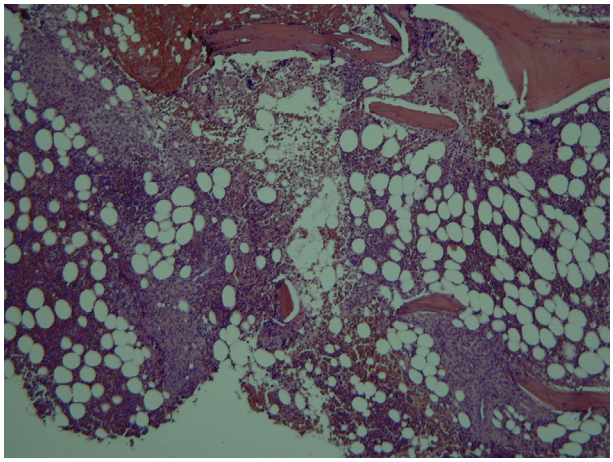

Fig. 1. Diffuse large B cell lymphoma with bone marrow involvement in hematoxylin and eosin stain.

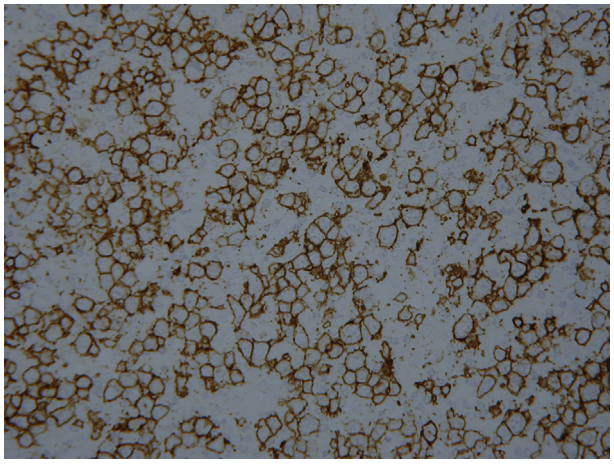

Fig. 2. CD-20 immuno-histochemical stain demonstration on diffuse large B cell lymphoma.

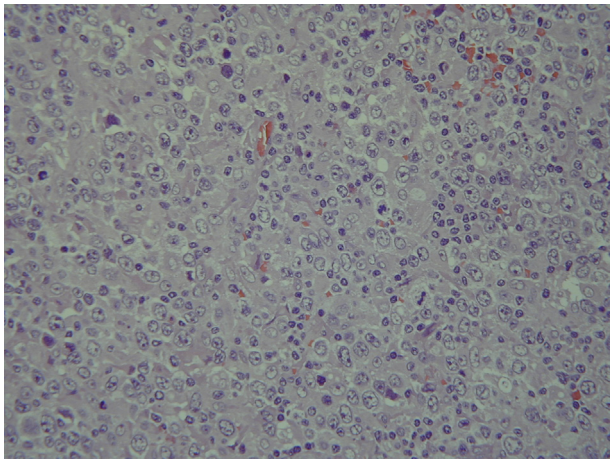

Fig. 3. Mast cell aggregation and infiltration, these cells were oval to spindle-shaped and containing moderate amount of pale cytoplasm.

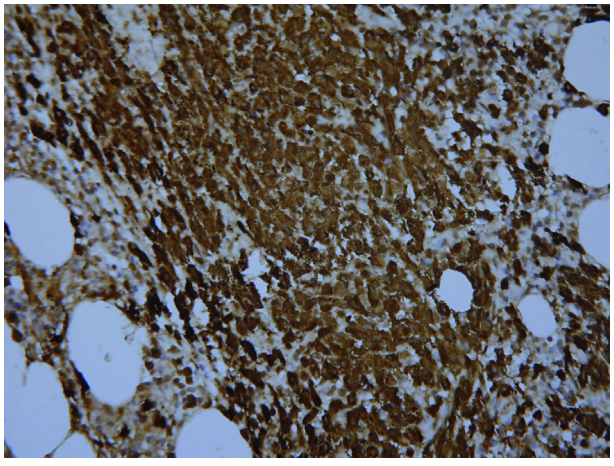

Fig. 4. Tryptase stain demonstration on mast cell.