Concomitant occurrence of multiple myeloma with diffuse large B-cell lymphoma

Editor,

Diffuse large B-cell lymphoma (DLBCL) is the most prevalent subtype of non-Hodgkin lymphoma (NHL), with a diverse pathobiology while multiple myeloma (MM), a neoplastic disease involving the clonal proliferation of terminally differentiated plasma cells, is the second most common hematologic neoplasm. The simultaneous presentation of these neoplasms is exceedingly rare, and we present such a case of synchronous DLBCL and MM in a male patient.

A 65-year-old male presented with testicular lump of 4 months duration, loss of testicular sensation, and no history of fever, weakness, bone pain, weight loss, or anorexia. Examination revealed a mobile, painless scrotal swelling, with associated right inguinal lymphadenopathy. There were no other palpable lymph node and no hepatosplenomegaly. Routine laboratory investigations showed no significant abnormality, except elevated serum lactate dehydrogenase and C-reactive protein levels. Computerized tomographic scan of the abdomen showed a single scrotal mass with right inguinal lymphadenopathy. Orchidectomy and block dissection of right-sided inguinal lymph nodes were performed, and excised tissue sent for biopsy. Grossly, a tumor mass was seen completely replacing the testis. Microsections showed sheets of large transformed lymphoid cells with prominent nucleoli and moderate to a large amount of amphophilic cytoplasm. Tumor infiltration into the capsule, tunica albuginea, and spermatic cord, as well as tumor metastasis to lymph nodes, was present.

Immunohistochemistry (IHC) of the tumor cells showed CD20 expression and was immunonegative for CD3, CD5, CD10, and CD23. MIB-1 labeling index was just above 50%. The case was diagnosed as DLBCL. Repeat IHC on testicular biopsy showed that the tumor cells were CD38 negative, ruling out the presence of an extramedullary plasmacytoma. Serum protein electrophoresis and immunofixation electrophoresis confirmed elevated levels of IgA kappa monoclonal proteins. Serum beta 2-microglobulin level was 2.2 mg/dL.

A final diagnosis of synchronous DLBCL and MM, IgA kappa, international staging system stage I was made. The patient showed clinical improvement after two cycles of R-CHOP chemotherapy but was lost to subsequent follow-up.

DLBCL is a heterogeneous group of neoplasms, comprising 30–40% of adult NHLs. This cytologically diverse group shows variable expression of pan-B-cell markers such as CD19, CD20, CD22, or CD79a, and a high proliferative index, with Ki-67 labeling >40% in most cases. IHC staining algorithms can be used to differentiate DLBCL into prognostically distinct groupings. MM is associated with clonal proliferation of plasma cells in the bone marrow and production of a monoclonal protein (IgG/IgA). Both DLBCL and MM arise from proliferation of a B-cell precursor, and initiation and progression of these neoplasms are driven by underlying genetic abnormalities. DLBCL commonly shows BCL2 and BCL6 gene abnormalities. MM often has a complex karyotype, with numerous chromosomal abnormalities and genomic heterogeneity.

MM has been associated with an increased risk of secondary malignancies, but the coexistence of lymphomas with myeloma is relatively rare, and the pathobiologic relationship of DLBCL with MM is not yet well understood. An extensive retrospective study showed the development of myeloma in 6 out of 4165 cases of B-cell lymphoma and 1 case of B-cell lymphoma developing in 804 patients with myeloma. Two of the patients developed
DLBCL, and the development of secondary malignancies was metachronous in 6 cases.[5]

Initial reports postulated a common clonal cell of origin for the coexisting malignancies, but further studies have demonstrated disparate clonal evolutions in concomitant malignancies, using immunoglobulin light and heavy chain isotype analysis or genotypic studies. These separate clonal origins indicate that DLBCL and myeloma evolve independently and exclude secondary transformation of a B-cell clone.[6]

Management of coexisting B-cell neoplasms poses a problem in the absence of uniform treatment guidelines. R-CHOP chemotherapy remains the treatment of choice in DLBCL,[10] Dexamethasone, cyclophosphamide, etoposide, and cisplatin regimen has been used in an earlier reported case of synchronous myeloma and DLBCL. After initial remission, there was relapse within 3 years.[2] In the current report, the initial clinical response to two cycles of R-CHOP chemotherapy could not be further evaluated as the patient was lost to follow-up.

The presence of a monoclonal gammopathy in serum or urine in a B-cell lymphoma warrants further investigation to rule out the possibility of myeloma. Urgent and intensive chemotherapy is required for such cases as they tend to have worse prognosis.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES


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